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Maharashtra College of Radiology

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"Maharashtra College of Radiology" [MCR] is an academic wing of the MSBIRIA established in 2020. The Maharashtra College of Radiology (MCR) is a distinguished institution dedicated to the education, training, and professional development of radiologists and imaging specialists in Maharashtra, India.

As a key academic body, MCR focuses on advancing the field of radiology through comprehensive educational programs, workshops, and continuous medical education (CME) activities by organizing conferences, symposia, and seminars that bring together experts from across the country and around the world.

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The Journal of Applied and Clinical Radiology is the official publication of the Maharashtra State Branch of the Indian Radiological and Imaging Association (MSBIRIA) and the Maharashtra College of Radiology (MCR). It serves as a platform for radiologists, radiology students, researchers, and healthcare professionals to share innovative research, clinical findings, and advancements in the field of radiology. It is a peer-reviewed platform dedicated to advancing knowledge in the field of radiology. It promotes continuous learning and professional development within the radiology community by providing high-quality articles that bridge the gap between research and clinical practice in radiology.

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Journal of Applied and Clinical Radiology: Vision to Reality

Anand Hatgaonkar

The Maharashtra State Branch of the Indian Radiological and Imaging Association (MSBIRIA) and the Maharashtra College of Radiology (MCR) proudly present to you the first issue of the "*Journal of Applied and Clinical Radiology*." This inaugural issue marks the beginning of a significant chapter in our shared journey to advance the field of radiology, offering a platform that supports innovation, fosters professional growth, and disseminates valuable knowledge.

The journal is published at a very crucial time when the practice of radiology is undergoing a great change, with advances in technology and increasing knowledge about disease processes. The main aim of this journal is to offer a forum for radiologists, researchers, and medical students a free platform to share their original research, case reports, and reviews in radiology. It is an attempt to close the gap between academic research and clinical practice to foster evidence-based practice and ongoing professional development among members of the radiological community that improves patient care.

In this inaugural issue, we are delighted to showcase articles submitted by the radiology community that challenge the edges of prevailing wisdom, such as in-depth case reports and case series with intricate clinical presentations and novel insights into the imaging findings. Every article has gone through a rigorous peer-review process to establish that it is of the highest academic integrity and clinical usefulness.

The challenge of starting this new academic venture stems from keeping up with new technology and meeting the timeline for publication. Confronting these challenges through continuous learning, teamwork, and support systems, we set off on this new academic adventure with publication partner Wolters Kluwer/Medknow.

The release of this journal would be impossible without giving credit to the senior radiology fraternity, Dr. Sandeep Kavthale, president elect IRIA, and all MSBIRIA members for trusting me and assigning me this herculean task of setting up this journal. Dr. Pradeep Gandhi, Dr. Pravin Sagole, Prof. Dr. Shailesh Sangani, and the MSBIRIA and MCR teams to actively encourage the launch of this new academic platform for the radiologists. It needs a lot of effort and complete dedication from day 1 of ideating the journal to narrowing its scope and standard and making this dream a reality. It is through the commitment of authors, reviewers, members of the editorial board, and the publication team that we were able to bypass these obstacles and make this dream a reality.

In addition, we understand that the success of this journal is dependent on ongoing improvement. Going forward, we are committed to making the processes and content of the journal better, guided by your feedback and needs as readers. Your participation is important, and we invite you to read and gain from the research shared and contribute your work and thoughts.

As we begin this endeavor, I offer our sincerest thanks to all who have worked towards making this vision a reality. If I

Editor in Chief, Journal of Applied and Clinical Radiology

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have inadvertently forgotten to mention anyone, then it must be regarded as my fault, and I wholeheartedly apologize.

We invite you to engage actively with this publication, share your perspectives, and participate in shaping a future where radiology continues to grow and thrive. Together, let us overcome challenges, support one another, and drive the field toward greater achievements.

With best wishes,

Dr. Anand Hatgaonkar
Editor in Chief, JAACR,
Professor & HOD Radiodiagnosis,
Datta Meghe Medical College, Nagpur.

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Fetus in fetu: A series of clinical cases and radiological insights

M. K. Jegannivas, Manisha Joshi, Dev Shetty, Tejaswi Bhukya, Mayuri Jugulkar

Abstract:

BACKGROUND: Fetus in fetu (FIF) is an exceptionally rare and intriguing congenital anomaly characterized by the presence of a malformed, parasitic twin within the body of a host twin. This condition poses unique diagnostic and therapeutic challenges, particularly in the realm of radiology. This condition is usually confused with teratoma and cystic meconium peritonitis. This study analyses and documents the various presentations and imaging features across different modalities in a series of cases presented at our institution.

OBJECTIVE: To review and summarize the imaging findings of FIF cases, highlighting the diagnostic features and differential diagnosis.

METHODS: We conducted an analysis of different radiological studies from a series of patients diagnosed with FIF at our institution. Cases were selected based on imaging findings consistent with the condition. Data including X-ray, ultrasound, computed tomography (CT), and magnetic resonance imaging (MRI) were analyzed to elucidate characteristic features and variations.

RESULTS: The case series includes four patients diagnosed with FIF, ranging from the antenatal period till adulthood. Imaging provided detailed anatomical localization and differentiation from other abdominal masses, such as teratomas. Key findings in the radiological imaging modalities included the presence of well-defined, encapsulated masses with varying degrees of organ differentiation and ossification.

CONCLUSION: Radiological imaging plays a crucial role in the diagnosis and management of FIF. X-ray, ultrasound, CT, and MRI each contribute unique insights into the condition's presentation and assist in differentiating it from other abdominal anomalies. Early and accurate imaging diagnosis is essential for effective surgical intervention and optimal patient outcomes. Continued research and case documentation will enhance our understanding and improve management strategies for this rare anomaly.

Keywords:

Fetus in fetu, parasitic twin, teratoma

Introduction

The Fetus in Fetu (FIF) is a rare entity in which one malformed vertebrate fetus is enclosed within the body of its twin. A spectrum of presentations has been described in the literature, although the embryonic pathogenesis and differentiation from a teratoma have not been well established.[1,2] The FIF has an incidence of 1 in 500,000, with a 2:1 male predominance.[3] The exact embryogenesis of FIF is controversial. The major conception is that this condition is secondary due to anomalous embryogenesis in a diamniotic monochorionic twin pregnancy in which a malformed monozygotic twin lies within the body of its fellow twin. Another school of thought is that it represents a highly organized teratoma.[4,5] Most occur in the abdomen, around 80% within the retroperitoneal cavity, while other rare locations include the cranial cavity, oral cavity, mediastinum, back, pelvis, sacrococcygeal region, and scrotum.[6] This condition (FIF) poses unique diagnostic and therapeutic challenges,

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particularly in the field of radiology. The FIF is usually confused with teratoma and cystic meconium peritonitis. In this study through a series of cases; we analyze and document the various presentations and imaging features of the FIF in various modalities and how these unique findings help radiologists in increasing their degree of confidence in diagnosing FIF.

Case Reports

Case 1

Our first case is a 4-day-old male infant presented with a large swelling over the back in the left paramedian area since birth; it is mostly covered with skin and has few appendages. The child is a term baby born via cesarean section at 39 weeks of gestation due to an abnormal presentation. On examination, the lump was round and tender with external morphological features of nipple-areolar complex and dysplastic hand with digits; it is associated with a small lumbosacral meningomyelocele [Figure 1]. Laboratory investigation revealed serum α-fetoprotein (AFP) of 2800 ng/ml (normal: 0–5.8 ng/ml) and β-human chorionic gonadotropin of 4.26 mIU/l (normal: 0–5.3 mIU/l). All other values, including complete blood count, urea, electrolytes, and liver function test, were within normal ranges.

The roentgenogram of the baby [Figure 2] showed soft-tissue swelling over the left parasacral region with few ossified bone-like structures. In view of the presence of bone-like corticated structures resembling long bones and vertebral bodies, the possibility of FIF was conveyed to the concerned clinicians.

The baby further underwent contrast-enhanced tomography of the abdomen, which revealed a mildly enhancing ill-defined partially calcified soft-tissue mass lesion with ossified bones in the left paramedian region over the back (left paraspinal region). Splaying of the lumbosacral posterior vertebral elements (from L4 level) was noted with herniation of the neural tissue and meninges into the subcutaneous plane. There were no fat density structures noted within the lesion; the intraspinal extension of the mass lesion was not seen [Figure 3].

The patient was then operated on, the mass was excised, and tissue samples were sent for histopathology, which later came as lumbosacral myelomeningocele with FIF with a vestigial tail.

Case 2

The second case was a 17-year-old male who presented to the surgery outpatient department (OPD) with a clinical history of a lump in the abdomen since childhood that was gradually increasing in size; associated with a dull aching type of pain that was on and off in nature. There was no history of fever, weight loss, altered bowel habits, or urinary symptoms; he had no other clinical complaints. On abdominal examination, as done in surgery OPD, there was a lump in the umbilical and left lumbar regions of the abdomen. It was hard to firm in consistency on palpation and was not moving with respiration.

The roentgenogram of the patient revealed a soft-tissue density lesion in the left lumbar region intermixed with a few discrete areas of calcification within, showing the appearance of limb bones and vertebrae [Figure 4]. The possibility of organized teratoma/FIF was conveyed.

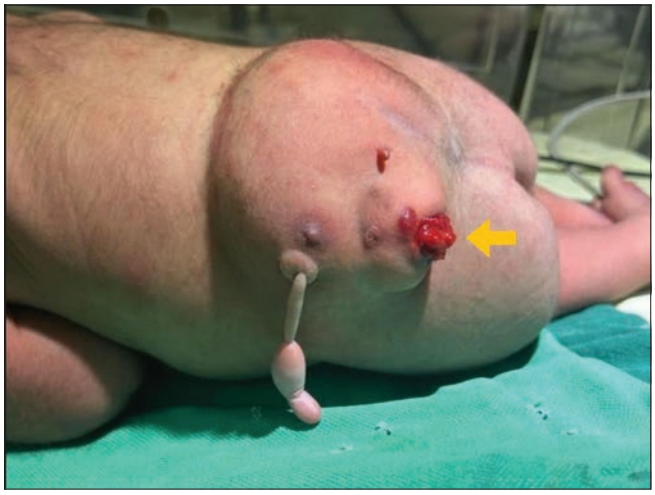

Figure 1: Clinical Photograph of the baby. Image of a 4-day-old male infant showing a large mass in the lumbosacral region with limb-like projection and a well-formed nipple-areolar complex on its surface; Note the reddish lobulated soft tissue in the mass which represents underlying spinal dysraphism (yellow arrow)

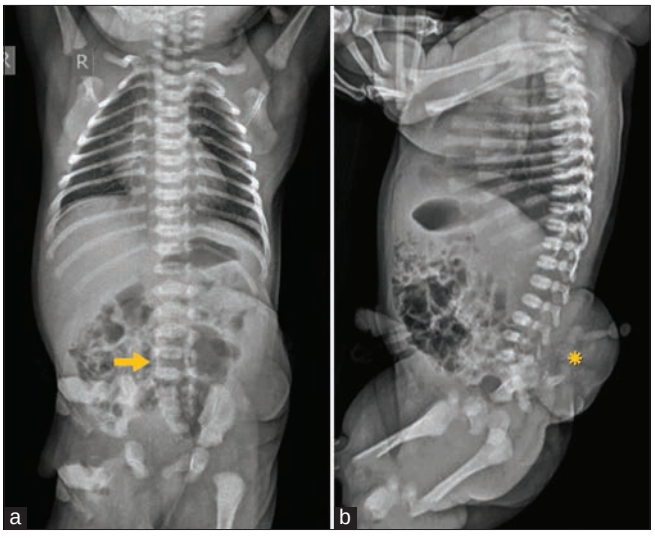

Figure 2: Infantogram anteroposterior (AP) and Lateral. (a and b) Infantogram AP and lateral views showing ill-defined soft-tissue density mass lesion in lumbosacral region (yellow asterisk) with few limb like areas of ossification within the mass; Note the defects in the posterior elements of lower lumbar and sacral vertebrae (yellow arrow)

The patient underwent contrast-enhanced computed tomography (CT) of the abdomen for further evaluation, which showed a large well-defined heterogeneous encapsulated soft-tissue mass in the left lower anterior pararenal space of the retroperitoneum; the mass was seen abutting the aorta, left common iliac artery, and left psoas muscle posteriorly, it showed intermixed areas of fat attenuation, foci of calcifications. The malformed calcified areas were resembling fetal spine and limb bones [Figure 5].

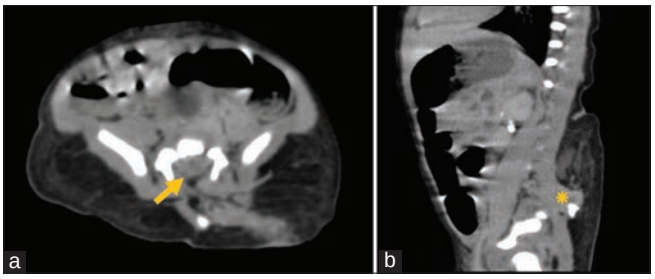

Figure 3: Axial and reformatted sagittal postcontrast computed tomography images. (a and b) Axial and sagittal images showing mildly enhancing soft-tissue lesion with ossified bones (yellow asterisk) in the left para median region over the back with splaying of posterior vertebral elements in the lumbosacral vertebrae (from L4 level) with posterior herniation of neural tissue and meninges (yellow arrow)

Case 3

A 28-year-old primi gravida at 26 weeks of gestation presented to our institution for the first antenatal ultrasonogram, which revealed a viable intrauterine fetus with a well-defined hypoechoic encapsulated mass lesion in the fetal abdomen in the left upper quadrant abutting the fundic bubble. The mass had a hypoechoic rim and a vascular supply mostly from the aorta. Multiple linear short and long hyperechoic areas were clustered in the center of the mass, showing some distal acoustic shadowing, which could probably represent long and short bones; thus, the possibility of FIF was raised against mature teratoma. No other congenital anomalies were detected in the fetus [Figures 6 and 7]. The family history was negative for congenital malformations, and there was no history of medication or any teratogenic drug use during pregnancy.

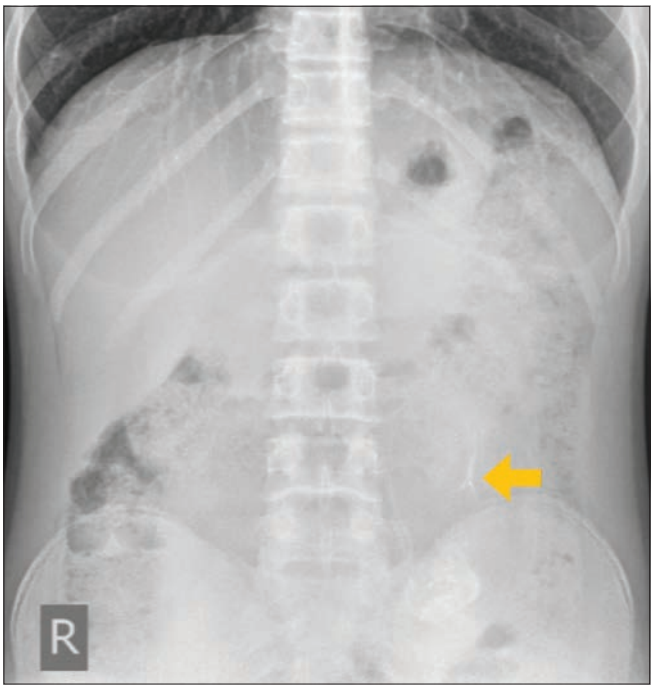

Figure 4: X-ray abdomen anteroposterior erect. X-ray showing well-defined calcified abdominal soft-tissue density mass with some components favoring fetal parts in the left lumbar region extending into the pelvis (yellow arrow)

The patient delivered a female baby at 38 weeks of gestation. After delivery, the baby underwent postnatal CT scan of the abdomen on day 3 of life, which revealed a large heterogeneous soft-tissue mass with fat attenuation and dysplastic fetal bones in the retroperitoneal region adjacent to the left kidney, mildly displacing the bowel loop, and visceral organs [Figure 8]. The diagnosis of FIF was made. The baby underwent resection and the histopathological report further confirmed the diagnosis.

Case 4

A 26-year-old female came to the gynecology OPD with complaints of dysmenorrhea and menorrhagia and was advised ultrasonogram of the pelvis which revealed a bulky uterus with few intramural fibroids, magnetic resonance imaging (MRI) of the abdomen and pelvis was requested for surgical planning.

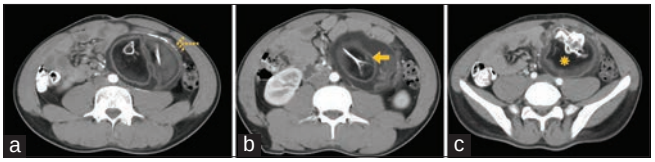

Figure 5: Axial postcontrast computed tomography images of the abdomen. (a-c) Image showing a well-encapsulated mildly enhancing lesion in the retroperitoneal region with areas of fat attenuation (yellow asterisk), foci of calcifications, and malformed calcified areas resembling fetal spine and limb bones (yellow arrow). The mass is displacing the bowel loops anteromedially and close to the aorta and left common iliac artery. Note the thin rim of peripheral calcification in the anterior wall of the mass (dotted yellow arrow)

MRI revealed a bulky heterogeneous uterus with few fibroids; the bilateral ovaries were normal [Figure 9]. On abdominal screening, there was an incidental finding of a large T1 and T2 hyperintense lesion in the left suprarenal region with few cystic areas (T2 hyperintense and T1 hypointense) and areas of calcifications (T1 and T2 hypointense), it was seen displacing the left kidney inferiorly. The calcifications were well-defined and appeared to have the shape of vertebral elements. There was suppression of signal intensity on fat suppression sequences [Figure 10] and no signal drop in the out-of-phase images, signifying the presence of macroscopic fat and no microscopic fat. The solid component showed no restricted diffusion [Figure 11].

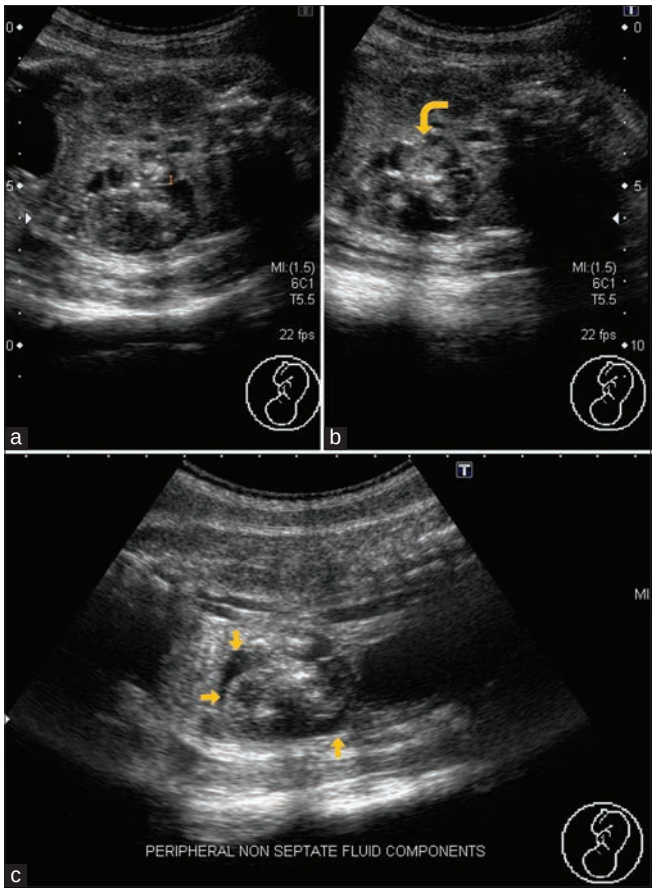

Figure 6: Antenatal sonogram of fetus at 26 weeks. (a and b) Ultrasonogram showing heterogenous hypoechoic encapsulated mass lesion in the fetal abdomen in left upper quadrant abutting fundic bubble and mildly displacing left kidney with multiple linear short and long hyperechoic areas clustered in the center of the mass showing some distal acoustic shadowing (yellow curved arrow)–probably represent long and short bones (c) Sagittal orientation showing peripheral fluid-filled cystic portion with no intervening septae (yellow arrows)

The left adrenal gland was not seen separately from the lesion, so the possibility of the adrenal FIF over the adrenal myelolipoma and the adrenal teratoma was made. Later, the patient underwent surgery and the mass lesion turned out to be a malformed parasitic twin (FIF).

Discussion

FIF is a rare developmental abnormality with < 200 cases reported in the literature,[7] and it has a slight male preponderance.[3] FIF occurs secondary to abnormal embryogenesis in a monochorionic diamniotic pregnancy in which a malformed parasitic twin is found inside the body of its partner as an abnormal fetiform mass.[8-10]

FIF is diagnosed in various age groups with a predominance in infancy; in the majority of cases, it is diagnosed younger than 18 months of age, with very few cases reported in adults. Among the four patients presented above, the age of presentation ranges from the antenatal period to adulthood. To the best of our knowledge and review, there are only a few cases reported in the adult population.[10-12]

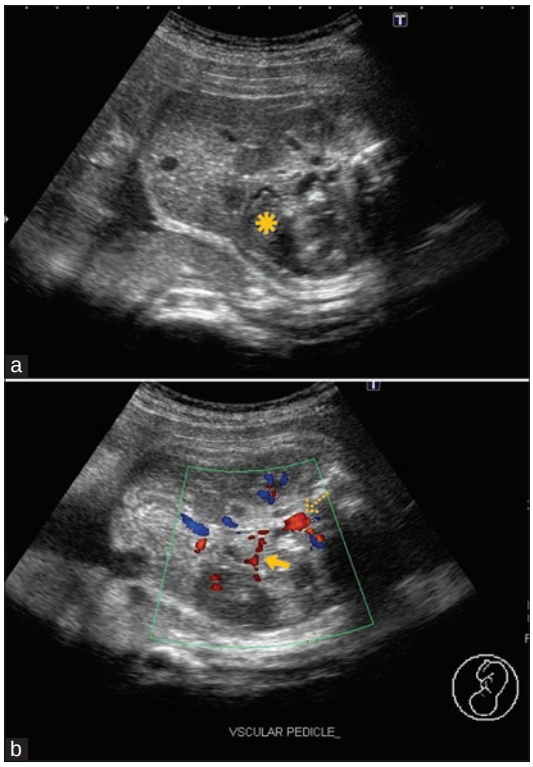

Figure 7: B mode and color Doppler images: (a and b) Encapsulated left upper quadrant mass (yellow asterisk) in the fetal abdomen with vascular supply (yellow arrow) likely arising from the aorta (yellow dotted arrow)

The diagnostic difficulties and differentials of the FIF depend upon the age and the location of the presentation of this condition. Most cases are diagnosed in the antenatal period and confirmed in the immediate postnatal period. Only a few cases are seen in adulthood presenting with symptoms of mass lesions (as seen in cases 1 and 2), and few are found as incidental findings (as seen in case 4).

The various imaging modalities, from simple radiography and ultrasound to advanced cross-sectional imaging techniques like CT and MRI, can help in the diagnosis of FIF with varying degrees of confidence. The soft-tissue density lesion with the presence of fetal bony parts like vertebra and long bones in the abnormal locations in the radiograph is the initial clue in the diagnostic evaluation of this condition. The presence of abnormal bones was seen in cases 1 and 2.

The FIF is commonly misdiagnosed as teratoma in the antenatal period. FIF and well-formed teratoma having all three germ layer components is a matter of dispute for their independent existence. "Willis criteria" explain the differences between the two based on the axial skeleton with vertebral axis development (explaining embryological development passing through the stage

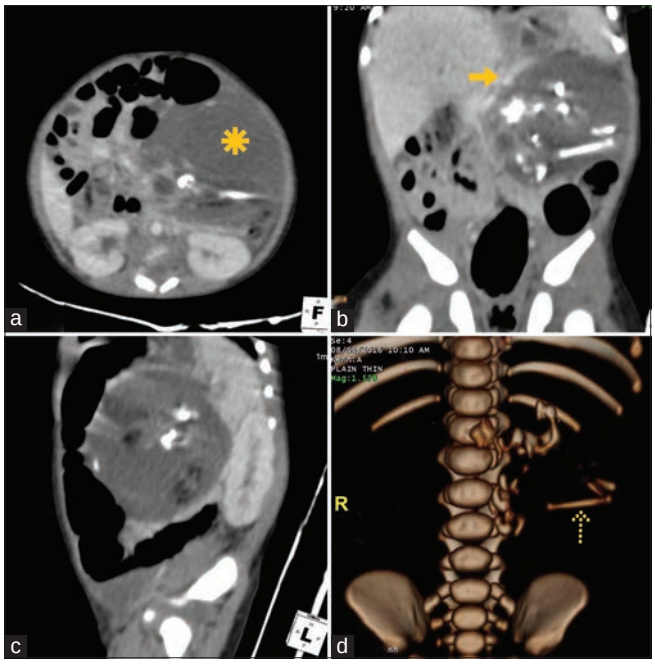

Figure 8: Postnatal contrast-enhanced computed tomography abdomen on day 3 of life. (a-c) axial, coronal, and sagittal reconstructed images of the abdomen showing a large heterogeneous mildly enhancing soft tissue mass with fat attenuation and malformed fetal bones in the intraperitoneal region anterior to the left kidney, mildly displacing the small bowel loops and visceral organs (yellow asterisk). Note the mass displacing the superior mesenteric and splenic vessels (yellow arrow). (d) Volume-rendered image showing malformed fetal bones (yellow dotted arrow)

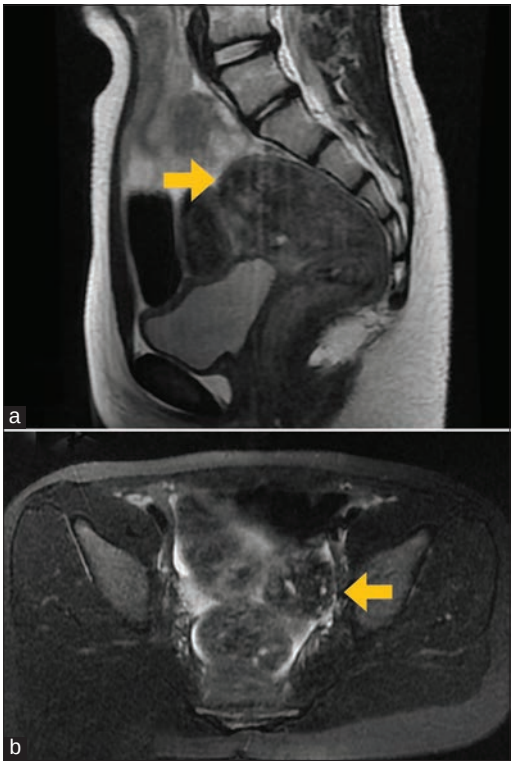

Figure 9: Magnetic resonance imaging Sagittal T2 and axial T2 fat saturated images of pelvis. (a and b) Images showing bulky heterogeneous uterus with well-defined fibroids in the fundus and posterior wall with few cystic areas within (yellow arrows)

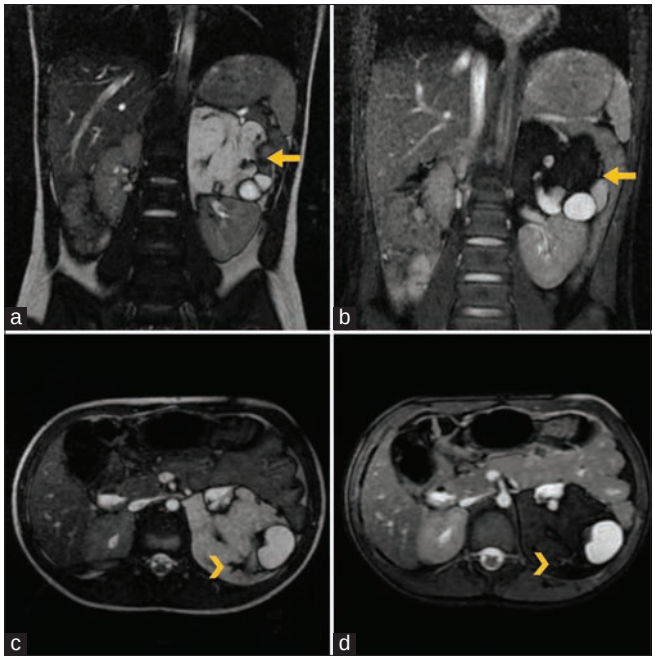

Figure 10: Magnetic resonance imaging two-dimensional fast imaging employing steady-state acquisition (2D FIESTA) and 2D FIESTA FAT-SAT (Fat Saturated) images. (a-d) Coronal and axial 2D FIESTA and 2D FIESTA FS images showing predominantly fat-containing solid lesion (yellow arrow) in the left suprarenal region displacing the left kidney inferiorly with few cystic (T2 hyperintense) areas and bony fragments resembling vertebrae (yellow arrowhead). Note left supra renal gland is not visualized separately

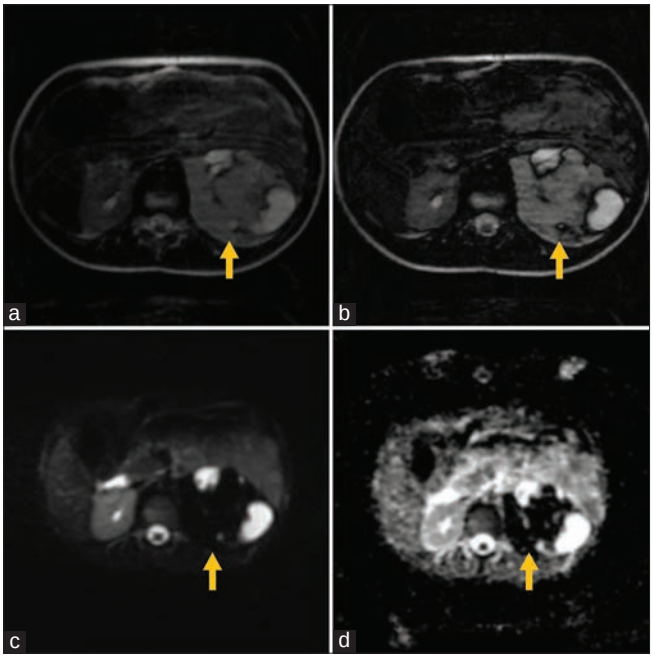

Figure 11: Magnetic resonance imaging in phase, out phase, diffusion-weighted imaging, and apparent diffusion coefficient (ADC) map images. (a and b) No signal drop in the in-phase and out-phase images, signifying the absence of microscopic fat (yellow arrows). (c and d) Diffusion-weighted images (b value 500) and corresponding ADC map showing no significant diffusion restriction (yellow arrows)

of the primitive streak) and an appropriate arrangement of other organs and limbs with respect to the axis in FIF.[13] To be called FIF, the mass must demonstrate true organogenesis. Nonvisualization of the vertebral axis on radiography or CT scan does not exclude FIF as the pathologist can see it.

FIF masses show varying degrees of organ system differentiation and deformity.[14] The FIF usually presents as a fetiform osseous mass, often in the abdomen of its host, with the retroperitoneum being the most common site (almost 80%). In three out of four cases in this series, the location is the retroperitoneum (including the adrenal gland). Different organs can be seen in FIF, including the vertebral column, limbs, central nervous system (CNS), gastrointestinal tract, vessels, genitourinary tract, and heart;[15,16] however, it is extremely rare in the pelvis, scrotal sac, sacrococcygeal region, mesentery, right iliac fossa, and cranial cavity.[15] In one of our cases, the FIF is seen located in the lumbosacral region associated with Spina bifida. In contrast, a teratoma is most commonly found in the sacrococcygeal region, gonads, mediastinum, and CNS. FIF and teratoma have similar sonographic features on ultrasound examination; hence, the risk of misdiagnosis always prevails.

To facilitate the distinction between the FIF and teratoma there are few specific findings in ultrasonogram. In FIF, the mass is divided into two parts: the peripheral fluid-filled cystic portion and the central solid portion "floating" within. The cystic portion is usually not divided by a septum. A teratoma is different in that it is often a multi-loculated cystic mass or a mixed mass of solid and cystic portion without clear border. The ultrasonogram findings in the case 3 of our study also revealed a mass lesion with peripheral cystic anechoic areas and central relatively solid hyperechoic areas with calcifications favoring FIF over teratoma. Similar findings are mentioned in a case report published by Kehal *et al*.[17]

Another possible differential is cystic meconium peritonitis. This condition is rare, and the most common site of occurrence of cystic meconium peritonitis is intra-abdominal (peritoneal), followed by the hollow viscus organ. The percentage of prenatally diagnosed cases using sonograms is very low; the presence of free air or ascites are some additional findings that favor this diagnosis. With time, the formation of loculated pseudocysts can be seen.[17]

A CT scan can give a more accurate diagnosis and define the relation of the FIF with the other intraabdominal structures. In addition, CT three-dimensional reconstruction can completely display the axial bone system and the limbs in the FIF, which are the core in the diagnosis as pointed out by Willis and Lewis.[12] It also facilitates the distinction between the calcification in the cases of FIF from those of a teratoma or cystic meconium peritonitis. In contrast with the bony calcification of FIF, the calcified features of teratoma have more of a tooth-like appearance, whereas those of cystic meconium peritonitis are amorphous, and deposited in a peripheral capsule. CT also offers a detailed view of the structures that compose the mass, its vascular anatomy, and its relationship with surrounding organs which are important information for surgical management.[18]

In recent years, MRI has also been utilized in the diagnosis of FIF as a problem solving tool, which can clearly identify the soft tissues and organs surrounding the FIF and delineate the tissue characterization, thereby providing valuable imaging data for the formulation of surgical strategies. In case 4, MRI helped us to differentiate the benign FIF from the malignant complex adrenal cortical lesions, which could have demonstrated restricted diffusion and microscopic fat. Compared to CT, MRI is an ideal imaging modality that avoids the need of iodinated contrast. It also eliminates the risk of ionizing radiation and considered safer in pregnancy.

The role of tumor markers is confined to the differentiation between FIF and other causes of intra-abdominal calcified masses, including teratoma, neuroblastoma, adrenal hemorrhage, meconium pseudocyst, and viral infections. The commonly used markers are β-human choriogonadotropin (hCG), maternal and host serum AFP, and urine homovanillic acid.[15]

The symptoms usually relate to the mass effect of the lesion. The blood supply of the FIF is usually derived from the abdominal wall plexus, as it is attached to the abdominal wall. The size and weight of FIF vary depending on the blood supply. However, the absence of an independent circulatory system could account for fetal growth retardation in almost all cases.[19]

The malignant transformation of FIF is not well documented till date, but it can be seen in teratoma. Therefore, differentiating FIF from teratoma plays a crucial role in patient management; since malignant teratoma warrants additional chemotherapy to prevent recurrence. Most of the FIF are well encapsulated, benign, and nonrecurrent thus complete surgical excision is the treatment of choice.

Conclusion

In this case series, we have presented histopathologically proven cases of FIF in various age groups ranging from antenatal, infantile, and adolescent to adult age groups; in various locations including the adrenal

gland, retroperitoneum, and paraspinal region. Conventionally, FIF and teratoma should be considered as the differentials of any calcified intra-abdominal mass seen on antenatal or postnatal imaging. Different imaging modalities, starting from simple radiographs showing calcified structures resembling the long bones and vertebrae, and ultrasounds showing encapsulated mass lesion with peripheral anechoic fluid areas to advanced cross-sectional modalities like CT, and MRI depicting the anatomy and tissue characteristics can aid us in diagnosis and differentiating FIF from the teratoma. The malignant potential of the teratoma necessitates to differentiate it from FIF. Although the presence of true organogenesis at the cellular level is the gold standard in diagnosing FIF; the various radiological modalities together have shown higher confidence level in diagnosing this condition. Therefore, understanding the imaging features in various modalities can help not only in early diagnosis but also in the timely management of this rare condition.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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Interesting case of small bowel gastrointestinal stromal tumor causing ileoileal intussusception

Bolisetti Suprabhat S, Seema Grover, Swapnil C. Patil, Snehagandha S. Satale

Abstract:

Gastrointestinal stromal tumors (GISTs) are the most common submucosal gastrointestinal mesenchymal stromal tumors and are usually seen incidentally on imaging studies. GISTs are most common in the stomach followed by the small intestine. Exophytic and mixed growth patterns are more common in small bowel GIST's. GISTs occur in middle-aged and elderly population. They have an equal preponderance in males and females. Smaller GISTs are homogeneous and well-defined. Larger GISTs are heterogeneous and lobulated with or without areas of ulceration, calcification, hemorrhage, or necrosis. Larger GISTs have an increased risk of malignancy. High-risk GISTs are more common in the ileum. Malignant GISTs commonly metastasize to the liver and peritoneum. Intussusception and subsequent obstruction is a very uncommon presentation of these lesions because of their tendency to grow in an extraluminal fashion. A multimodality imaging approach may be needed for the diagnosis. Computed tomography scan is the imaging modality of choice as it depicts well both the exophytic and endophytic components of the lesion. We present a case of small bowel (ileal) GIST causing intussusception.

Keywords:

Gastrointestinal stromal tumor, intussusception, small bowel

Introduction

Gastrointestinal stromal tumors (GISTs) are the most common gastrointestinal submucosal mesenchymal tumors.[1,2] These are usually detected incidentally on imaging studies done for nonspecific symptoms. They are generally large on detection due to their submucosal location and lack of aggressive features.[3] They are either exophytic, intramural, or mixed/combined/endophytic in location. Larger lesions can cause hemorrhage, ulceration, bowel obstruction, and rupture. GISTs causing intussusception in adults are uncommon.

Case Report

A 28-year-old married nulligravida female was referred to our department for an ultrasound of the abdomen and pelvis for symptoms of abdominal pain and discomfort for a few days. Her transabdominal and transvaginal USG examination revealed an approximately 41 m m × 26 mm × 25 mm (cep halocaudal × transverse × anteroposterior) well-defi ned hypoechoic solid appearing lesion in the lower abdomen. It was surrounded by a bowel loop and revealed foci of calcification [Figure 1a]. The lesion was seen separate from the urinary bladder, uterus, and ovaries. Her dedicated computed tomography (CT) scan study – plain and contrast (using both oral and intravenous contrast) revealed an enhancing well-defined intussuscepting soft-tissue density ileal lesion with no proximal bowel dilatation. The lesion revealed foci of calcification [Figure 1b-i]. There were no fat attenuation areas within. The patient

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underwent surgery which confirmed an intussuscepting ileal lesion [Figure 1j-m] and was resected followed by anastomosis. Histopathology revealed a low-grade gastrointestinal tumor of the ileum [Figure 1n and o]. Her subsequent immunohistochemistry studies were negative for malignancy.

Discussion

GISTs are arising from the interstitial cells of Cajal which are the electrical pacemakers of the gut. GISTs are commonly seen in the stomach followed by the small intestine.[1,2] Burkill *et al*.,[4] however, found small bowel dominance of GISTs in their cohort of 116 patients. They are usually detected incidentally on imaging due to nonspecific symptoms. GISTs are usually large on detection due to their submucosal location and lack of aggressive features. They are exophytic, intraluminal, or mixed-in locations. Exophytic and mixed growth patterns are more common in small bowel GISTs.[5] Larger GISTs can present due to intestinal obstruction, necrosis, hemorrhage, or metastatic disease.

Intussusception and subsequent obstruction is a very uncommon presentation of these lesions because of their tendency to grow in an extraluminal fashion.[5] Very few cases of small bowel intussusceptions from stromal tumors in adults have been described in medical literature. Intussusception is rather infrequent in adults, accounting for 0.1% of all surgical admissions and 1%–5% of mechanical bowel obstructions.[6] Fujimoto *et al*.[6] have reported a case of primary GIST of the third part of duodenum causing duodenojejunal intussusception.

GISTs are more common in middle-aged and elderly population.[1] They have an equal preponderance in males and females. Few articles in the past have, however, cited male preponderance.[7] They are generally solitary, however, multiple lesions can be seen in syndromic conditions. Smaller GISTs (<5 cm) are generally homogenous and well-defined. Larger GISTs are lobulated and heterogeneous with areas of calcification, necrosis, ulceration, or hemorrhage. The risk of malignancy increases with larger size and presence of heterogeneity. Malignant GISTs commonly

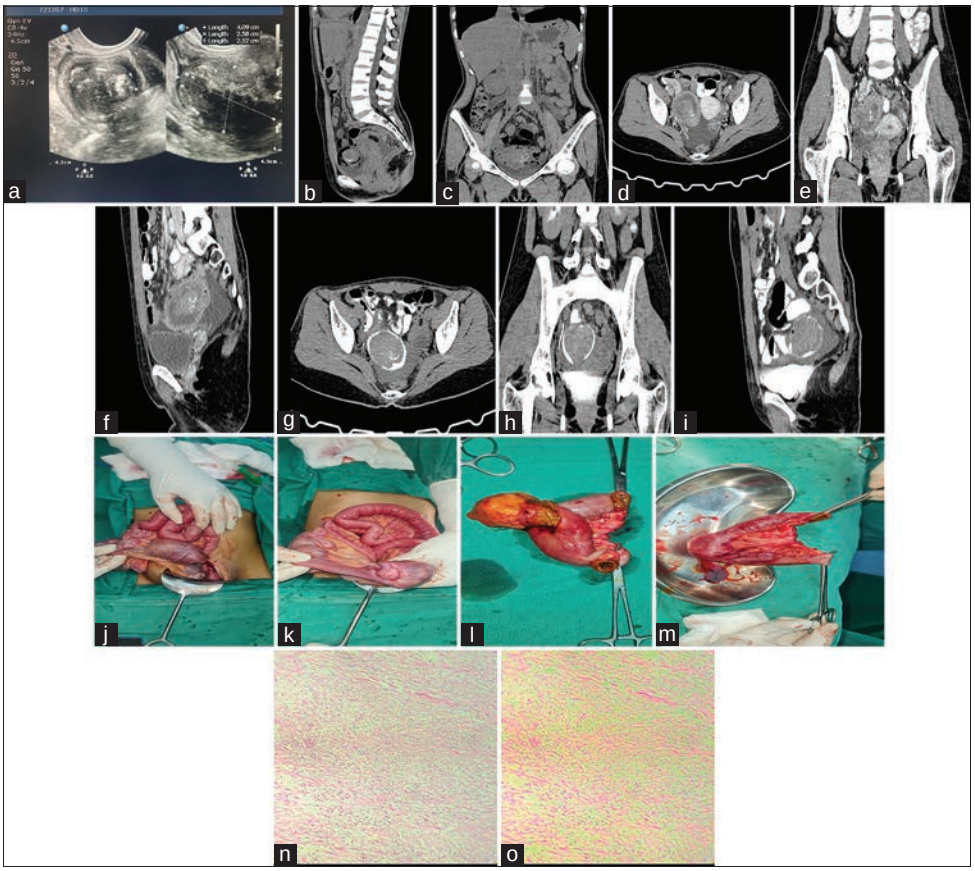

Figure 1: (a) Transvaginal ultrasonography study revealing a heteroechoic appearing solid mass lesion with calcification in the lower abdomen surrounded by bowel loop, (b and c) plain computed tomography (CT) sections revealing a predominantly hypodense solid bowel mass lesion with calcification surrounded by bowel loop, (d-f) contrast-enhanced CT sections revealing heterogeneous enhancement of the lesion seen on axial sections, (g-i) delayed contrast-enhanced sections revealing intraluminal oral contrast filling the bowel loops surrounding the lesion confirming the intussusception, (j-m) intraoperative images showing the intraluminal ileal lesion causing ileoileal intussusception, (n and o) histopathological slides revealing findings of a low-grade gastrointestinal tumor whose immunohistochemistry studies later revealed benignity

metastasize to the liver and peritoneum.[4] High risk GISTs are more common in the ileum.[1]

Necrosis may lead to enteric fistulation.[8] Necrosis was seen as areas of breakdown/nonenhancement observed in 39% of small bowel GISTs.[4] Calcification is seen in 7%–22% of small bowel GISTs.[9] Calcification is more common in larger GISTs and by itself is not significant prognostic factor histopathologically.[10] Calcification within the tumor is occasionally recognized in association with tumor necrosis.[8] GISTs are histologically classified as spindle cell, epitheloid cell, or mixed cell type.[11]

Multimodality imaging approach may be required for the diagnosis. CT scan is, however, the modality of choice as it depicts the exophytic and endophytic components if any, and also identifies areas of hemorrhage, necrosis, calcification, obstruction, and metastatic disease if any. Smaller GISTs are generally well-defined and reveal homogenous moderate enhancement. They tend to show arterial enhancement with an early draining vein. The larger GISTs reveal lobulated contours and heterogeneous enhancement.

Conclusion

GISTs generally cause nonspecific symptoms and are seen incidentally on imaging studies. GIST in a relatively younger age group with calcification, ileal location, nonmalignant, and relatively small in size to cause intussusception is what makes this case interesting.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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Antenatal ultrasound diagnosis of isolated fetal urinary bladder exstrophy at 15 gestation weeks

Sonal Omprakash Gupta, Shafali Jain[1], Kavita Aneja[2], Himani Pandey[3]

Abstract:

Fetal bladder exstrophy (FBE) is a rare and complex congenital malformation of the urogenital system. The features of FBE include the absence of the anterior bladder wall, eversion of the posterior bladder mucosa, exposure to the ureter and urethra, and direct fusion of the posterior bladder wall with the lower abdominal wall skin. FBE is usually detected at the fetal anomaly ultrasound assessment at 18–20 gestation weeks but can be suspected in antenatal ultrasound as early as 14–15 gestation weeks. In this report, we describe a case of isolated fetal urinary bladder exstrophy detected antenatally using two-dimensional ultrasound and color Doppler assessment at 15 gestation weeks and describe the ultrasound findings that supported the diagnosis of FBE.

Keywords:

Antenatal ultrasound, aorto-umbilical angle, fetal Doppler, umbilical artery, urinary bladder exstrophy

Introduction

Fetal bladder exstrophy (FBE) is a rare and complex congenital malformation of the urogenital system that includes the absence of the anterior bladder wall, eversion of the posterior bladder mucosa, exposure to the ureter and urethra and direct fusion of the posterior bladder wall with the lower abdominal wall skin.[1] FBE has a morbidity rate of 1/10,000–1/50,000 and an approximate incidence ratio of 1.5/1–5/1 for males compared to females.[2] The etiology is mostly unknown and genetic or environmental factors such as smoking, alcohol, infections, or teratogenic drugs are probably causal. It is hypothesized that the failure of migration of mesenchymal cells between the abdominal ectoderm and the cloaca during the 4th week of gestation causes a series of abnormalities such as eversion of the bladder on the abdominal surface, inferiorly displaced umbilicus, divergence of pubis, and abnormal external genitalia.[3,4] FBE can occur as an isolated phenomenon and is usually detected at the fetal anomaly ultrasound assessment at 18–20 gestation weeks but can be suspected as early as 14–15 gestation weeks. FBE has a low association with aneuploidy and a low recurrence rate. In this report, we describe a case of isolated fetal urinary bladder exstrophy detected antenatally using fetal ultrasound assessment.

Case Report

A G2 P1 L0 A0 pregnant woman aged 26 years with a history of previous unexplained neonatal death presented for antenatal assessment at 15 gestation weeks. She did not have a history of smoking, alcohol consumption, infections, diabetes, thyroid disorders, hypertension, consanguinity, or consumption of any medications and had a body mass index of 15.2 kg/m^2. The fetal ultrasounds were done by a fetal radiologist with over 23 years of experience in fetal sonography using a GE Voluson E6 machine with C1-6D convex

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probe, 9 L-D high-frequency linear probe, and RIC 5–9 transvaginal four-dimensional (4D) probe. The woman had presented for a nuchal translucency (NT) scan at 15 gestation weeks. Ultrasound assessment showed a crown-rump length of 92 mm and an early anomaly scan was done using 2D ultrasound and color Doppler. A small soft-tissue bulge measuring 0.60 cm ×0.43 cm was identified in the suprapubic anterior abdominal wall [Figure 1] on the sagittal image. The umbilical artery (UA) normally forms an acute angle with the aorta [Figure 2]. We found an increase in the aorto-umbilical angle (K-angle)[5] with the UA touching the ischium on the para-sagittal view [Figures 3 and 4]. The cord X cross-over at the abdominal insertion site was normal, with no evidence of omphalocele, cloacal, or spinal defects. The amniotic fluid volume and the fetal kidneys appeared normal. The urinary bladder remained nondistended on repeated scans over 2 h. An isolated FBE was suspected, and the woman was scheduled for a follow-up anomaly scan at 18 gestation weeks. She presented at 21 gestation weeks for the Targeted Imaging for Fetal Anomalies (TIFFA) scan. The TIFFA scan showed a soft tissue bulge measuring 1.04 cm ×1.17 cm in the lower anterior abdominal wall [Figure 5] with cord insertion just cranial to it (low insertion of cord) [Figure 6]. The anterior abdominal wall showed two bulges, extrophied bladder, and genitalia [Figure 7]. There was an undulating course of intra-abdominal UAs, closely placed UA on axial view with long limbs not forming intra-abdominal Y [Figure 8]. There was a persistent absence of distended bladder between the two UA and the UA was touching ischial tuberosity [Figure 9] on para-sagittal and low axial view. The amniotic volume (amniotic fluid inde×14), kidneys, spine, and bowel loops were normal. The fetal head, heart, stomach, and all four limbs were normal and there was no growth lag at 21 weeks. Four-dimensional (4D) ultrasound studies confirmed low insertion of the cord [Figure 10] and that the urinary bladder is bulging anteriorly from the anterior lower abdominal wall [Figure 11]. The couple was informed that the fetus

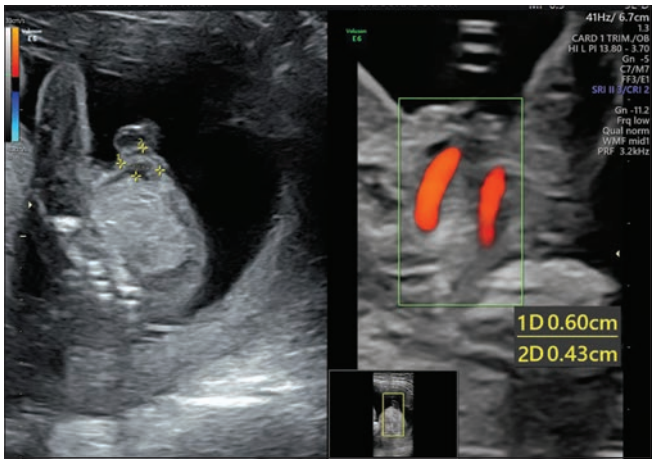

Figure 1: Crown-rump length 92 mm, corresponding with 15 weeks 1 day showing 0.6 mm ×0.4 mm sized cystic out-pouching from the lower anterior abdominal wall and absence of bladder between the two parallel emerging umbilical arteries

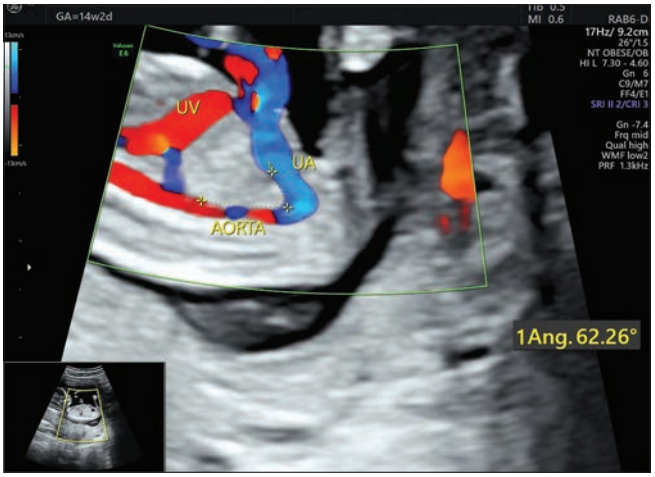

Figure 2: Normal equilateral triangle formed by umbilical artery, umbilical vein, and aorta with acute aorto-umbilical angle. UA: Umbilical artery, UV: Umbilical vein

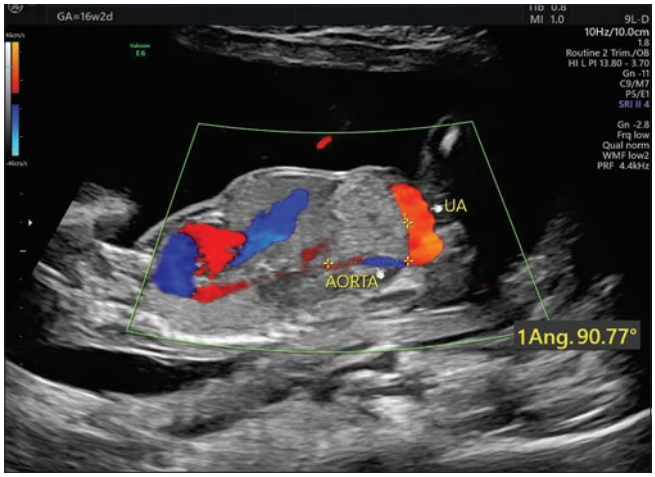

Figure 3: Increased aorto-umbilical angle. UA: Umbilical artery

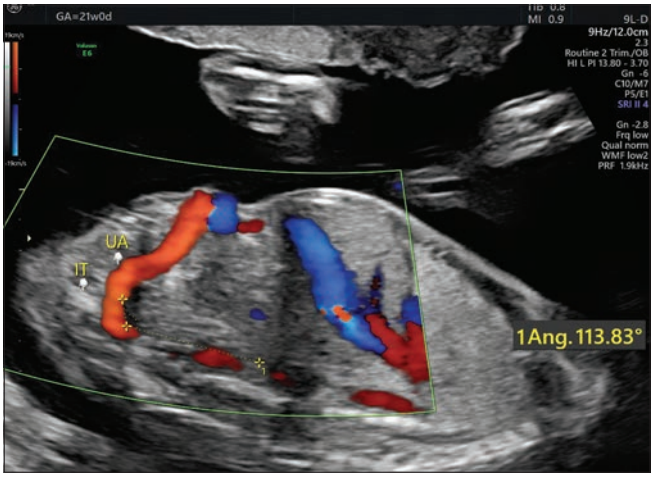

Figure 4: Increased aorto-umbilical angle, umbilical artery touching ischial tuberosity with low cord insertion. UA: Umbilical artery, IT: Ischial tuberosity

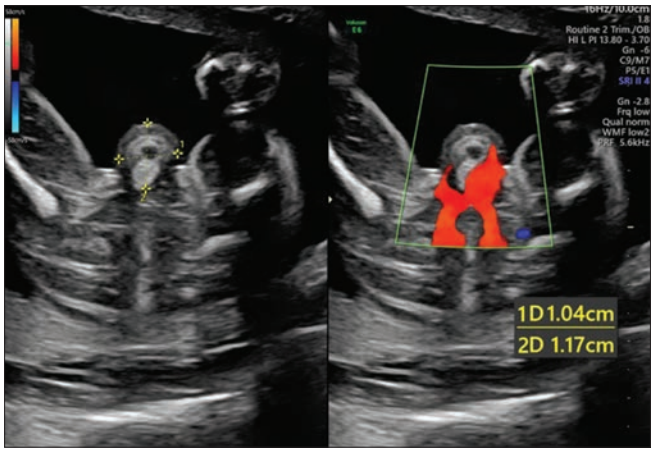

Figure 5: At 21 weeks, a 1.04 cm ×1.17 cm sized echo complex, predominantly cystic lesion at the site of cord insertion over the lower anterior abdominal wall. Umbilical arteries are playing by the side of the lesion

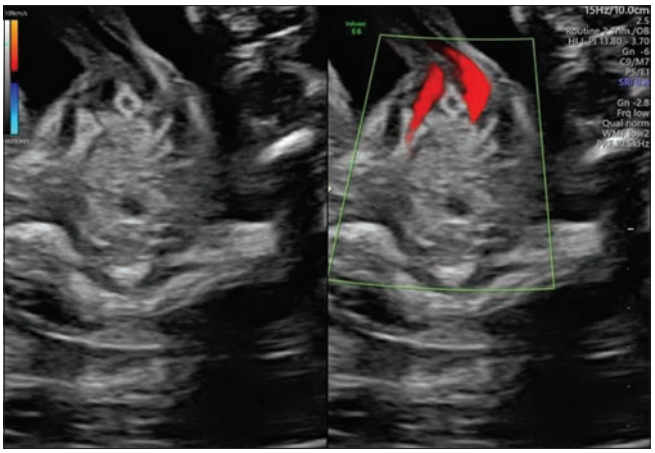

Figure 6: Tiny cystic area at the site of cord insertion between the two umbilical arteries is at the level of the abdominal wall and not completely intra-abdominal

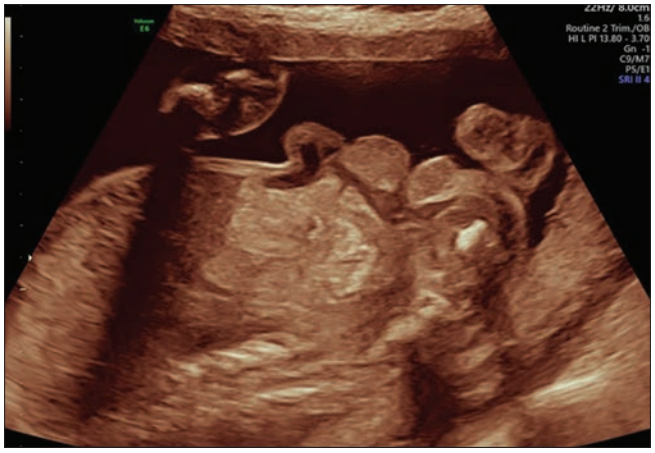

Figure 7: Midsagittal view of the anterior lower abdominal wall shows two ventral bulges: Exstrophy of the bladder and genitalia

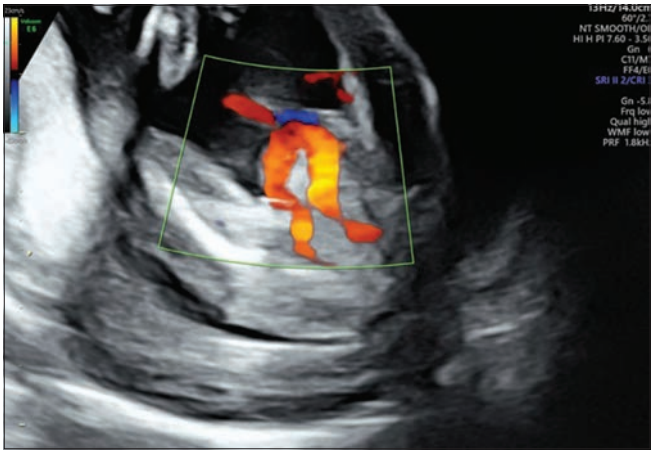

Figure 8: Axial view of pelvis shows long limbs of umbilical arteries, converging outside anterior abdominal wall, and forming "V" instead of "Y." Absence of cystic bladder in between the two vessels

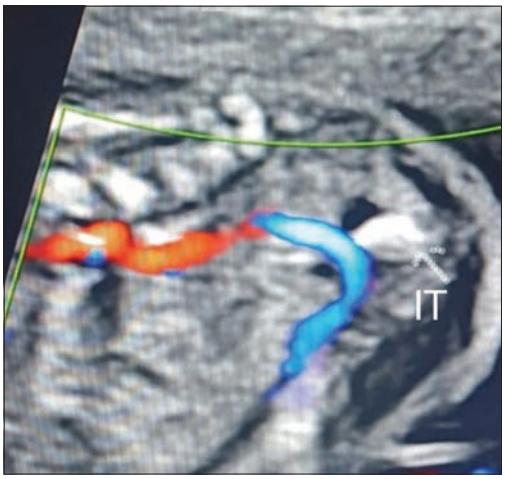

Figure 9: Umbilical artery reaching ischial tuberosity. IT: Ischial tuberosity

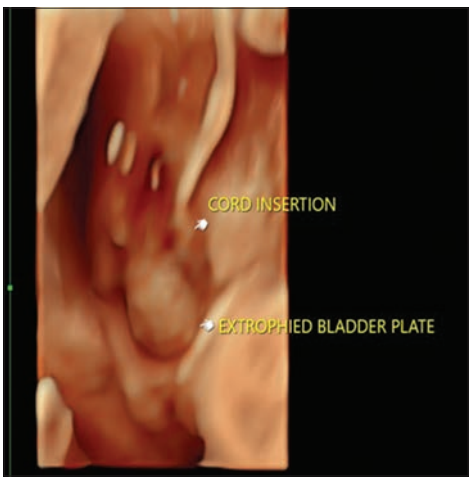

Figure 10: Lateral oblique four-dimensional image to show cord insertion just above the defect

was diagnosed with bladder exstrophy accompanied by abnormal external genitalia and was advised further evaluation by fetal magnetic resonance imaging (MRI) and amniocentesis. The necessity of postnatal surgical interventions, possibly multiple surgical interventions, was explained but the woman opted for termination of the pregnancy. The gross findings were confirmed

by a physical examination of the abortus after the termination of the pregnancy [Figures 12 and 13]. We obtained informed consent from the woman for the anonymous use of the ultrasound images for academic and educational purposes.

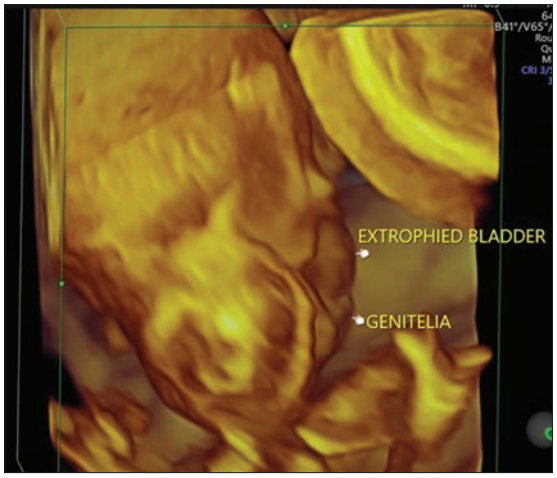

Figure 11: Urinary bladder is bulging anteriorly from the anterior lower abdominal wall (Exstrophy of bladder)

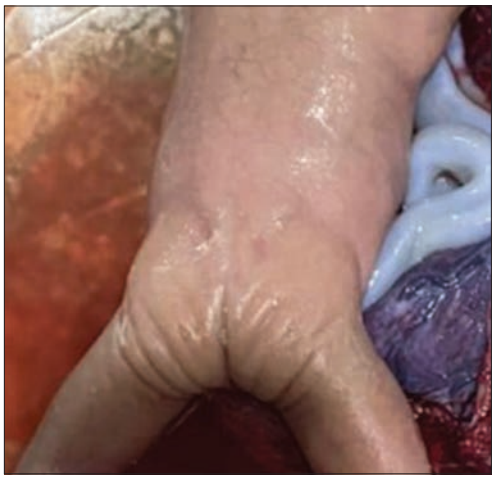

Figure 12: Abortus showing no spinal defect

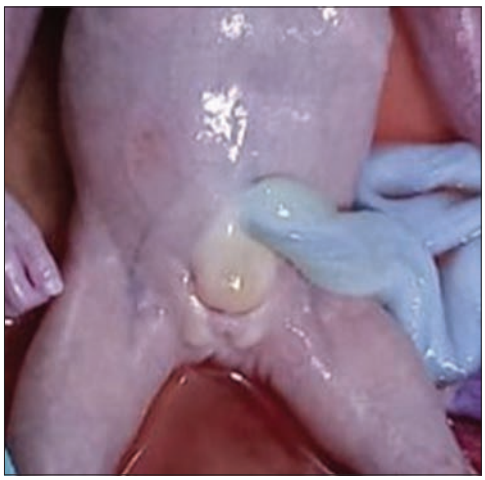

Figure 13: Abortus showing extrophied urinary bladder from anterior lower abdominal wall

Discussion

Early identification of isolated bladder exstrophy at 15 gestation weeks, demonstration of the abnormality using different ultrasound planes, and use of 2D, 4D, and color Doppler studies to confirm the diagnosis are strengths of this case report. In addition, we report on the K-angle, which was described as a differentiating sign in color Doppler in 2017.[5] Exstrophy bladder often has a variable ultrasound appearance as a cystic or soft-tissue ventral mass. The ventral mass may not be visible in the NT scan at 11–14 gestation weeks as the intra-abdominal pressure may not be enough to cause the posterior wall to bulge in early pregnancy.[6] A urinary bladder that is not seen on ultrasound even though the kidneys and amniotic fluid index are normal increases the suspicion of bladder exstrophy. Low insertion of the umbilical cord[6] helps in the early diagnosis of exstrophy bladder between 12 and 18 gestation weeks when there is no visible or obvious bulge from the lower anterior abdominal wall. This case had a visible soft-tissue mass at 15 weeks, nondistended urinary bladder, and other signs on color Doppler including an altered intra-fetal course of UAs, UAs along the sides of bulging mass, and widening of the UA–aorta angle suggestive of bladder exstrophy. Caudal extension of UAs up to ischial tuberosity as seen in this case substantiates the diagnosis in a difficult case of small, isolated bladder exstrophy in the mid-trimester scan.[7] Mapping the course of the UA and the UA–aorta angle using color Doppler studies is useful to differentiate bladder exstrophy from an isolated or simple omphalocele.[5,8] The intra-fetal course of the UA is normal in omphalocele which is a differential diagnosis for bladder exstrophy.

The OEIS complex which includes the combined occurrence of omphalocele, exstrophy bladder with or without cloacal exstrophy, imperforate anus, and spinal anomalies must be ruled out in such cases.[9] The OEIS complex is often associated with renal anomalies and has a sporadic occurrence with unclear etiology although teratogenic effects of medications, defects in blastogenesis, and mutations in genes like HLXB9 have been proposed as potential causal factors. This case did not have spinal or renal abnormalities, imperforate anus, or cloacal exstrophy ruling out the possibility of OEIS complex.

The absence of a bladder in prenatal ultrasonography is an initial clue to diagnosing bladder exstrophy.[10] The fetal bladder can usually be completely visible on ultrasound at 14 weeks of gestation, and the bladder fills and empties every 30–40 min. In FBE, the posterior

wall of the bladder adjoins the abdominal wall, and the ureter on the bladder wall is exposed. Fetal urine drains directly into the amniotic cavity and urine does not fill the bladder. The presence of urine directly jetting into the amniotic cavity during fetal examination is a diagnostic sign of FBE that can help avoid waiting for possible bladder filling or multiple targeted ultrasounds to confirm the diagnosis. FBE must be suspected when the bladder between the bilateral UAs is not filled continuously for 30–60 min without obvious abnormality in both kidneys of the fetus and normal amniotic fluid volume.[11] However, an absent bladder may be misclassified as an emptying bladder as the fetal urinary bladder can have an alternating pattern of filling and emptying, especially if fetal kidneys are normal with normal amniotic fluid volume. Prenatal ultrasound of FBE often shows that the bladder between the two UAs in the lower abdomen is not continuously visible. Color Doppler studies of the UAs must be done when the bladder is not visible.

The umbilical cord insertion-to-genital tubercle length of fetuses with bladder exstrophy is below the fifth percentile of the general population and can be used as an objective ultrasound parameter in prenatal assessment to diagnose suspected cases in early pregnancy.[11] An equilateral triangle of the aorta, UA, and umbilical vein can be seen in color Doppler ultrasound in normal pregnancies.[10] This equilateral triangle can deform when the fetal bladder is everted and can be used as a diagnostic sign for FBE.[10] A low-set umbilicus is another unique characteristic of bladder exstrophy, but identification varies with the experience and skill of the sonographer. An MRI is recommended if the prenatal ultrasonography does not detect a lower abdominal mass in fetuses suspected to have FBE. The MRI can identify ureteral and bladder wall abnormalities without being affected by maternal obesity, amniotic fluid volume, and fetal position.

Conclusion

We recommend careful ultrasound assessment of the fetal bladder, the K angle, and color Doppler studies of the UAs at 14–22 gestation weeks for early identification of FBE. The absence of a urinary bladder or ejection of urine into the amniotic cavity on ultrasound with normal kidneys and amniotic fluid volume and low insertion of the umbilical cord helps in the early diagnosis of exstrophy bladder between 12 and 18 gestation weeks when there is no visible bulge from the lower anterior abdominal wall.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

Acknowledgment

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Nil.

Conflicts of interest

There are no conflicts of interest.

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A launching rocket – Endometrioid carcinoma

Shrishail Adke, Ajith Varrior

Abstract:

Endometrial and cervical cancers are common gynecological malignancies that often present similarly and have considerable overlap, yet they require different treatment strategies. When tumors grow large enough to involve both the endometrium and the cervix, determining their origin can be challenging. We report the case of a 51-year-old female who presented with complaints of vaginal bleeding for the past 5 months and had a hemoglobin level of 4 g/dL. During a per speculum examination, a fragile mass was found occupying the entire vagina. A per vaginal examination could not assess the cervix or the size of the uterus. A magnetic resonance imaging (MRI) revealed a well-defined heterogeneous lesion of intermediate T2 signal in the vagina and fornices, causing distension. The lesion extended into the endometrial cavity without invading the myometrium or cervical stroma. It showed enhancement and diffusion restriction on imaging. A biopsy of the lesion suggested endometrioid carcinoma, and the patient was started on chemotherapy, resulting in near-complete resolution of the lesion. Certain imaging features can help differentiate between cervical and endometrial origins, such as the epicenter of the tumor and the pattern of enhancement on dynamic contrast MRI. Another differential diagnosis to consider is adenoma malignum of the cervix.

Keywords:

Cervical carcinoma, endometrioid carcinoma, launching rocket, gynecological tumors

Introduction

Endometrial and cervical carcinomas represent significant gynecological malignancies in the peri- and postmenopausal population, often presenting with overlapping clinical and imaging characteristics. However, distinguishing between these tumors is crucial, as their management and prognosis differ depending on the tumor type. Here, we present a case of an elderly female presenting with vaginal bleeding and a mass involving the cervix, vagina, and endometrial cavity. Key radiological features for differentiating these tumor types are briefly discussed.

Case Report

A 51-year-old postmenopausal woman presented with complaints of vaginal bleeding for the past 5 months, accompanied by a low hemoglobin level of 4 g/dL. A speculum examination revealed a fragile mass occupying the entire vagina. On vaginal examination, the cervix could not be reached, and the size of the uterus could not be assessed.

A magnetic resonance imaging (MRI) of the pelvis showed a well-defined, heterogeneous lesion with intermediate T2 signal in the vagina and fornices, causing distension. The lesion extended superiorly into the endometrial cavity, resulting in mild distension. Importantly, there was no invasion of the myometrium or the cervical stroma (stage IIa). The portion of the cervix extending into the vagina was surrounded by the tumor, exhibiting a "target" appearance on axial sections. There was no breach of the T2 hypointense rim of the vaginal wall [Figure 1]. The junctional zone is maintained. The lesion was isointense to muscle on the T1 sequence and exhibited

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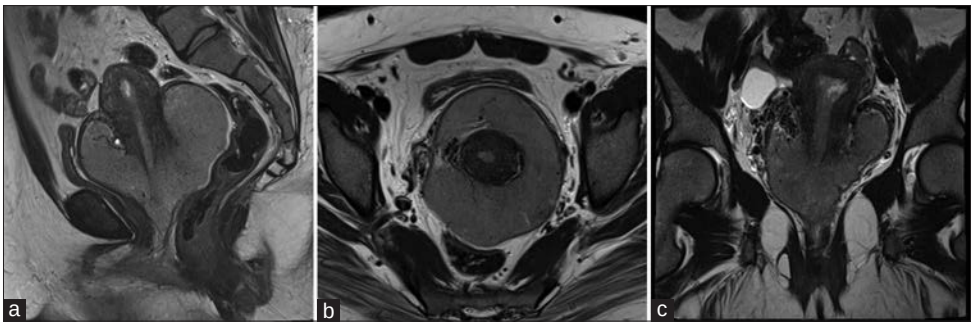

Figure 1: Sagittal (a), axial (b), and coronal (c) sections on T2 sequence of the magnetic resonance imaging pelvis showing a well-defined heterogeneous T2 intermediate lesion in the vagina and the fornices causing its distension. Superiorly, it is extending into the endometrial cavity causing mild distension. There is no myometrial or cervical stromal invasion. There is a targetoid appearance on axial sections due to the cervix surrounded by the tumor in the vagina (b). There was no breach of the T2 hypointense rim of the vaginal wall

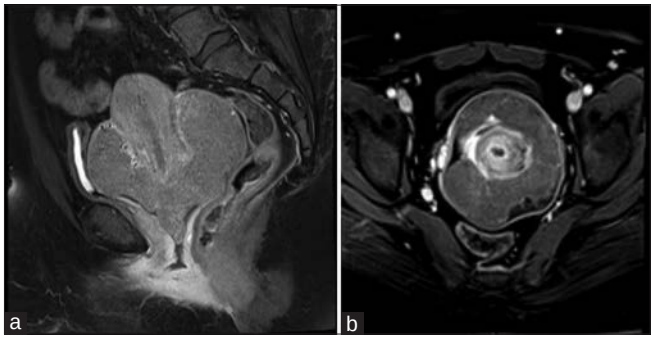

Figure 2: Sagittal (a) and axial (b) sections on postcontrast T1 sequence of the magnetic resonance imaging pelvis showing enhancement of the lesion with few nonenhancing areas in the periphery. The enhancement was less compared to the myometrium and the cervical stroma

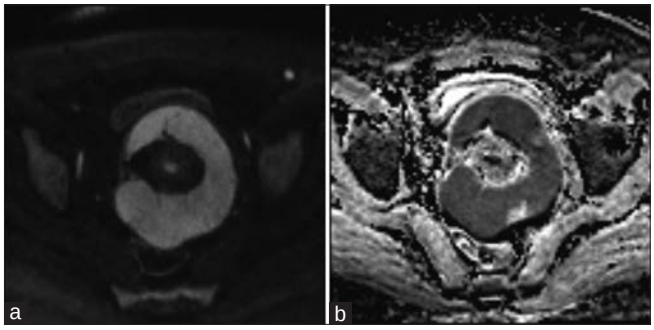

Figure 3: Axial diffusion-weighted sequence of the magnetic resonance imaging pelvis showing true diffusion restriction (a) with corresponding drop on apparent diffusion coefficient (b)

postcontrast enhancement that was less than that of the myometrium and cervical stroma [Figure 2]. In addition, the lesion showed diffusion restriction with a corresponding drop on the apparent diffusion coefficient map [Figure 3].

The bulk of the tumor in the vagina, along with the endometrial component, resembled a launching rocket, where the body of the rocket represented the tumor within the endometrial cavity, while the fumes represented the lesion within the vaginal cavity and fornices [Figure 4].

Histopathological findings from the biopsy of the lesion were suggestive of endometrioid type of endometrial carcinoma, which showed strong positivity for estrogen receptor (ER) and vimentin (Vim) and weak positivity for p16 and carcinoembryonic antigen (CEA). The patient was started on dexamethasone and a neoadjuvant chemotherapy comprising of paclitaxel and carboplatin regimen. A postchemotherapy MRI revealed near-complete resolution of the lesion, with a nabothian cyst in the anterior cervical lip [Figure 5]. Surgery was deemed unnecessary due to postchemotherapy resolution.

Discussion

Endometrial and cervical cancers are prevalent gynecological malignancies with overlapping clinical presentations, yet they necessitate distinct treatment approaches.[1,2] Differentiating the primary site of origin becomes particularly challenging when tumors are large and involve both the endometrium and cervix. The endometrioid variant of endometrial carcinoma is often linked to increased estrogen exposure, commonly associated with hormone replacement therapy, obesity, tamoxifen use, early menarche, late menopause, nulliparity, endometrioma, or ovarian granulosa cell tumors. There is an increased risk in patients with hereditary nonpolyposis colorectal cancer. In contrast, cervical carcinoma is strongly associated with human papillomavirus (HPV) infection (HPV 16, 18).

Treatment for endometrial carcinoma typically involves total hysterectomy with bilateral salpingo-oophorectomy and lymph node dissection. Early-stage cervical cancer is generally managed with radical hysterectomy, lymphadenectomy, and radiotherapy, while advanced cervical cancer is primarily treated with chemoradiation.[3]

MRI is the gold standard for noninvasive imaging in diagnosing pelvic malignancies, with specific features aiding in distinguishing between cervical and endometrial origins. It depicts the extent of myometrial invasion and cervical involvement.[4]

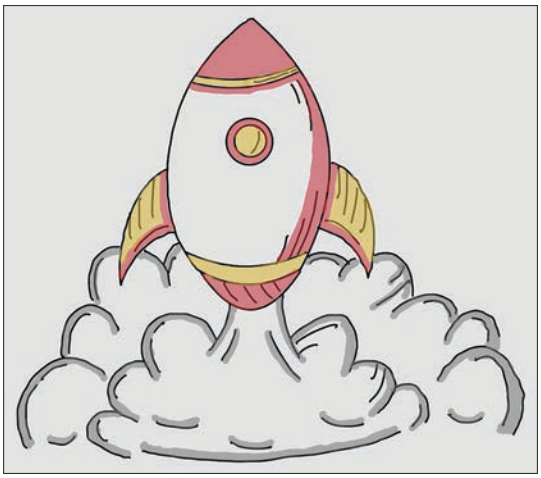

Figure 4: Schematic representation of a launching rocket where the body of the rocket signifies the lesion within the endometrial cavity causing its distension. The fumes from the rocket represent the lesion within the vaginal cavity and the fornices

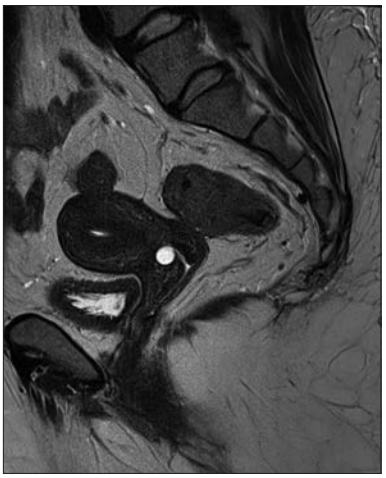

Figure 5: Sagittal T2 sequence of the magnetic resonance imaging pelvis postchemotherapy showing near-complete resolution of the lesion with a T2 hyperintense cyst in the anterior lip of the cervix

MRI is recommended in type I endometrial cancer to identify patients with stage Ia who would not benefit from lymphadenectomy. Furthermore, in stage I, if the tumor is confined to the endometrium, a fertility-sparing treatment can be offered to the patient. In type II endometrial cancer, MRI is used to detect extrauterine spread.

In cases of cervical carcinomas, MRI is used to accurately determine the tumor size, parametrial and pelvic side wall invasion, and lymph nodal involvement. Fertility-preserving treatment for cervical carcinoma includes radical trachelectomy and is employed when the tumor size is <2 cm (for vaginal route-VRT) or 4 cm (for abdominal route-ART) and the tumor distance from the internal os is >1 cm (VRT) or 0.5 cm (ART).[4]

The tumor's epicenter offers the highest sensitivity and specificity for differentiation.[3] Morphologically, endometrial carcinoma often appears longitudinal, while cervical carcinoma typically has a round or oval shape.[2] Deep myometrial invasion and tumor: uterus volume ratio >0.13 correlated with high-grade endometrial carcinoma.[4] On early dynamic contrast-enhanced MRI (DCE-MRI), cervical carcinoma is characterized by hypervascularity and full-depth cervical stromal invasion. In contrast, endometrial carcinoma commonly shows cavity distension, myometrial invasion, and peripheral rim enhancement. Retained secretions are another feature occasionally observed in cervical carcinoma. A 7-point scoring system has been proposed for differentiation: a score >4 indicates cervical origin, while a score below 4 suggests an endometrial origin.[3] Furthermore, quantitative perfusion parameters derived from DCE-MRI are significantly lower in endometrial cancers compared to cervical cancers.[2]

There are certain limitations of MRI when it comes to assessing myometrial invasion in endometrial carcinoma such as isointense tumors on T2 sequence, poor endometrial–myometrial interface due to fibroids or adenomyosis, and thin myometrium at the cornua or in the atrophic uterus. Diffusion-weighted images and DCE can help in detecting and delineating the tumor.[4]

Adenoma malignum of the cervix is a rare and aggressive form of cervical cancer with a poor prognosis due to its propensity for early peritoneal spread and distant metastases.[5] On MRI, it typically presents as a multicystic tumor (hyperintense on T2-weighted images) with a solid component invading the cervical stroma.

Immunohistochemistry plays a key role in the histopathological diagnosis of these tumors. Endometrial carcinoma is usually positive for ERs, progesterone receptors, and Vim but negative for CEA and p16. In contrast, cervical carcinoma often shows positive staining for p16 and CEA, with ER being either negative or focally positive, although some overlap can occur. In addition, HIK-1083 positivity is a distinguishing feature of adenoma malignum.

Conclusion

Although there might be an overlap in the imaging findings of an endometrial and cervical carcinoma, careful examination and certain key features help in distinguishing them. The epicenter of the tumor, enhancement characteristics on a dynamic contrast MRI, and involvement of adjacent structures aid in identifying the origin of the tumor.

Declaration of patient consent

The authors certify that they have obtained all appropriate

patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest
There are no conflicts of interest.

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Cystic echinococcosis/hydatid disease of thyroid gland

Zainab, Indraneel Borekar, Yogendra Sachdev

Abstract:

Cystic echinococcosis/hydatid disease of the thyroid gland is a rare occurrence with prevalence ranging between 0 and 3.4%. We present to you a case of primary hydatid disease of the thyroid gland in a 65-year-old male patient who was clinically diagnosed with a thyroid nodule. The patient underwent fine-needle aspiration cytology-thick gel-like material was aspirated from the cystic nodule and many protoscolices with hooklets, detached refractile hooklets, and fragments of laminated membranes were seen microscopically which were suggestive of hydatid cyst.

Keywords:

Echinococcosis, hooklets, hydatid, protoscolices, thyroid

Introduction

Hydatid cyst is a parasitic disease caused by echinococcus granulosis. It has a high incidence rate in Mediterranean countries, the Middle East, South America, New Zealand, Australia, and Southeast Asia. Hydatid cyst develops most often in the liver (65%) and lungs (25%) in human beings.[1] Hydatid cyst involvement of the thyroid is rather rare. There are approximately 160 thyroid hydatid cases reported in the literature.[2] In patients with diagnosed hydatid cysts in the liver, systemic evaluation is necessary to rule out the involvement of other organs. Among patients presenting with growths located in the neck, primary hydatid cysts of the thyroid gland must be considered in endemic regions. Hydatid cysts in the thyroid may grow rapidly and result in compressive and obstructive problems. Furthermore, rupture of hydatid cysts can trigger complications such as anaphylaxis, shock, and death.[3]

Case Report

A 65-year-old male patient presented with painless swelling in the front of the neck for the past 6 months with no complaints of dysphagia/dyspnoea/hoarsness of voice. The patient had no hyper/hypothyroidism-related symptoms. On examination, it was a smooth well-defined swelling showing movement on deglutition. Blood investigations showed normal T3, T4, and TSH levels [Figure 1].

On ultrasound examination

A well-defined multiloculated anechoic cystic lesion with internal septations was noted involving the left lobe of the thyroid gland with extension into the isthmus. The lesion did not take internal/peripheral vascularity on color Doppler and was causing a mass effect on the trachea in the form of the subtle tracheal shift to the right [Figures 2-4].

On contrast-enhanced computed tomography

A well-defined multiloculated cystic lesion with few internal septations was noted involving the left lobe and isthmus of the thyroid gland on postcontrast study. The lesion did not show peripheral wall or septal enhancement [Figure 5].

On magnetic resonance imaging

Evidence of well-defined multivesicular T1 iso-to-hypointense, T2 hyperintense

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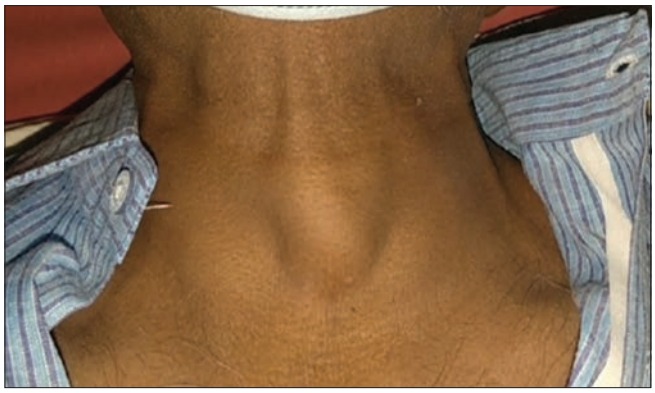

Figure 1: Swelling in the front of the neck which showed movement on deglutition.

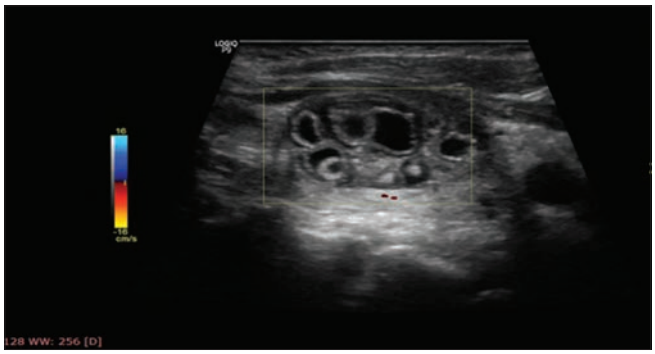

Figure 3: The lesion is not taking internal or peripheral vascularity on color Doppler ultrasound.

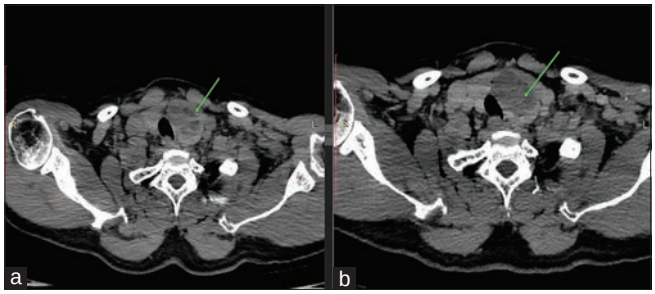

Figure 5: (a) Axial plain CT image of neck shows a well-defined multiloculated cystic lesion with few internal septations involving left lobe and isthmus of thyroid gland causing mass effect on trachea in the form of tracheal shift to the right. (b) Postcontrast axial image shows no peripheral wall or internal septal enhancement

cystic lesion noted involving the left lobe of the thyroid gland and isthmus. Multiple varying-sized intralesional daughter cysts were noted with perilesional T2 hypointense rim, possibly fibroblastic pericyst. The lesion is causing a mass effect on the trachea in the form of tracheal shift to the right [Figure 6, 7].

Discussion

Hydatid disease is a parasitic infection with worldwide distribution, especially in sheep- and cattle-rearing regions of Australia, South America, the Middle East, South Africa, Eastern Europe, and the Mediterranean region. Hydatid cysts of *Echinococcus granulosus* can be developed in

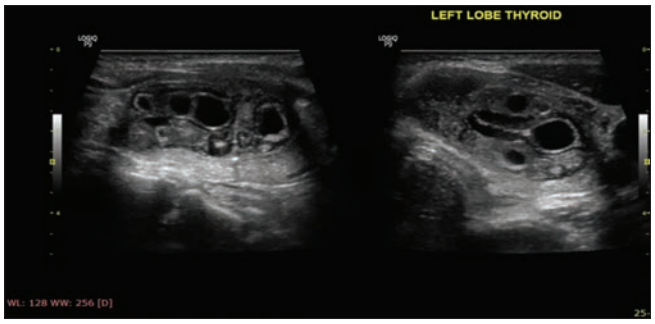

Figure 2: Well-defined multiloculated anechoic cystic lesion with internal septations in the left lobe of thyroid gland.

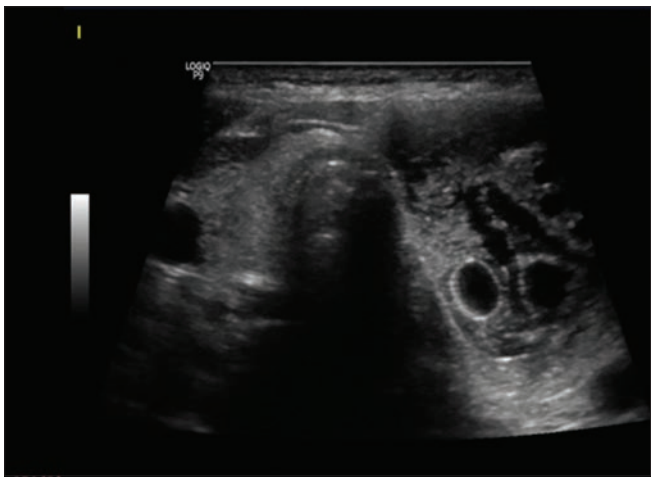

Figure 4: The lesion is causing mass effect on trachea in the form of subtle tracheal shift to the right.

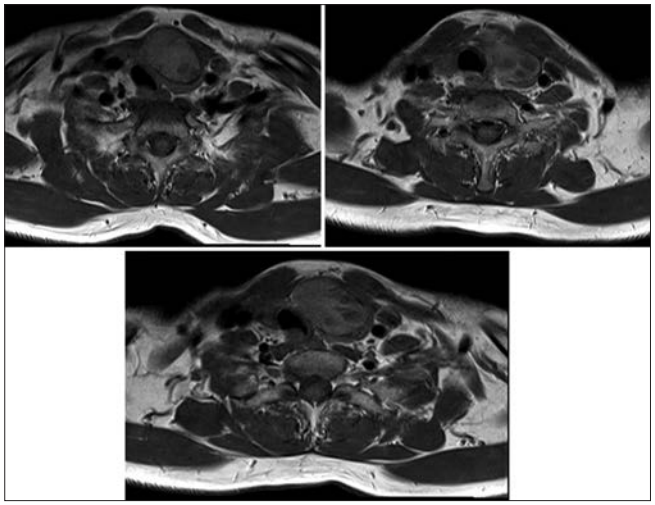

Figure 6: Axial T1W1 of neck shows well-defined multivesicular iso-to-hypointense lesion involving left lobe and isthmus of thyroid gland causing mass effect on trachea in the form of tracheal shift to the right.

any part of the body. Multiorgan involvement has been reported in 20%–30% of hydatid disease cases.

The parasitic embryo can enter the systemic circulation and lodge in the thyroid gland after either bypassing

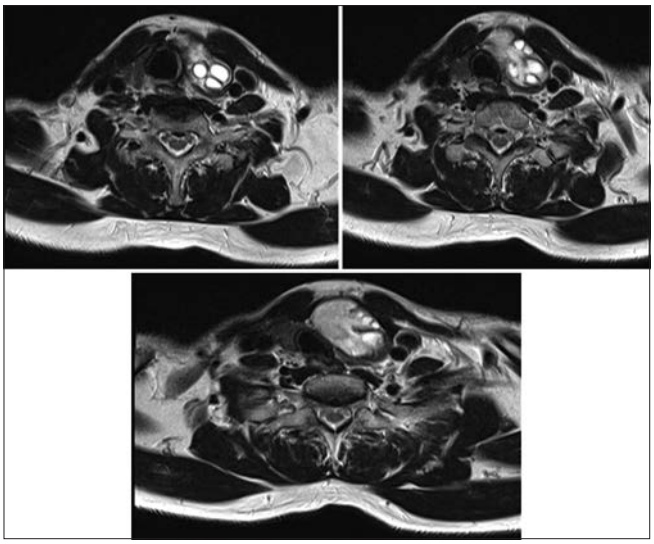

Figure 7: Axial T2WI of neck showing hyperintense lesion involving the left lobe and isthmus with multiple varying sized hyperintense intralesional daughter cysts and a perilesional hypointense rim possibly fibroblastic pericyst. Right lobe of thyroid gland and overlying strap muscles appear normal.

(primary type) or passing through (secondary type) the hepatic microcirculation. A high blood flow rate in the thyroid gland may be responsible. However, the small diameter of the thyroid arteries explains the rarity of the disease. Hydatid disease caused by *E. granulosus* is often manifested by a slow-growing cyst mass. The cyst might remain clinically silent for a long time period. It may suddenly increase in size after years of dormancy. When it increases in size, it may adhere to the surrounding structures, such as the trachea, esophagus, carotid sheath, recurrent laryngeal nerve, and strap muscles, in a similar manner with thyroid carcinoma. Consequently, the patient may present with pressure symptoms and signs such as dyspnea, paralysis of the vocal cord, hoarseness, or dysphagia. Complications include anaphylaxis by spontaneous or surgical rupture of the cyst, pyogenic abscess in secondary infected cysts, compression symptoms, and internal organ damage.

Diagnosis of hydatid disease has been greatly facilitated with ultrasonography, computed tomography (CT), and magnetic resonance imaging (MRI). Ultrasonography is highly efficient in detecting germinal vesicles in cystic lesions, which is important for a preoperative diagnosis of hydatidosis. Although cystic echinococcosis typically consists of a single unilocular cyst, 20%–30% of cases present with multiple cysts in the same or multiple organs. The multiple hypoechogenic images noted within the cystic lesions were thought to represent germinal vesicles of a hydatid cyst. CT scan and MRI are complementary studies. They provide a precise assessment of the extension into the soft tissues and the calcifications of the peripheral rim of the cyst. The signal from the cysts is inhomogeneous of low intensity on T1-weighted and high intensity on T2-weighted images. Hydatid disease may mimic benign or malignant tumors, cysts, abscesses, and other lesions.

Management of thyroid hydatid cyst is surgical. Complete excision should be the procedure of choice. The commonly held opinion is that echinococcal cysts should be radically removed whenever possible, the authors recommend subtotal thyroidectomy, especially when the cyst is small and confined to the thyroid gland. Chemotherapy is necessary to avoid recurrence.[4]

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Conflicts of interest
There are no conflicts of interest.

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A case of wandering spleen complicated by pseudocyst: Radiological features and management

Shrishail Adke, Chandrasekhar I. Khot, Ajith Ramakumar Varrior, Padma Vikram Badhe

Abstract:

Wandering spleen is a rare condition characterized by the absence of the spleen in its normal anatomical position due to congenital or acquired abnormalities of its suspensory ligaments. This anomaly predisposes the spleen to torsion and infarction due to the formation of a long vascular pedicle. It can also be rarely complicated by pseudocyst formation. The condition is most commonly observed in multiparous women aged 20–40 years and is often asymptomatic, though it may present with acute or intermittent abdominal pain. Diagnosis relies on radiological imaging, including ultrasound, CT, and nuclear scans, which identify the ectopic spleen and assess complications. Management primarily involves surgical intervention, with splenopexy preferred in viable spleens and splenectomy reserved for cases with infarction or massive splenomegaly.

Keywords:

Ectopic spleen, pseudocyst, splenectomy, wandering spleen

Introduction

Wandering spleen is a condition, where spleen is not seen in its normal anatomical location of left upper quadrant and is seen in ectopic location in lower abdomen or pelvis. It mainly results from the abnormality of suspensory ligaments of spleen. Pseudocyst in a wandering spleen is a very rare entity. Here we present a rare case of Pseudocyst in a wandering spleen.

Case Report

A 16-year-old girl presented with a history of intermittent abdominal pain for the past 1 year. The pain has increased in intensity over the past week.

On examination, there was fullness noted in the left lower abdomen.

Imaging findings

An ultrasound [Figure 1] at our center revealed the spleen to be absent at the splenic fossa, with a structure suspected to be an ectopic spleen located inferior to the left kidney and extending into the pelvis. A large hypoechoic pseudocyst with echogenic material was noted within this ectopic spleen. Transvaginal ultrasound which would have shown pelvic extension and the relation of this cyst with ovaries and other pelvic organs better could not be done as the patient was an unmarried female.

A contrast-enhanced computed tomography (CECT) abdomen [Figure 2] was carried out for further evaluation. The CT image shows the stomach occupying the splenic fossa and the superior pole of the spleen located inferior to the left kidney. The pancreatic body and tail were displaced inferiorly along with the splenic vein, superior mesenteric vein, and their confluence. An

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ectopic spleen was noted in the pelvic region with a pseudocyst inside.

Magnetic resonance imaging images [Figure 3] of the abdomen revealed the stomach in the splenic fossa. Pelvic images showed splenic parenchyma in the pelvis, with a T2 hyperintense cyst displacing both ovaries and the uterus posteriorly. The ovaries were seen separately from the lesion, ruling out an ovarian cyst with torsion.

Based on these findings, she was diagnosed to have wandering spleen with pseudocyst.

The patient underwent total elective splenectomy, 2 weeks after administering the prophylactic pneumococcal, meningococcal, and *Haemophilus Influenza* B vaccine. Intraoperative findings confirmed the imaging results, and the postoperative course was uneventful.

Discussion

Wandering spleen (ectopic, floating, or proptotic spleen) is a condition where the spleen is not seen in its normal anatomical location in the left upper quadrant and is instead seen in an ectopic location in the lower abdomen or pelvis.[1] It mainly results from an abnormality of the suspensory ligaments of the spleen, which results in its abnormal mobility. The spleen starts to develop from mesenchymal cells in the dorsal mesogastrium in the 5th week of gestation.[2] The suspensory ligaments of the spleen, mainly the gastrosplenic and lienorenal ligaments, develop from the dorsal mesogastrium and are responsible for fixing the spleen in its normal position. There may be congenital absence or maldevelopment of these ligaments or acquired laxity of these ligaments caused by pregnancy or conditions causing splenomegaly.[3] A long vascular pedicle may form containing splenic vessels secondary to the abnormality of these ligaments, predisposing the wandering spleen to torsion and consequent splenic infarction.[4]

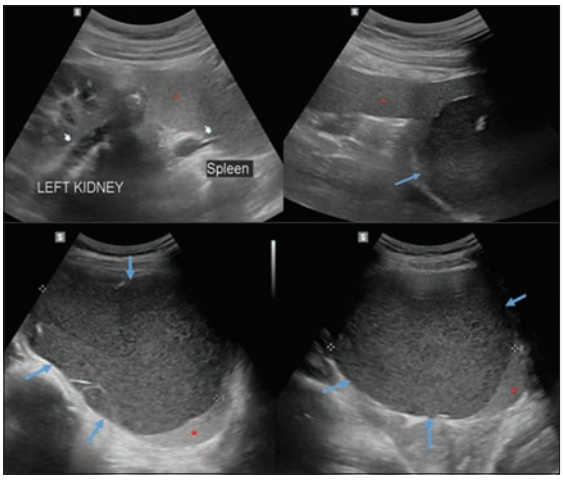

Figure 1: Ultrasound image shows splenic superior pole (red star) inferior to left kidney. Large hypoechoic pseudocyst with echogenic material within it (blue arrow)

It is a rare condition with a reported incidence of <0.5%. It has a female preponderance, with multiparous women being most commonly affected.[4] The most common age group for presentation is 20–40 years. It is generally asymptomatic and has variable presentations. It may present as a painful abdominal mass due to acute torsion or intermittent abdominal pain due to episodes of torsion and spontaneous detorsion, making clinical diagnosis elusive.

Radiological investigations play a crucial role.[5] A plain radiograph may show the absence of a splenic shadow in the left upper quadrant and an ectopic soft-tissue opacity in the abdomen. Ultrasound examination shows an absent spleen in the splenic fossa with the finding of a comma-shaped characteristic spleen in an ectopic location. The mobility of the wandering spleen can be demonstrated in the right decubitus position. CECT plays a crucial diagnostic role in confirming the ectopic spleen. The extent of ischemia can be determined in the setting of splenic infarction or torsion. The Whirl sign can be seen due to the twisting of the vascular pedicle. A Tc-99 m sulfur colloid liver–spleen scan is useful in confirming an abnormal mass in the abdomen as the spleen.[6]

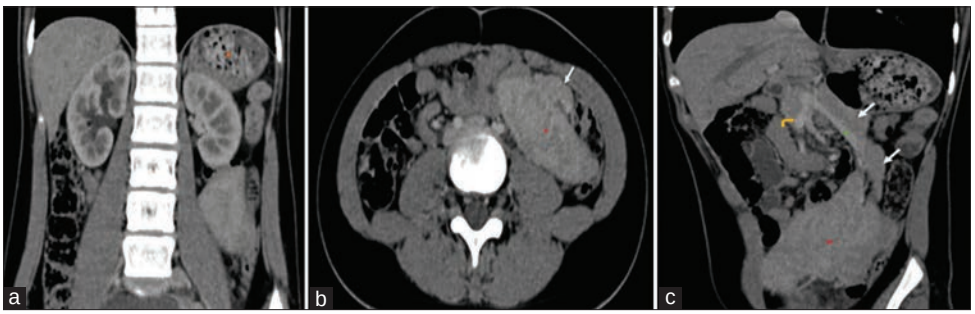

Figure 2: CT image (a) shows the stomach (orange star) occupying the splenic fossa and splenic superior pole inferior to the left kidney. Pancreatic body and tail (white arrows) displaced inferiorly along with splenic vein (green star) and merging with superior mesenteric vein (yellow curved vein) (b) to form superior mesenteric–splenic vein confluence. Ectopic spleen (red star) (c) in pelvic region

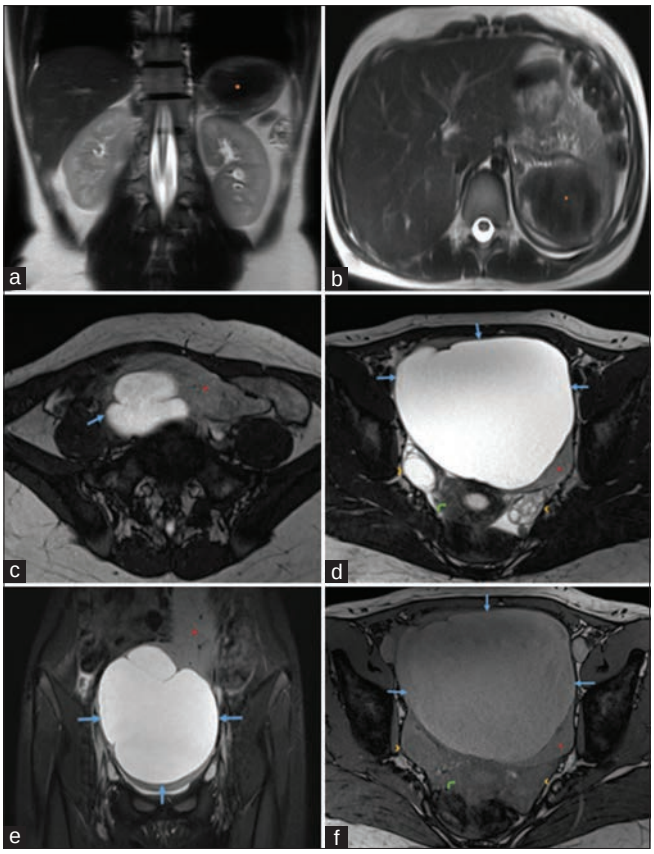

Figure 3: Magnetic resonance imaging T2W images of the abdomen in the coronal and axial plane (a and b) reveal splenic fossa occupied by the stomach (orange dot). Axial, coronal T2W images (c-e) and axial T1W image (f) of the pelvis reveal splenic parenchyma in the pelvis (red star), hyperintense cyst (blue arrows) displacing both ovaries (yellow arrowheads) and uterus (green curved arrow) posteriorly. Both ovaries (yellow arrowheads) are seen separately from the lesion

Cysts of the spleen are classified as parasitic and nonparasitic cysts. The most common cause of parasitic cysts is a hydatid cyst. Nonparasitic cysts are classified into true cysts and pseudocysts based on the presence or absence of an epithelial lining. Pseudocysts of the spleen are rare, with a reported incidence of <1% in splenectomy specimens.[7] They are usually posttraumatic, inflammatory, or degenerative. Serum CA 19-9 levels are high in true cysts, while they are rarely raised in pseudocysts due to the absence of an epithelial lining.[7]

Management is mainly surgical, with splenopexy being the treatment of choice. However, splenectomy is performed in cases of splenic infarction and in massive splenomegaly, where splenopexy is not feasible.[1] Alternate treatment options are percutaneous aspiration and drainage and laparoscopic fenestration and partial splenectomy. However, they carry the risk of infection, recurrence, and hemorrhage in complicated and large lesions.[2]

In our case, complete splenectomy was done instead of partial splenectomy with complete cystectomy as there was a future risk of cyst recurrence, torsion, infarction, and intraoperative hemorrhage.

Teaching points

1. In the absence of a history of splenectomy, wandering spleen or asplenia should be suspected if the spleen is not visualized in splenic fossa on imaging
2. In female patients having cystic lesions with acute abdomen, an important differential is an ovarian cyst with torsion. Other differentials listed below should also be kept in mind.

Final diagnosis

Wandering spleen complicated by pseudocyst.

Differential diagnosis list

- Ovarian cystic lesions
- Pancreatic cystic lesions
- Mesenteric cyst
- Intestinal duplication cyst.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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Ollier's disease: A rare insight into imaging features

Aisha Lakhani, Prashant Onkar, Kajal Ramendranath Mitra, Azeem Merchant

Abstract:

Ollier's disease, also known as multiple enchondromatosis, is a rare congenital disorder with no identifiable cause. A nonossifying chondrocyte mass or a chondrocyte developing hamartomatously in the metaphysis are the clinical manifestations of this condition. The current study describes the case of a 21-year-old male patient who presented with pain, functional and aesthetic concerns in his left hand as a result of soft tissue swelling and was diagnosed with Ollier disease based on clinical observations and imaging findings. Ollier's disease is uncommon in general, and as a result, it is less recognized among orthopedic surgeons. Physicians should counsel patients to have routine follow-up testing because of the increased risk of malignant transformation. To understand more about this disease, the present case study details a patient's case preceding reviewing relevant literature to help in clinical diagnosis and management.

Keywords:

Benign bone tumor, multiple enchondroma, Ollier's disease

Background

Ollier described an uncommon disease characterized by several unilateral benign enchondromas principally located in the metaphyseal regions, in 1889.[1] The disease pathogenesis is still unknown; however, somatic mosaic mutations in IDH1 and IDH2 have been linked to Ollier's disease and Maffucci syndrome.[2] Clinical and radiological examinations identify Ollier disease rather than genetic testing. The lesions often develop in the medullary cavity and expand in the cortex, producing a distinctive endogenous bone mass. It develops as localized pain, swelling of the bones, palpable bone mass, and deformity.[3] The incidence of chondrosarcoma is significant, with transformation rates ranging from 5% to 50%.[4] The current case report discusses an Ollier disease and summarize to help understand this rare condition.

Case Report

A 21-year-old man was sent to the radiology department for hand radiographs because of soft-tissue swelling in his left hand, which caused him discomfort, functional problems, and cosmetic issues. He experienced difficulties operating with his left hand on the computer. On examination, we found swellings and deformities on his dorsal hand and the proximal phalanx of the middle finger. There was no history of injury, systemic symptoms, or operations. The skin and tendons were unaffected. Laboratory studies were within normal findings.

Multiple well-defined lytic expansile lesions (enchondroma) affecting the metacarpi and phalanges of the middle and ring fingers of the left hand with narrow transition zones were seen on radiographic images of the hand. The lesion is causing endosteal scalloping with cortical thinning of the proximal phalange of the middle

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finger [Figure 1a]. Although the underlying skeletal deformities altered soft-tissue contour, no soft-tissue mass or calcification was seen. On magnetic resonance imaging (MRI), lesions appeared hypointense on T1 and hyperintense on T2 with peripheral contrast enhancement and no adjacent soft-tissue involvement or perilesional edema. The imaging findings show no signs of aggressive activity [Figure 1b-d]. The case was diagnosed as a benign bone tumor based on the clinical and imaging characteristics.

We obtained a biopsy from nodules on the left hand under brachial plexus anesthesia to rule out malignancy. Histologic findings revealed benign cartilaginous patterns of multiple enchondromatosis [Figure 1e]. No hemangiomas were evident, suggesting that Ollier's disease was the correct diagnosis.

Discussion

Several enchondromatosis, known as Ollier's disease, is an uncommon, sporadic, nonheritable skeletal illness marked by multiple enchondromas. Ollier initially characterized it in 1899.[2] Long, flat bones such as the pelvis, tibia, and femur, as well as small bones such as the hands and feet, are the most common sites for multiple subperiosteal cartilaginous deposits (enchondromas). Lesions have been seen to manifest unilaterally or bilaterally, with a significant predominance of unilateral lesions.[5] Clinically, patients manifest multiple tubular and flat bone enchondromas, predominantly unilateral, within the first decade of life. Individuals may have skeletal abnormalities such as bending, shortening, pathological fractures, and asymmetric deformity. Frequently, the physis is disturbed, resulting in irreversible growth abnormalities and deformities throughout the patient's life.[6] Ollier's disease significantly increases the chance of malignant transformation, with enchondromas most often converting into chondrosarcomas between 40 and 50.[7,8] The disease should be evaluated using clinical description, radiographic imaging, and pathologic findings.

Ollier's disease is frequently diagnosed using imaging modalities such as radiograph, computed tomography (CT), MRI, and nuclear medicine. Due to its low cost and great sensitivity, radiograph is often the first diagnostic modality, with CT as a follow-up. Radiographs of the diaphysis or metaphysis reveal well-defined expansile lesions with central "rings and arcs" or scattered calcification indicative of degeneration and poor vascularity across the affected bones without cortical penetration. Ollier's disease is characterized by radiolucent channels in the metaphysis of long tubular bones.[9,10] CT is advantageous for determining the size and position of soft tissues in regions where radiography is inaccessible, such as the pelvis– analysis of mineralization patterns aided in predicting chondrosarcoma malformation. Chondrosarcoma is

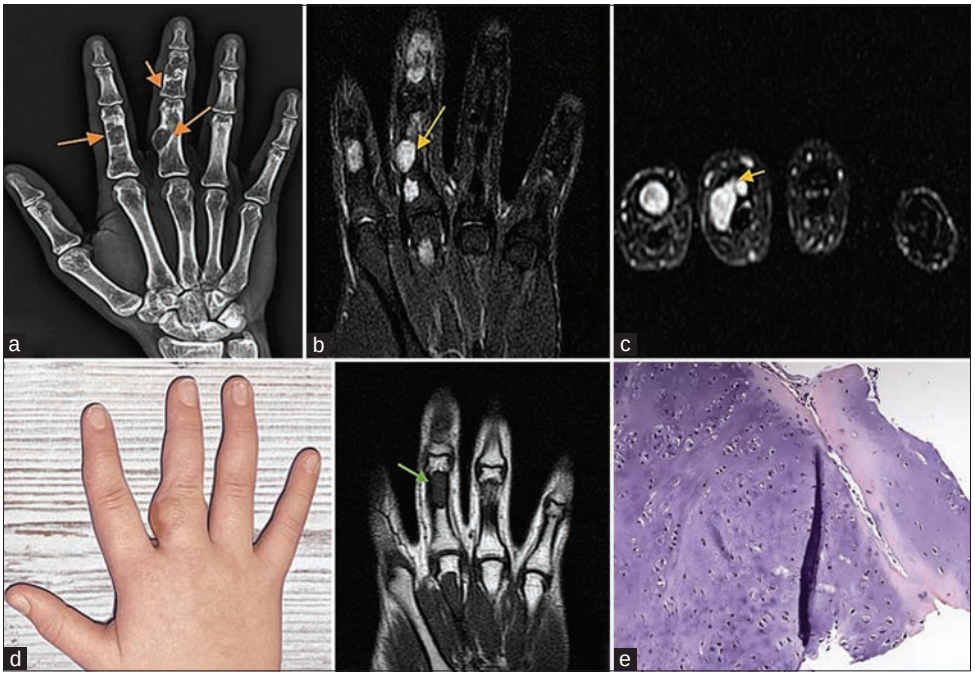

Figure 1: (a) Dorso-palmar radiograph of left hands showing multiple lytic lesions expanding the middle finger metacarpal and phalanges and index finger phalanges (orange arrow) (b and c) Coronal and axial STIR images show hyper-intense lesion (yellow arrow) involving the medullary cavity suggestive of enchondroma in Ollier's shows high degree of cellularity (d) Coronal T1-weighted magnetic resonance image shows well defined lobulated homogenous hypo-intense (green arrow) lesion noted in intramedullary cavity of metacarpal and phalanges (e) H&E image shows Enchondroma arising in the setting of Ollier disease. A cartilaginous tumor with a higher degree of cellularity, composed of spindled chondrocytes with moderate nuclear atypia and an open chromatin pattern with myxoid stroma

often suspected when a cortical breakthrough and soft-tissue involvement are seen.

On MRI, the lesion produces a higher signal on T2-weighted images and a low signal on T1 imaging, similar to other hyaline cartilage foci. Technetium-99m methylene diphosphonate radionucleotide bone scintigraphy is utilized to evaluate the severity of the disease. Reactive bone, ossified enchondromas, and local hyperemia may impact radio-pharmaceutical absorption.[10] Clinical manifestations of new or increased pain are the most sensitive indicators of chondrosarcoma transformation. Emission CT revealed osteoporosis or pathological fracture. Emission computed tomography (ECT) is beneficial in evaluating a variety of lesions that mimic Ollier's illness. In positron emission tomography, enchondromas have recently shown a low degree of 18F-fluorodeoxyglucose absorption. The most sensitive approach for identifying lesions and neoplasia is bone scintigraphy.[5,6]

Multiple hereditary exostoses, a condition in which lesions form on the surface rather than in the center of the bones, is a differential diagnosis for Ollier's disease (with enchondromatosis, lesions occur in the center of the bones).[9] Maffucci syndrome is another kind of multiple enchondromatosis marked by numerous enchondromas associated with soft-tissue hemangiomas and, occasionally, lymphangiomas. Histopathologic examinations are often not performed since multiple enchondromatosis is diagnosed clinically and by distinct radiographic manifestations. Histopathologic examinations are required when malignant change is suspected.[9,10]

Conclusion

Ollier's disease is an uncommon tumor that is easily identified clinically and radiographically by the specific position of enchondromas all through the skeleton, as seen in this case. A thorough grasp of clinical signs and radiographic findings may help to avoid the use of other unnecessary diagnostic imaging modalities. However, in more complicated and painful situations, other diagnostic imaging modalities such as MRI, ultrasonography, and scintigraphy may be used. With more reports and studies, radiographic diagnosis is becoming more precise, and gene mutation studies are becoming more trustworthy, which may provide insight into the most appropriate treatment, avoiding malformation enchondromas. These statistics help patients obtain a more precise diagnosis and achieve a higher success rate with therapy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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Perineural spread in a case of pituitary adenoma

Revati Tekwani, Ajith Varrior

Abstract:

Perineural spread (PNS) is commonly observed with head-and-neck malignancies such as squamous cell carcinoma, adenoid cystic carcinoma, and nasopharyngeal carcinoma. Pituitary adenomas are not known to have PNS, and only a single case has been reported previously. We report a case of a 27-year-old woman who presented with the complaints of headache and visual disturbances. Magnetic resonance imaging of the pituitary showed a macroadenoma extending along the trigeminal nerve through the foramen ovale, the Meckel's cave, the prepontine area, and the superior orbital fissure on the right. Histopathology of the postresection specimen revealed a pituitary adenoma. Imaging features of PNS include thickening with the diffuse enhancement of the nerve, nodularity, enlargement/erosion of the foraminae, effacement of the perineural fat pad, and denervation changes in the muscles. PNS in a tumor has to be actively searched for and reported, as it alters the prognosis and the management.

Keywords:

Cranial nerves, perineural spread, pituitary adenoma

Introduction

Cranial nerve involvement can be attributed to a multitude of pathologies ranging from benign etiologies such as infection, inflammation, granulomatous diseases, and benign nerve tumors to malignant etiology, which can involve the nerve primarily or secondarily in the form of perineural spread (PNS) or metastasis. The PNS of a tumor can be anterograde or retrograde.[1] Although commonly associated with squamous cell carcinoma and salivary gland tumors, PNS has not been associated with pituitary adenomas. We report a case of a pituitary macroadenoma with extension along the cranial nerves.

Case Report

A 27-year-old woman presented with the complaints of headache for 1 year and visual disturbances for 20 days. She had irregular menstrual cycles since past few months. On examination, there was bitemporal hemianopia. There was no other neurological deficit. A magnetic resonance imaging of the pituitary showed a well-defined lobulated T2 hyperintense and T1 isointense lesion in the sella and the suprasellar region [Figure 1]. Laterally, the lesion was extending into the right cavernous sinus with encasement of the internal carotid artery. There was an extra-axial extension into the right inferior temporal region with mass effect. On the left, it was extending till the lateral tangent line. It was compressing the optic chaisma with effacement of the third ventricle. Inferiorly, there was an extension into the sphenoid sinus. The lesion was extending along the mandibular division of the right trigeminal nerve with widening of the foramen ovale [Figure 2]. It was also extending into the Meckel's cave and the prepontine area on the right [Figure 3]. Anteriorly, it was extending into the right superior orbital fissure and into the intraconal compartment [Figure 4]. Based on the above imaging findings, the differentials

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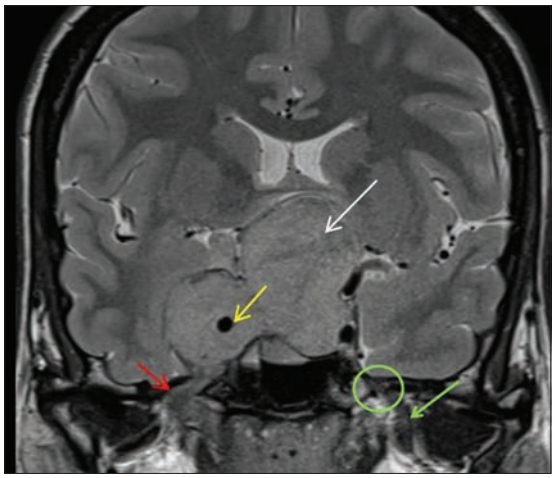

Figure 1: Coronal T2-weighted sequence of the magnetic resonance imaging pituitary shows a well-defined lobulated hyperintense lesion in the sella and suprasellar region (white arrow). There is extension along the mandibular division of the right trigeminal nerve along the foramen ovale with loss of perineural fat pad (red arrow). There is widening of the foramen. There is encasement of the right internal carotid artery (yellow arrow). The contralateral normal mandibular nerve (green arrow) is seen passing through the intact foramen ovale (green circle)

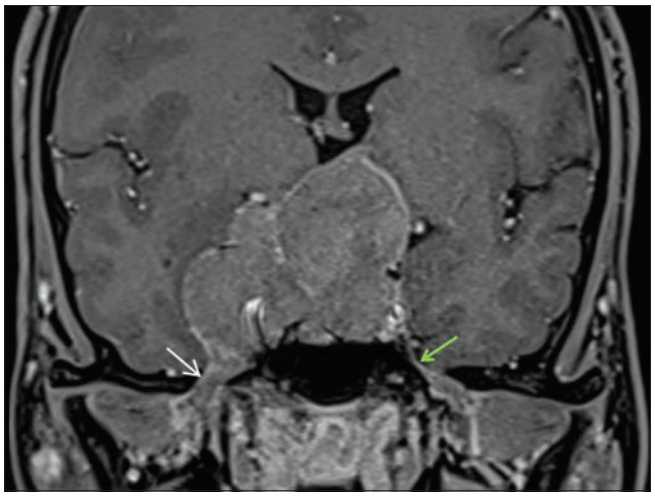

Figure 2: Coronal postcontrast T1 sequence of the magnetic resonance imaging Pituitary shows heterogenous enhancement of the lesion along with enhancing thickening of the mandibular division of the right trigeminal nerve (white arrow) through the enlarged foramen ovale. The contralateral normal mandibular nerve is highlighted by green arrow

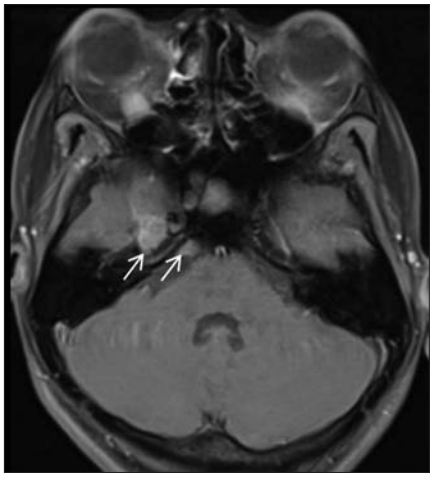

Figure 3: Axial postcontrast T1 sequence of the magnetic resonance imaging Pituitary shows enhancing thickening of the right trigeminal in the Meckel's cave and extending inferiorly into the right prepontine region (white arrows)

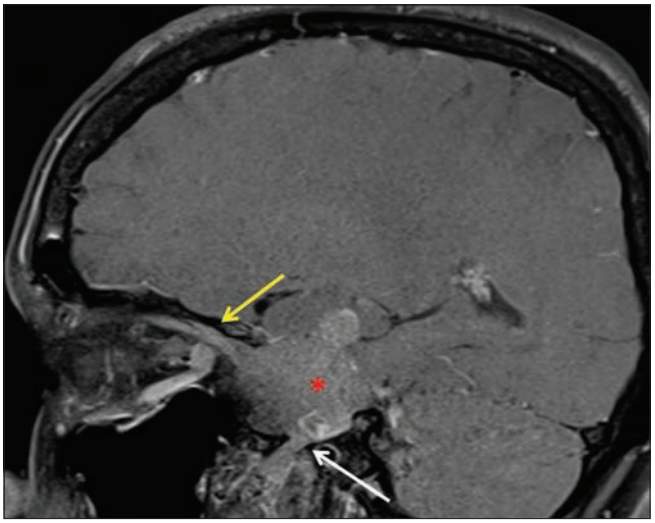

Figure 4: Sagittal postcontrast T1 sequence of the magnetic resonance imaging Pituitary shows the tumor (*) extending inferiorly along the thickened enhancing right mandibular nerve, through the foramen ovale into the infratemporal fossa (white arrow). Anteriorly, there is an extension along the superior orbital fissure (yellow arrow)

were a pituitary macroadenoma with PNS or a collision tumor of a pituitary adenoma with a schwannoma. The patient underwent a transsphenoidal resection of the tumor. Histopathology was suggestive of a pituitary adenoma. Postsurgery, the patient was asymptomatic.

Discussion

PNS is a common entity observed with head-and-neck malignancies.[1] Perineural invasion is a termed reserved for the involvement of any of the layers of the nerve on histopathology and can involve small unnamed nerves. It is not evident on imaging. Compared to that, PNS is the extension of the tumor along a large, typically named nerve which is seen radiologically.[1] Certain tumors that are notorious for PNS include squamous cell carcinoma, adenoid cystic carcinoma of the salivary glands, lymphoma, melanoma, nasopharyngeal carcinoma, and few cases of meningioma.[1,2] The trigeminal and the facial nerves are most commonly involved cranial nerves. It is of paramount importance to detect PNS as it affects the prognosis and the surgical management. Clinically, the nerve involvement is typically mild, slowly progressive, and persistent.[1]

Certain radiological features observed with PNS include thickening with the diffuse enhancement of the nerve, nodularity, enlargement/erosion of the foraminae,

effacement of the perineural fat pad, and denervation changes in the muscles.[1-3] Except for denervation changes, the rest all of these features were present in our case. There can be skip lesions along the course of the nerve. It is crucial to examine the contralateral side and also to keep an eye on anatomical variants.

PNS is not commonly associated with pituitary adenomas. A similar case was reported in 2018,[4] where there was perineural extension along the trigeminal nerve in a case of a pituitary adenoma with apoplexy.

Collision tumors is a term used for two different tumors occurring in the same anatomical region. In the pituitary gland, adenomas are associated with other tumors.[5] Differentials for sellar/suprasellar masses with PNS include hypothalamic-chiasmatic glioma and lymphoma. Sarcoidosis can also present with thickening and enhancement of the pituitary stalk with enhancement of the cranial nerves and the dura.[6] Pituitary tuberculoma masquerading as a macroadenoma has been reported and should be considered as a differential, especially in the Indian subcontinent.[7]

Certain pitfalls include perineural vascular plexus (peripheral enhancement with a hypointense nerve with maintained fat pad), neuritis (shows enhancement, however, no thickening) due to infection/inflammation, or radiotherapy.[1]

Conclusion

PNS of malignancies, which is commonly associated with head-and-neck tumors, significantly alters their management and prognosis. It is odd to see PNS with pituitary adenomas; however, one should be able to identify the radiological features or nerve involvement and to differentiate it from the mimics.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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Maharashtra State Branch of the Indian Radiological and Imaging Association

The Maharashtra State Branch of the Indian Radiological and Imaging Association (MSBIRIA) is a prominent regional chapter of the Indian Radiological and Imaging Association (IRIA), the national organization representing radiologists and imaging specialists in India. Established in 1931 to promote the advancement of radiology and imaging sciences.

After the formation of Maharashtra state in 1960 the state radiology chapter was known as MSRA (Maharashtra State Radiology Association) and renamed MSBIRIA subsequently. MSBIRIA plays a crucial role in the professional development of its members through various educational programs, conferences, and workshops.

MSBIRIA conduct Academic and Welfare activities for Radiologists in Maharashtra and promote social, medical and welfare services for the needy people. First Maharashtra State Radiology Association conference was held at Mumbai then called Bombay in 1973. MSBIRIA is committed to fostering excellence in radiology by encouraging research, disseminating knowledge, and upholding the highest standards of clinical practice. The branch provides a platform for radiologists to collaborate, share insights, and stay updated with the latest advancements in the field of radiology. It also serves as a bridge between its members and the broader national and international radiology communities.

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