

Fetus in fetu - the included twin/ entrapped twin: a case report

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Fetus in fetu is a rare congenital anomaly with fewer than 200 cases reported worldwide. It is a part of spectrum of anomalous embryogenesis. We report a case of an 18-month old boy with abdominal distension. Ultrasonography, X-ray, and computed tomography (CT) scan revealed a retroperitoneal mass suggestive of fetus in fetu. with evidence of organogenesis and a vertebral column, which differentiates it from an organized teratoma. The mass was resected in toto. [Paediatr Indones. 2023;63:418-24; DOI: <https://doi.org/10.14238/pi63.5.2023.418-24>].

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Fetus in fetu is part of a spectrum of malformations, which result from abnormal embryogenesis in a mono chorionic diamniotic pregnancy. The spectrum ranges from conjoined twins at one end, parasitic twins, and embryonic vestigial fetal inclusions/fetus in fetu in between, to fetiform teratoma at the other end. Conjoined twins are a rare developmental anomaly in which twins do not separate completely. There are two types: symmetrical (common) and asymmetrical (rare). Asymmetric twins are of two types- parasitic twin (heteropagus) and fetus in fetu.¹ Fetus in fetu was first described by Johann Friedrich Meckel in the late 1800s.² The incidence is 1 in 500,000 births, with a 2:1 male predominance.³ The majority of cases are detected before 18 months of age as a slow growing intraabdominal mass.⁴ It is regarded as a parasitic twin of diamniotic mono chorionic monozygotic twins.⁵ Less than 200 cases have been reported in the literature

worldwide. Willis was the first to coin the term fetus in fetu.⁶ Most cases are detected in infancy, however, it can be detected at any age, with the oldest reported case at 47 years of age.

The case

An 18-month-old male presented with abdominal distension on the right side of midline, which had persisted since birth. The parents noted that they had felt a lump in the child's right abdomen. There was no history of trauma, fever, loose stools, constipation, vomiting, hematemesis, melena, jaundice, weight loss, or loss of appetite. There were no urinary complaints or history of third degree consanguineous marriage of the parents. Birth history consisted of a full term, normal, vaginal delivery; the baby cried immediately after birth. Developmental history was normal. Liver and renal function tests, as well as hemogram were

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normal. Plain erect abdominal radiograph revealed multiple radiopacities in the right hypochondriac and lumbar regions (**Figure 1**).

Ultrasonography (US) revealed a large, well-defined, solid mass of heterogeneous echotexture measuring approx. 12 x 8 x 10 cm (length x antero-posterior x transverse) in the retroperitoneum of the right upper abdomen supra-renal renal region extending inferiorly in the right lumbar region, compressing and displacing the right kidney inferolaterally just beneath the lateral abdominal wall (**Figure 2**). The pancreas was compressed and displaced to the left. Superiorly, the mass was causing extrinsic compression on the medial surface of right hepatic lobe and gall bladder. The echogenic areas of the mass were suggestive of fat, while the hypoechoic areas suggested a soft tissue component, and linear echoreflexive areas of calcification or bone. No significant vascularity was noted within the mass. The possibility of retroperitoneal teratoma was raised.

Computed tomography (CT) scan of the abdomen and pelvis revealed a large, well-defined mass, with lobulated outlines of approximately 12.7 x 9.1 x 10.1 cm (craniocaudal x transverse x antero-posterior) in the retroperitoneum of the right hypochondrium, right lumbar and umbilical region (**Figure 3**). Superiorly, it was causing extrinsic compression on the right hepatic lobe and gall bladder. Inferiorly, it was causing extrinsic compression on the right kidney, which was displaced to the right inferolaterally. A mass effect was noted on the right renal artery, which was compressed and displaced inferiorly. It received an arterial supply from the abdominal aorta whose branches were seen in the soft tissue component of the mass. The portal vein was displaced anteriorly. The hepatic portion of the inferior vena cava (IVC) was markedly compressed and displaced anteriorly (**Figure 4**). The infrahepatic portion of the IVC was compressed and displaced laterally, just beneath the right lateral abdominal wall up to the level of the renal hilum. The infrarenal IVC



Figure 1. Plain radiograph erect abdomen showing multiple radiopacities in the right hypochondriac and lumbar regions.

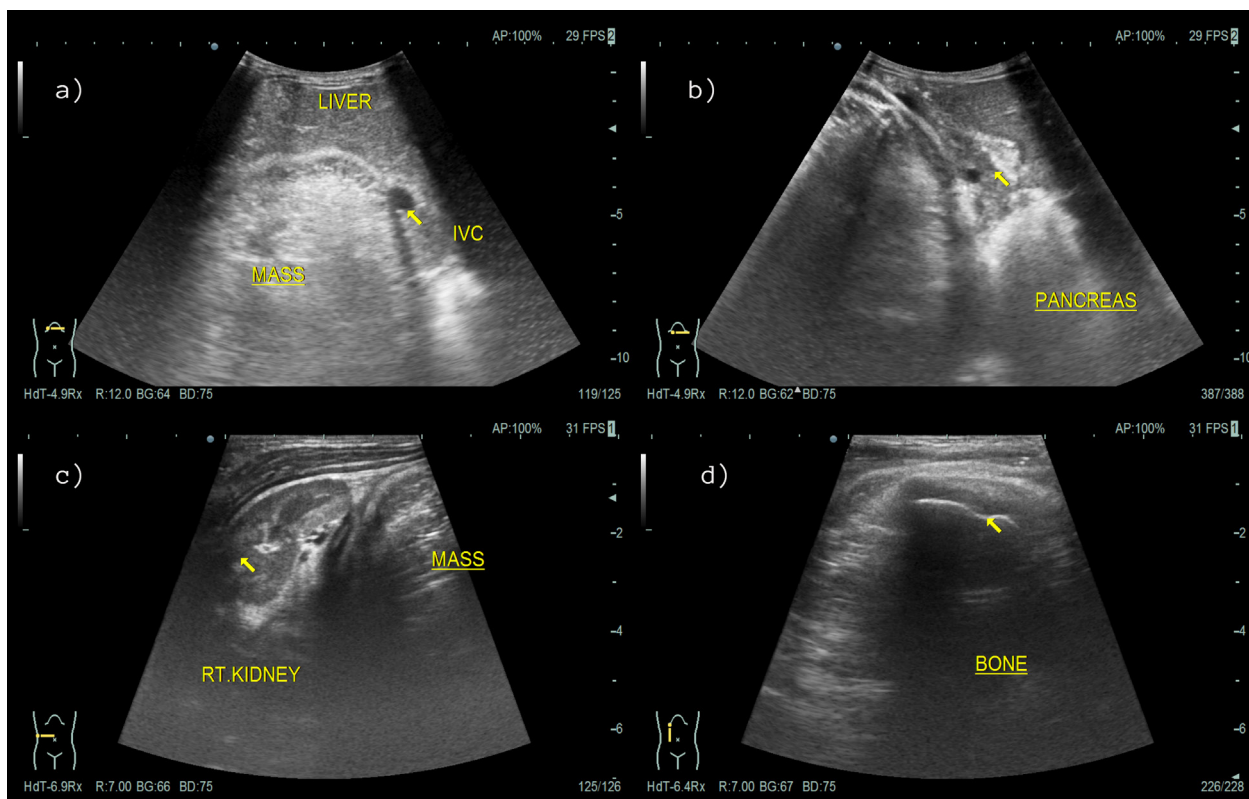


Figure 2. Transabdominal ultrasound using 3-5Hz curvilinear probe was performed. (a) It revealed an ill-defined heterogeneous retroperitoneal mass in the right hypochondrium inferior to the right lobe of liver showing echogenic fat. (b) Medially, the mass caused a mass effect on the pancreas. (c) The right kidney was displaced inferolaterally and was just beneath the lateral abdominal wall. (d) The mass revealed linear echorefective areas within, representing bony structures.

was coursing medially to reach its normal position at the level of aortic bifurcation. A mass effect was seen on the adjoining bowel loops and colon. The mass showed a fat component (CT value: -100 to -200 Hounsfield units), and multiple areas of soft tissue components (CT value: 20 to 40 Hounsfield units) showing mild post-contrast enhancement and multiple bones. Vertebral bodies, pelvic bones, scapulae, ribs, femur, tibia, fibula, metatarsals and phalanges were identified (Figure 3). The rest of the abdomen was normal. The right adrenal gland was distinctly not seen. Diagnosis of fetus in fetu (FIF) in the retroperitoneum of the aorto-caval region was made. Differential diagnosis included a well-organized teratoma. The patient underwent surgery and the mass was resected in toto (Figure 5). Arterial feeders from the mass emanated from the right renal artery mass, which was adherent to the IVC.

This case report followed all ethical standards for research. Written informed consent was obtained from the patient's parents for publication, along with relevant images. Patient identity was not disclosed.

Discussion

Variations of abnormal conjoined twinning include asymmetric conjoined twins (parasitic twin), enclosed fetus in fetu, teratoma, and acardiac twins. Such fetuses vary in the extent of damage or defect as well as site of union to the normal twin. The absence of a functional heart and competent brain in these examples suggest that their etiology is a primary cardiac malformation with a secondary disruption in brain development.⁷ Fetus in fetu is part of a spectrum of anomalous embryogenesis, which includes

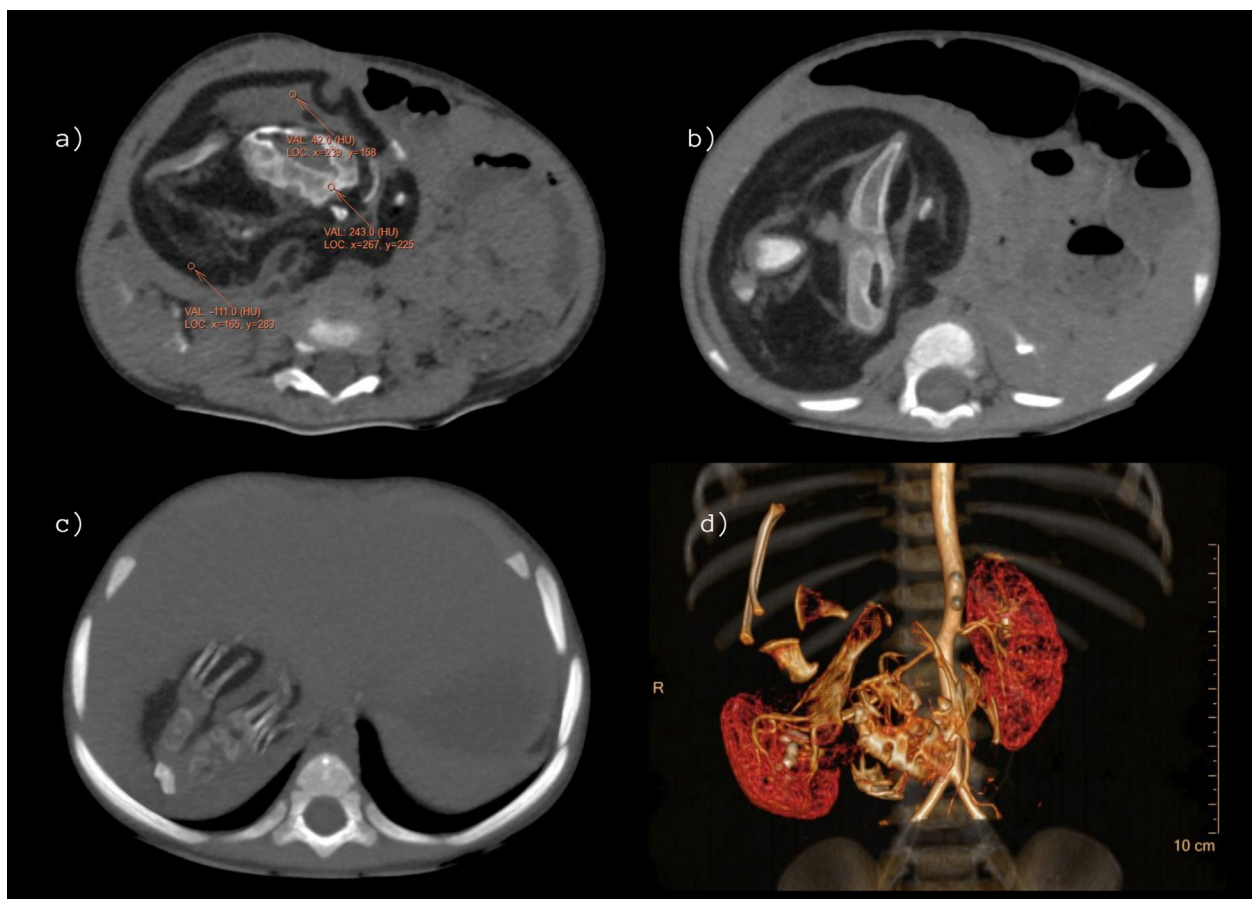


Figure 3. Non-contrast computed tomography (NCCT) of abdomen and pelvis showing: (a) multiple areas of fat, bony (vertebral bodies), and soft tissue components; (b) pelvic bones; (c) digits and tarsal bones; (d) coronal 3D bony reconstruction of the mass in relation to the abdominal aorta and the renal arteries bilaterally.

twin reversed arterial perfusion, organized teratoma, embryonic vestigial fetal inclusions, parasitic fetus, and conjoined symmetric twins.⁸

According to the monozygotic twin theory, fetus in fetu is a result of unequal division of totipotent cells of the blastocyst. Fetus in fetu represents a diamniotic monozygotic twin that gets engulfed into the host twin's body after anastomosis of vitelline circulation. The existence of multiple fetus in fetu in a single host and fetus in fetu coupled with teratoma can be explained by the monozygotic dichorionic diamniotic twin theory.⁹

If a blastocyst derived from the conceptus embeds into another blastocyst instead of implanting in the endometrium, or if blastocysts are encompassed by another implant close together, it leads to the inclusion of a cell mass within a maturing sister

embryo, resulting in the formation of a fetus in the fetus. The fetus in fetu initially develops in a manner similar to that of normal fetus. Genetic marker studies and genotyping suggest that the fetus in fetu and host infant are genetically identical. This finding supports the monozygotic twin theory.¹⁰ The presence of a vertebral column in a fetus indicates that it descended from the primordial streak. Fetus in fetu can be located at sites where teratomas are often found, such as the retroperitoneum and ovaries. Fetus in fetu can be seen along with teratomas. Teratoma formation can occur following removal of fetus in fetu.¹¹

Spencer was one of the first to propose criteria for classifying any fetiform mass as a fetus in fetu. According to the criteria it must (a) be enclosed within a distant sac, (b) be covered partially or completely by skin, (c) have recognizable anatomic

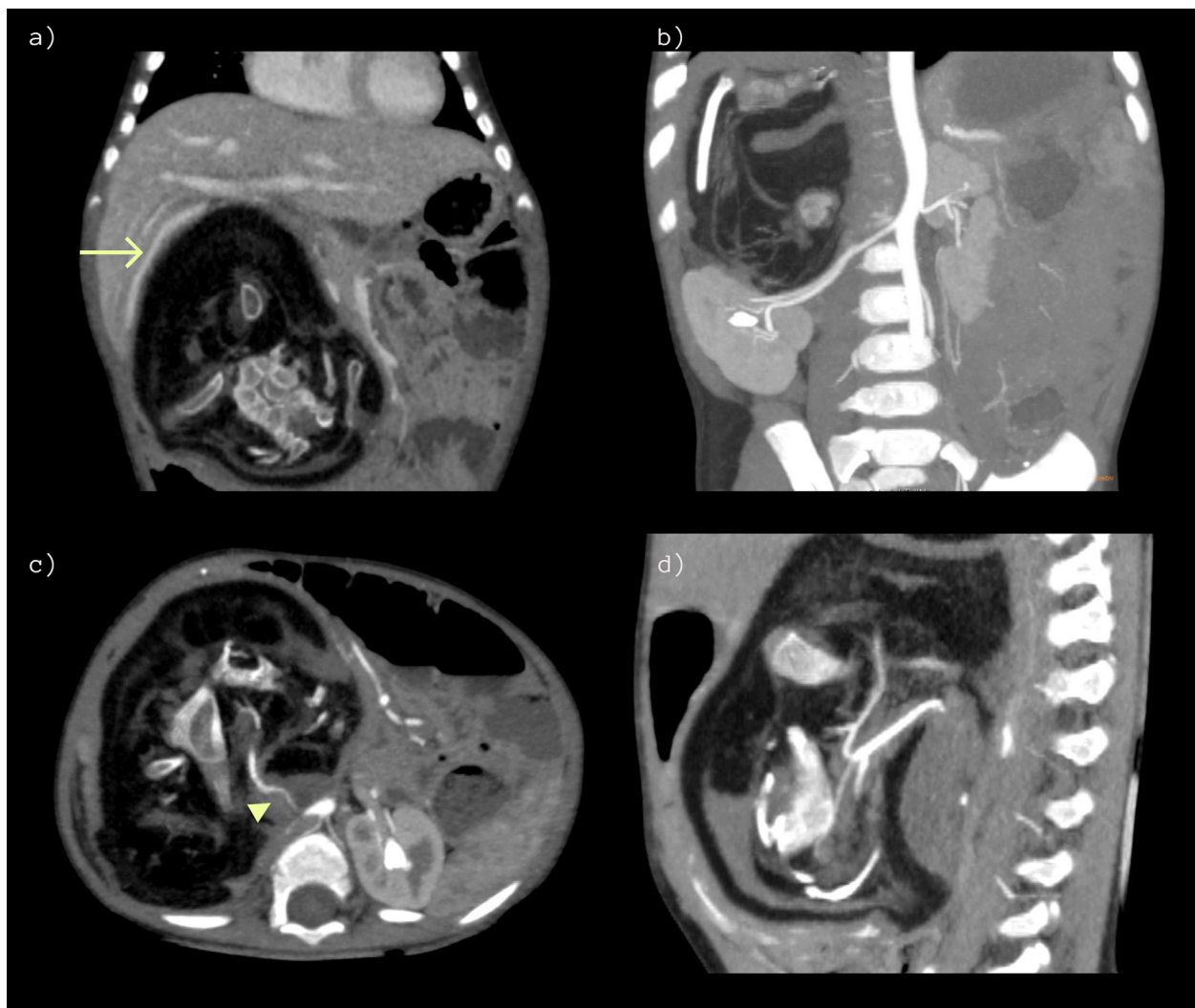


Figure 4. Contrast enhanced CT abdomen and pelvis including: (a) venous phase showing compression and displacement of the hepatic portion of IVC; (b) arterial phase showing inferiorly displaced right kidney and right renal artery; (c) and (d) arterial feeders (arrowhead) to the mass arising from the proximal portion of right renal artery.

parts, (d) be attached to its host by a few relatively large blood vessels, and (e) be adjacent to one of the conjoined twins' attachment sites.¹²

Willis proposed criteria in 1935 to differentiate fetus in fetu from teratoma. The criteria are: (i) the presence of a separate spinal column indicating that the fetus has passed through the primary stage after gastrulation involving formation of the neural tube, metamerization, and symmetrical development around this axis, and ii) development of organs in a synchronized manner, indicating they have achieved same degree of maturation.¹³

Fetus in fetu is usually seen in the upper retroperitoneum, but has been rarely found in the gastrointestinal tract (GIT), central nervous system, lungs, genitourinary tract, adrenal glands, pancreas, liver, spleen, lymph nodes, pelvis, scrotal sac, mesentery, right iliac fossa, oral cavity, and sacrococcygeal regions.¹⁴ The common retroperitoneal location can be explained by the development of the superior mesenteric artery from the vitelline circulation.⁴ Absence of an independent blood supply explains the fetal growth restriction in all cases.

Brain tissue and intestine were detected in about

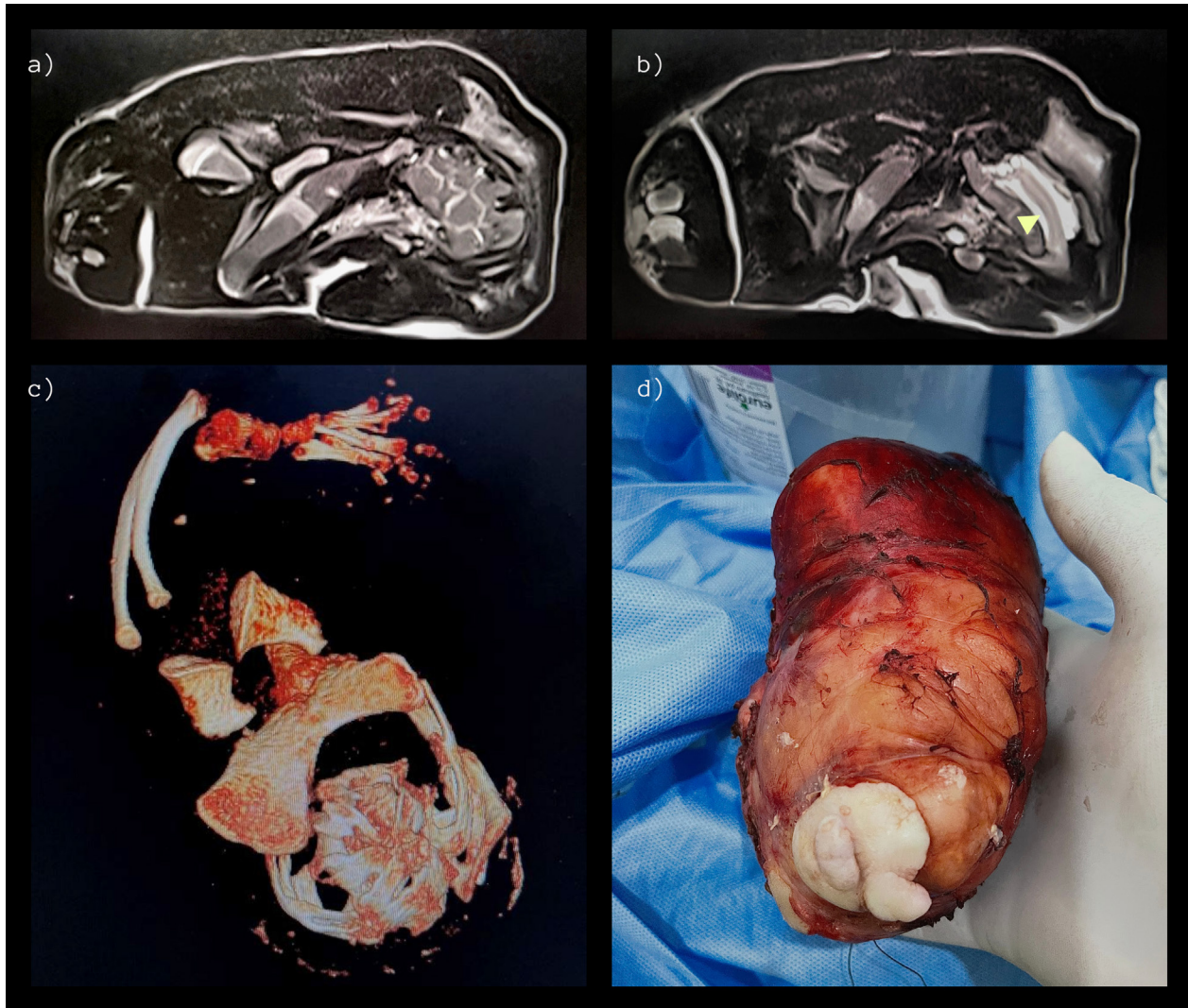


Figure 5. (a) and (b) T2 weighted image (T2WI) magnetic resonance imaging of the post-operative specimen, arrowhead reveals vertebral column; (c) NCCT of the post-operative specimen depicting the bony organization; (d) gross specimen of the retroperitoneal mass.

half of cases. Thyroid, parathyroid, pancreas, kidney, spleen, adrenal, testis, ovaries, tongue, salivary glands, urinary bladder, lymph nodes, teeth, and trachea are rare, but have also been identified. The mass may have an umbilical cord with 2 vessels, but placenta or chorionic villi are not seen.⁷ Different organs have been reported in fetus in fetu, with vertebral columns (91%) and limbs (82.5%) being the most commonly visualized, followed by CNS (55.8%), GIT (45%), and vessels (40%).¹⁴ Fetus in fetu is usually anencephalic, and enclosed within a sac containing fluid or sebaceous material.¹⁵ An umbilical cord-like structure connects the fetus in fetu and the host. It is

usually acardiac. In children, a fetus in fetu presents due to its mass effect, causing abdominal distension, feeding difficulty, dyspnea, and mass effect on kidneys. The main blood supply of the sac is from the plexus, where it is attached to the abdominal wall.⁵

Imaging by radiograph, ultrasound, CT scan, and MRI can assist in making a correct diagnosis of fetus in fetu. Abdominal radiograph can identify a vertebral column, long bones, or both. Rarely, a vertebral column may not be identified if it is underdeveloped and dysplastic. CT scan can identify a vertebral column, axial bones, long bones, fat, and soft tissue components. Ultrasound is useful for detecting

echogenic fat, linear echoreflexive bones, soft tissue components, and cystic structures. MRI detects fat and bones. A 3D CT scan is extremely useful in pre-operative evaluation, as the fetus in fetu mass reveals all its contents, its blood supply and its relations with respect to other abdominal structures of the patient.

Complete surgical resection along with the surrounding sac is suggested, as the mass can grow and cause mass effect on adjoining structures. Leakage of sac contents can cause infection, hemorrhage, and pleuro-peritoneal inflammation. Though benign, a single case of malignant degeneration was reported.¹¹ The treatment of choice would include complete resection of the tumor with an intact membrane. Due to the risk of malignant recurrence, interval monitoring of alpha-fetoprotein levels should be done.

Conflict of interest

None declared.

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Disclaimer

The views and opinions expressed in this article are those of the authors and not an official position of the institution.

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