Case Report

Fetus-in-fetu in the Pelvis: Report of a Case and Literature Review

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Abstract

Introduction: Fetus-in-fetu is an extremely rare condition in which a malformed fetus is found in the body of its twin. To our knowledge, fewer than 100 cases have been reported. Wide variations of presentation have been described, although its embryo-pathogenesis and differentiation from a teratoma have not been well established. Clinical Picture: We describe a male neonate with a fetoid-like mass in his pelvis associated with bilateral undescended testes. The mass was detected on prenatal ultrasound scans. The diagnosis of fetus-in-fetu was considered prenatally and confirmed on a computed tomography scan after birth. Outcome: The mass was successfully excised. Histological examination, accompanied by a review of the literature, confirmed that the mass had features consistent with a fetus-in-fetu. Conclusions: Although an extremely rare clinical entity, fetus-in-fetu can be diagnosed prior to surgery with current imaging modalities. When it arises in the retroperitoneum of a male infant, it can hinder the descent of the testes. Complete excision is curative.

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Key words: Antenatal diagnosis, Teratoma, Undescended testes

Case Report

Baby A was delivered at 37-week and 4-day gestation via elective caesarean section. His mother was a healthy 33-year-old with a previous uneventful pregnancy. There was no history of consanguinity and no family history of multiple pregnancies. In the current pregnancy, there was no maternal illness, history of drug intake or exposure to radiation. A fetal anomaly scan at 20-week gestation showed a grossly normal, singleton fetus. An intra-abdominal mass in the fetus was detected on a growth scan at 31 weeks. A repeat scan at 33 weeks confirmed the presence of a cystic mass with solid components. It was behind the bladder and measured 65 mm x 31 mm with a volume of 25 cm^3. The solid component within measured 29 mm x 16 mm x 16 mm. The mass was thought to represent either a well-differentiated teratoma or a fetus-in-fetu.

Baby A weighed 3200 grams at birth and had good APGAR scores. Abdominal examination revealed a large pelvic mass. Both his testes were not palpable. Beta-human chorionic gonadotropin (β-HCG) was raised at 63.5 IU/L and α-fetoprotein (AFP) was >176,750 μg/L.

Postnatal ultrasonography confirmed the presence of a lower abdominal mass, measuring 30.8 mm x 38.2 mm in transverse diameter and 32.3 mm x 37.1 mm in anterior-posterior diameter. There appeared to be a segment of the spine lying within the mass and a spinal cord within the bony vertebrae (Fig. 1). There was also an umbilical cord-like pedicle. A computed tomography (CT) scan of the abdomen showed a large 60 mm x 60 mm x 50 mm heterogeneous well-encapsulated pelvic-abdominal cystic mass, displacing the bladder anteriorly and the bowel loops superiorly. There was an oval soft tissue mass with ossific components within the cyst. The ossific components resembled that of a vertebral column with a rudimentary rib cage, as well as a femur (Fig. 2). A preoperative diagnosis of fetus-in-fetu was made.

Elective laparotomy was performed. We found a large retroperitoneal cystic mass that arose from the pelvis and extended superiorly to just below the bifurcation of the aorta. It was densely adherent to the sacrum. The bladder was displaced anteriorly and was floppy and distended. The mass represented a tense amniotic sac with a fetoid

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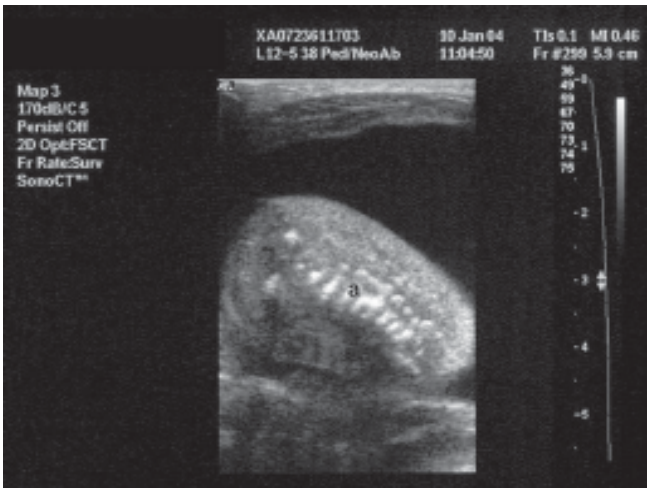

Fig. 1. Ultrasound picture showing a complete vertebral column (a) within the cystic mass.

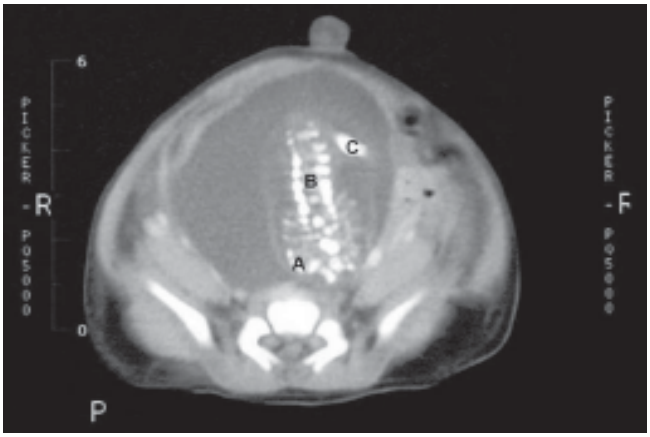

Fig. 2. Computed tomography scan of baby A's abdomen showing the fetoid mass within its amniotic sac, and the presence of a rudimentary rib cage (A), the vertebral bodies (B), and a left femur (C).

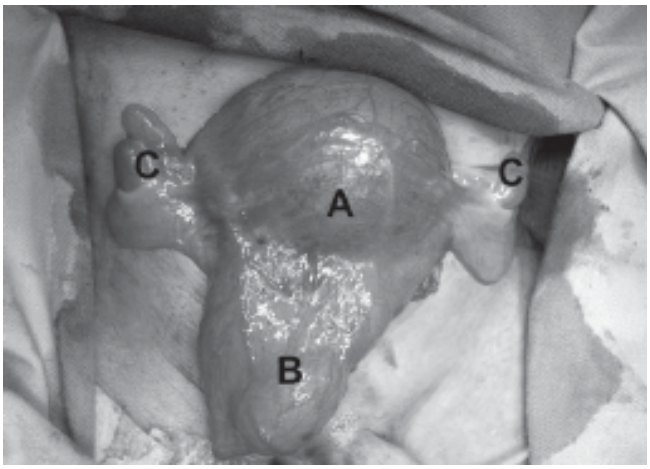

Fig. 3. Intraoperative picture demonstrating the cystic mass (A) in relation to the bladder (B) and bilateral intra-abdominal testes (C).

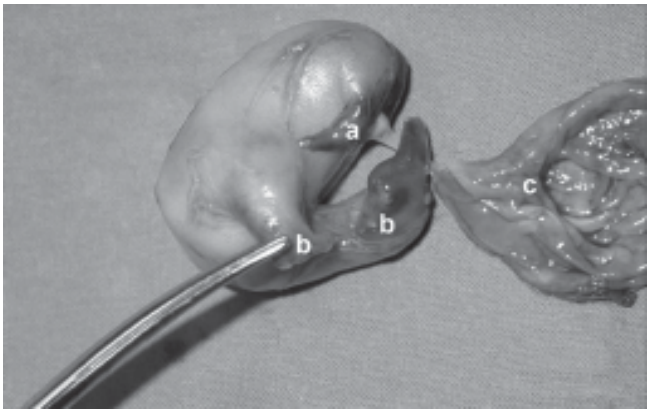

Fig. 4. Photograph of the dissected fetus demonstrating its rudimentary right upper limb (a), the lower limbs with malformed digits (b), and its attachment to the amniotic sac (c) via an umbilical cord-like structure.

mass within. The sac contained clear fluid and the fetus within had grossly visible limbs. The blood supply was via multiple small posteriorly situated vessels. Both testes were intra-abdominal, each measuring about 0.5 mLs (Fig. 3).

The cystic component was decompressed and the mass dissected off the retroperitoneum after ligating the feeding vessels. Both testes were brought down and anchored to the scrotum. Baby A did well after the surgery and was discharged on the 6th postoperative day.

Macroscopic examination revealed a soft tissue mass resembling a fetus attached to the membranous sac via a 10 mm x 1 mm cord-like structure. The mass weighed 20 grams and measured 45 mm x 25 mm x 25 mm. There were 2 malformed lower limbs: the right measured 20 mm long, with a foot measuring 12 mm, and the left measured 35 mm long with a 14-mm foot. There were 4 rudimentary digits on the left foot and a big toe with 2 rudimentary digits on the right foot. There was also a rudimentary right upper limb measuring 10 mm long and 2 mm in diameter with a finely tapered distal end. A raw area at the cephalic end measured 13 mm in diameter (Fig. 4). Further dissection of the specimen revealed a spinal cord housed in the complete spinal column. The body cavity showed a tubular branched structure with a proximal blind end and the distal end opening to the external surface. No kidney, heart or gonad was identified.

Microscopic examination confirmed the presence of amniotic membrane, dermal epithelium lining with normal subcutaneous tissue, bone and cartilage. Portions of colon with myxomatous wall, spinal cord with occasional neuro-epithelial rosettes and ganglia, as well as minimal amounts of marrow tissue were also identified. Fibroblast cultures taken from the skin confirmed a male 46XY karyotype.

Three months after the surgery, baby A's serum AFP was 69.0 μg/L and β-HCG was <1.2U/L. He remained well at a review 9 months after the surgery.

Discussion

Fetus-in-fetu, a term quoted by Lewis,[1] was first described by Meckel to describe a rare condition in which a malformed parasitic twin was found inside the body of its partner, usually in the abdominal cavity. It represents an aberration of monozygotic diamniotic twinning in which unequal division of the totipotent inner cell mass of the developing blastocyst leads to the inclusion of a smaller cell mass within a maturing sister embryo.

It has an incidence of 1 in 500,000 births,[2] with fewer than 100 reported cases worldwide. The majority of these cases present in infancy, with the oldest reported case occurring in a 47-year-old man.[3] Thakral et al[4] reported equal male and female predisposition but Patankar et al[5] and Federici et al[6] noted a 2:1 male predominance. This is in contrast to teratomas, which show a female preponderance. The commonest presentation is an abdominal mass that is typically located in the upper retroperitoneum. Other more unusual sites, including the cranial cavity, oral cavity, sacrococcygeal region and scrotum, have been reported.[3-6] In contrast, teratomas usually arise in the lower retroperitoneum. The fetus is usually single, although up to 5 fetuses have been found in the skull of a newborn.[7]

It remains controversial whether fetus-in-fetu is a distinct entity or represents a highly organised teratoma. Du Plessis et al[8] reported an interesting patient with both a well-formed fetus-in-fetu and a malignant teratoma, stating that that was "a potential triplet situation gone awry, resulting in the host, his parasitic twin and a teratoma arising from a third embryo which may have escaped the influence of a primary organizer". Previous investigators have hypothesised that fetus-in-fetu results from a modified process of twinning, and have traced a progression from normal twins to conjoined symmetrical twins, through parasitic fetuses and fetal inclusion, and finally to teratoma.[9]

Willis[10] believes that teratomas originate from the early separation of a focus of multi-potential tissue in the growing embryo that develops in a chaotic way in the host organism. According to Willis, the presence of an axial skeleton distinguishes a teratoma from a fetus-in-fetu.[10] The presence of a separate spinal column indicates that the fetus has passed through a primary stage of gastrulation, involving formation of neural tube, metamerisation, and symmetrical development around this axis. However, not all the reported cases of fetus-in-fetu have an identifiable spine.[6,11-13] Metamerisation is believed to have nevertheless occurred in the presence of well-differentiated limbs with hands, phalanges, and nails. The presence of nervous tissue plexuses in the digestive tube, and melanocytes in the skin also implied the existence of notochord during development.[12] Federici et al[6] and Eng et al[11] proposed that in the presence of structures with an advanced grade of fetal organisation such as eyes, parts of the central nervous system, well-developed limb-like processes, skin and colon, the diagnosis of fetus-in fetu can be applied, even in the absence of a real axial structure.

Willis' theory has been supported by genetic, cytogenetic, and serologic assessments. Studies on blood group systems and chromosome cultures from the host and fetus showed no difference between the two.[2,14] When intra-abdominal gonads were found, these corresponded histologically to the sex of the bearer.[11,13] These studies virtually excluded dizygosity or aneuploidy, and indicated that the fetus-in-fetu was derived from the same zygote tissue as its host.[11] Our baby and his included fetus shared the same 46XY karyotype, thereby supporting Willis' theory. Early incorporation of the parasitic twin into the abdomen of the host would explain the ultrasonographic finding of a singleton fetus at 20 weeks of gestation.

Symptoms of fetus-in-fetu relate mainly to its mass effect and include abdominal distension, feeding difficulty, emesis, jaundice, pressure effects on the renal system and dyspnoea.[1,3,6,13] de Lagausie et al[12] reported signs of maceration with the threat of consumptive coagulopathy in their patient. Occasionally, the anomaly is asymptomatic.[15] In our patient, the mass hindered the descent of both testes. To our knowledge, this is the first reported case where a fetus-in-fetu had resulted in bilateral undescended testes in the host. In addition, our patient also had a floppy and distended bladder, consistent with the longstanding mass effect.

The preoperative diagnosis of fetus-in-fetu hinges on the radiological findings. Plain abdominal X-rays may be helpful in diagnosis, with up to about half of the cases showing the presence of a vertebral column and axial skeleton.[13] However, up to the mid-1990s, fewer than a quarter of the cases were diagnosed prior to surgery.[6,13] The use of CT scans has since enhanced the accuracy of preoperative diagnosis. The ability to diagnose fetus-in-fetu on prenatal ultrasonography was first reported by Nicolini et al in 1983.[16] This had accurately suggested the diagnosis of fetus-in-fetu in our patient. We were then able to provide appropriate prenatal counselling. In recent years, magnetic resonance imaging has also been used to diagnose 4 cases of fetus-in-fetu.[13]

Serum AFP levels in the host may be normal or elevated in cases of fetuses-in-fetu.[11,12,15] The maternal AFP level was also raised in at least 1 case.[17] We expect tumour marker levels to normalise postoperatively.

The fetus is typically suspended by a pedicle within a complete sac containing fluid or sebaceous material. The umbilical cord-like structure may consist of only 2 vessels.[1-3,5,6,11-13] There was no placenta or chorionic villi at the point of attachment of sac to the host, except in 1 case in which the primitive chorionic villi were seen.[18]

Definite vascular connections to the host are rarely described.[2,3,5,13] Usually, the wall of the capsule is thickened at the point of attachment of the pedicle and is oriented towards the superior mesenteric artery or the base of mesentery.[3,17] Mohta et al[17] hence proposed that the vascular supply to the mass is usually from the superior mesenteric artery. We tend to agree with Heifetz et al[19] who reported that the predominant blood supply appears to be derived from the plexus where the fetus-in-fetu and the sac are attached to the host's abdominal wall.

Cases of reported fetuses-in-fetu weighed between 13 grams[20] and 2000 grams.[5] The size of the fetus is likely to be related to its blood supply. Fetuses with distinct vascular connections to the host are relatively bigger with better-developed features.[5,11,13] Our included fetus had recruited its blood supply from the pre-sacral plexus but weighed only 20 grams. The absence of umbilical vessels and a definite vascular connection explained its growth retardation and arrest in organ system differentiation.

It did, however, have distinct fetoid features with a complete spinal column. It was anencephalic and acardiac. Its upper limb was less developed compared to the lower limbs. These are features shared amongst most fetuses-in-fetu.[1-3,6,10-13] The absence of a cardiovascular system may lead to the misdiagnosis of acardiac fetus.[3,13] Although the gross morphology is otherwise similar, an acardiac twin fetus frequently has an abnormal karyotype. The karyotype of fetus-in-fetu is normal and similar to that of the host. Sutthiwan et al[21] reported a case of ectopia cordis in their fetus-in-fetu, although there was no mention of microscopic confirmation of cardiac tissue. To our knowledge, there has only been 1 reported case of cardiac activity in a resected fetus. That fetus had a pseudo-cardiac mass with a heartbeat that was of a different rate to that of its host. Histopathology of the mass showed characteristics of cardiac muscles.[12]

Although there was little evidence of organogenesis in our included fetus, mature somatic tissues from ectodermal (neuro-epithelium, ganglia), endodermal (colon) and mesodermal (bone and cartilage) origins were present. These, together with the presence of a complete vertebral column with appropriate arrangement of the limbs around it, distinguish our fetus from a teratoma.

Complete excision of the surrounding membrane, as in our case, should ensure definitive cure.[1,10,15,19] Hopkins et al[15] reported malignant recurrence following resection of a fetus-in-fetu. This was presumably caused by transformation of adherent membranes remaining at the surgical site. In view of this report, early and frequent postoperative surveillance is recommended, especially if the sac cannot be completely resected. This may be achieved with moderate sensitivity using serum tumour marker concentrations.[15] To detect tumours that do not secrete AFP or β-HCG, regular imaging may be warranted. As our patient's AFP levels normalised after the surgery, we elected to monitor him with postoperative AFP levels and to perform repeat imaging should the AFP levels become elevated.

In conclusion, fetus-in-fetu is a rare and interesting entity that typically presents as an abdominal mass in infancy or early childhood. When it arises in the retroperitoneum, it may hinder the descent of the testes in a male infant. Using current imaging modalities, it can be diagnosed fairly accurately before surgery. Complete excision is curative and allows confirmation of the diagnosis.

REFERENCES

1. Lewis RH. Foetus in foetu and retroperitoneal teratoma. Arch Dis Child 1960;36:220-6.
2. Grant R, Pearn JH. Foetus-in-foetu. Med J Aust 1969;1:1016-20.
3. Dagradi AD, Mangiante GL, Serio GF, Musajo FG, Menestrina FV. Fetus-in-fetu removal in a 47-year-old man. Surgery 1992;112:598-692.
4. Thakral CL, Maji DC, Sajwani MJ. Fetus-in-fetu: a case report and review of the literature. J Pediatr Surg 1998;33:1432-4.
5. Patankar T, Fatterpekar GM, Prasad S, Maniyar A, Mukherji SK. Fetus-in-fetu: CT appearance—report of two cases. Radiology 2000;214: 735-7.
6. Federici S, Ceccarelli PL, Ferrari M, Galli G. Zanetti G, Domino R. Fetus-in-fetu: report of three cases and review of literature. Pediatr Surg Int 1991;6:60-5.
7. Kimmel DL, Moyer EK, Peale AR, Winborne LW, Gotwals JE. A cerebral tumor containing five human fetuses. A case of fetus-in-fetu. Anat Rec 1950;106:141-58.
8. Du Plessis JPG, Winship WS, Kirstein JD. Fetus in fetu and teratoma. S Afr Med J 1974;48:2119-22.
9. Gross RE, Clatworthy HW. Twin fetuses-in-fetu. J Pediatr 1951;38: 502-8.
10. Willis RA. The borderland of embryology and pathology. 2nd ed. Washington DC: Butterworths, 1962:442-62.
11. Eng HL, Chuang JH, Lee TY, Chen WJ. Fetus-in-fetu: a case report and review of the literature. J Pediatr Surg 1989;24:296-9.
12. de Lagausie P, de Napoli Cocci S, Stempfle N, Truong QD, Vuillard E, Ferkadji L, et al. Highly differentiated teratoma and fetus-in-fetu: a single pathology? J Pediatr Surg 1997;32:115-6.
13. Hoeffel CC, Nguyen KQ, Phan TT, Truong NH, Nguyen TS, Tran TT, et al. Fetus-in-fetu: a case report and literature review. Pediatrics 2000;105:1335-44.
14. Boyce MJ, Lockyer JW, Wood CB. Foetus in foetu: serological assessment of monozygotic origin by automated analysis. J Clin Pathol 1972;25: 793-8.
15. Hopkins KL, Dickson PK, Ball TI, Ricketts RR, O'Shea PA, Abramowsky CR. Fetus-in-fetu with malignant recurrence. J Pediatr Surg 1997;32:1476-9.
16. Nicolini U, Dell'Agnola CA, Ferrazzi E, Motta G. Ultrasonic prenatal diagnosis of fetus-in-fetu. J Clin Ultrastruct 1983;11:321-2.
17. Mohta A, Shrivastava UK, Sodhi P, Upreti L. Fetus-in-fetu. Pediatr Surg Int 2003;19:499-500.
18. Lamabadusuriya SP, Atukorale AW, Soysa PE, Walpita PR. A case of fetus in fetu. Arch Dis Child 1972;47:305-7.
19. Heifetz SA, Alrabeeah A, Brown BS, Lau H. Fetus-in-fetu: a fetiform teratoma. Pediatr Pathol 1988;8:215-26.
20. Lee EY. Foetus in foetu. Arch Dis Child 1965;40:689-93.
21. Sutthiwan P, Sutthiwan I, Tree-trakan T. Fetus in fetu. J Pediatr Surg 1983;18:290-2.