Pediatr Surg Int (2009) 25:289–292
DOI 10.1007/s00383-009-2328-8

CASE REPORT

Fetus in fetu: two case reports and review of literature

Jamir D. Arlikar · Shivaji B. Mane · Nitin P. Dhende · Yogendra Sanghavi · Arvind G. Valand · Pradip R. Butale

Accepted: 16 January 2009 / Published online: 31 January 2009

Abstract Fetus in fetu is a rare disorder. Its embryopathogensis and differentiation from teratoma has not been well established. It is a parasitic twin of a diamniotic monozygotic twin. Here we report, two cases of fetus in fetu with review of literature. In case report 1, a 2-year-old boy was referred for asymptomatic lump in abdomen since birth. X-ray showed the mass in the abdomen with some calcification and fluid inside. CT scan reported a heterogenous mass in the retroperitoneum with bony malformation. CT showed presence of three vertebrae in it. After surgically excising the mass and opening the sac it showed presence of trunk and two limbs with one of the limbs having a nail. Histopathology showed presence of GI tract. In case report 2, 4 month female was found to have lump in the abdomen by housemaid while bathing. X-ray showed a soft tissue shadow while ultrasonography revealed cystic mass arising from right kidney. CT suggested cystic mass with calcification not arising from kidney. During exploration whole mass was excised and there was frank fetus inside it. Histopathology confirmed presence of four vertebral bodies with germ layers. Although fetus in fetu is rare condition, correct diagnosis using imaging can be made before surgery. Complete excision is curative.

J. D. Arlikar (✉) · S. B. Mane · N. P. Dhende · Y. Sanghavi · A. G. Valand · P. R. Butale
Grant Medical College, Mumbai, India
e-mail: jamirarlikar@gmail.com

J. D. Arlikar · S. B. Mane · N. P. Dhende · Y. Sanghavi · A. G. Valand · P. R. Butale
Sir JJ Group of Hospitals, Mumbai, India

Introduction

Fetus in fetu is a rare condition with less than 200 cases reported in the world to the best of our knowledge. Meckel first described this condition. It was Willis who coined the term Fetus in fetu. In this condition, malformed parasitic twin is found inside the body of its partner usually the abdominal cavity. Though the most common site is the abdominal cavity, it has also been found in posterior mediastinum, neck, and in sacrococcygeal region. Commonest site is in the retroperitoneum (80%). Majority of cases occur in infancy, but can be detected at any age.

Case report 1

A 2-year-old boy had history of lump in abdomen since birth. Initially it was small, but gradually increased to attain the present size of 20 × 20 cm. The child had no other symptoms. The mass had variegated consistency, relatively fixed, and non tender. Radiograph showed calcified mass pushing the intestine. USG revealed a large, hyperechoic, and heterogeneous mass in the abdomen. CT suggested a mass in the retroperitoneum with calcified tissue and presence of vertebrae inside. So, a differential diagnosis of teratoma and fetus in fetu was considered. In exploratory laparotomy there was a mass in the retroperitoneum in close proximity with the intestine, pancreas, and stomach. It was almost 20 × 18 cm in size, partly cystic and partly bony. There was no feeding vessel to it. It had a covering around it, which accidently ruptured releasing serous fluid. In toto excision of mass was done. After opening the sac it had a limb-like structure along with nail with a poorly formed head. Also it was easy to make out the back and part suggestive of spine. Radiology of the

Fig. 1 Case 1—CT scan showing mass in the abdomen with three vertebrae

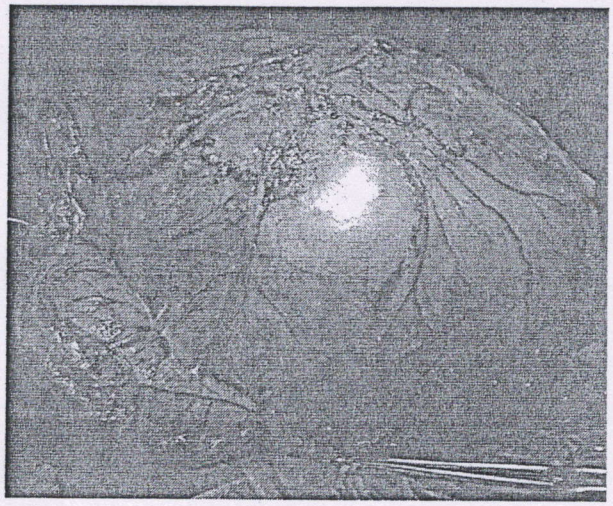

Fig. 2 Case 1—encapsulated mass in close relation with the intestine

mass confirmed the presence of bony limbs and two vertebrae. Histopathology also showed the presence GI organs inside the mass. The patient was discharged without complications and is being followed up since the last 11 months without any recurrence on ultrasonography (Figs. 1, 2, 3).

Case report 2

A 4-month-old female was diagnosed with a lump in the abdomen. She had no other complaints. Abdominal examination showed a mass in the right hypochondrium,

Fig. 3 Case 1—after removing the capsule, back and limbs with nail

soft and cystic in consistency. X-ray was suggestive of soft tissue shadow with no calcification. Ultrasonography of abdomen showed cystic mass arising from kidney. But CT scan of abdomen revealed that the mass was separate from the kidney and filled with cystic fluid without any calcification. So with diagnosis of teratoma and keeping in mind possibility of fetus, in fetu exploratory laparotomy was done. It was retroperitoneal mass in close relation with small bowel and right kidney. The mass was excised completely. There was no feeding vessel to it. Once outer cyst wall was opened there were fetal parts. We could identify fetal skull with limbs. Histopathology confirmed the presence of four vertebrae with three germ layers in the mass. The patient was discharged without any complications and is following with us for last 24 months without any recurrence (Figs. 4, 5, 6).

Discussion

Fetus in fetu is very rare condition. The overall incidence is 1 in 5,00,000 live births [1]. It is the malformed monozygotic diamniotic twin which is found inside the body of a living child or sometimes in an adult. It is thought to result from unequal division of totipotent inner cell mass of the developing blastocyst which results in small cell mass within a maturing sister embryo. The ultimate result is vestigial remnant of a diamniotic monochorionic twin that is located within the body of an otherwise normally developed twin [2]. Mostly, it is a single parasitic twin, but can range from 2 to 5. The organs present can be vertebral column, limbs, central nervous system, gastrointestinal tract, vessels, and genitourinary tract. Classically they are anencephalic. An unusual feature of our first case was the presence of limb with nail and the presence of well

developed intestine and the absence of other well-differentiated organs.

Presentation is usually before age of 18 months, but the oldest reported case is of 47-year-old adult. Thakral et al. [3] reported equal male and female predisposition but Patankar et al. and Federici et al. [4] noted a 2:1 male predominance. The common presentation is mass [3] most commonly in the abdomen, almost 80% in retroperitoneum [5] but sometimes can present as mass in skull [6–9] in 8%, sacrum [10–14] in 8%, scrotum [15] in 1%, mouth [16] in 1%, posterior mediastinum [17] in 1%, liver [18] one report. One of our cases had presentation at the age of 4 months and other at the age of 24 months though he had abdominal lump since birth. Symptoms of fetus in fetu relate mainly to its mass effect and include abdominal distension, feeding difficulty, pressure effects on renal system, and dyspnoea [4, 19, 20]. Preoperative diagnosis is now possible with advent of CT. Plain abdominal X-ray may be helpful in diagnosis, with half of them showing vertebral column and axial skeleton. Though rare anomaly fetus in fetu can be identified in preoperative period radiologically [9, 21–23]. Most commonly they are found in upper retroperitoneum; both of our cases had mass in the retroperitoneum.

Intra-abdominal fetus is usually contained in a complete sac, without any major vascular connections to the host. Predominant blood supply appears to be derived from the plexus where the fetus in fetu and the sac are attached to the abdominal wall [24], as in our case. Complete excision of the mass is curative. Complete excision of surrounding membrane should ensure definitive cure [19, 24–26]. Hopkins et al. [19] reported a malignant recurrence following resection of fetus in fetu. This was presumably caused by transformation of adherent membranes remaining at surgical sites. One should not try to deflate the capsule or membrane. If possible, the membrane should be removed intact. Cases of reported fetus in fetu weighed between 13 g [27] and 2,000 g [28].

Pathological controversy arises during differentiation of fetus in fetu from a mature or well organized teratoma. According to Willis (1935), presence of axial skeleton with vertebral axis with an appropriate arrangement of other limbs and organs, with respect to axis goes more in favor of fetus in fetu as it indicates abortive attempt after the stage of primitive streak formation [2]. In one of our cases, as described in literature, the vertebral column was detected by pathologist. So it was in accordance with Willis theory. It was radiolucent on radiography as it was less calcified [29]. Occasional cases have been reported where no spinal column was found on radiography [2]. However, review of literature showed that in about 9% of cases of fetus in fetu, there was no vertebral column, even on pathologic examination [30]. This has led to another definition proposed by Gonzalez-Crussi [30]:"Fetus in fetu is applied to any structure in which the fetal form is in a very high development of organogenesis and to the presence of vertebral axis" [30]. Our first case falls in the category of fetus in fetu by definition proposed by Gonzalez-Crussi. On the other hand, teratoma is an accumulation of pluripotential cells in which there is neither organogenesis nor vertebral segmentation [31]. Another important aspect of fetus in fetu is that they never become malignant whereas teratoma is known to become malignant [25]. Some supporters of teratoma theory have suggested that the fetus in fetu mass represents a well-differentiated, highly organized teratoma [24]. So it remains controversial whether fetus in fetu is a distinct entity or represents a highly organized teratoma. Du Plessis et al. [32] reported an interesting patient with both 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 teratoma

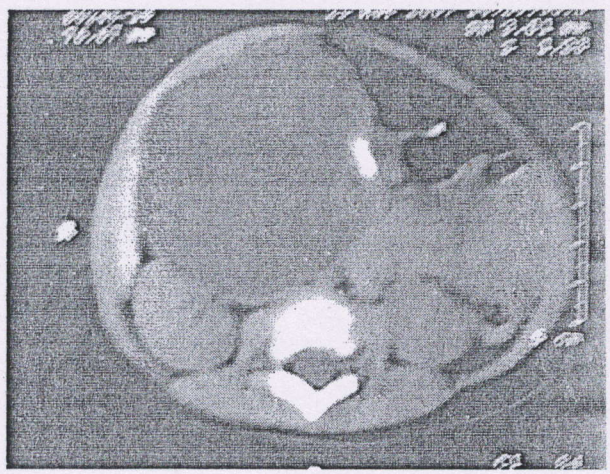

Fig. 4 Case 2—CT scan depicting mass in the abdomen without any calcification

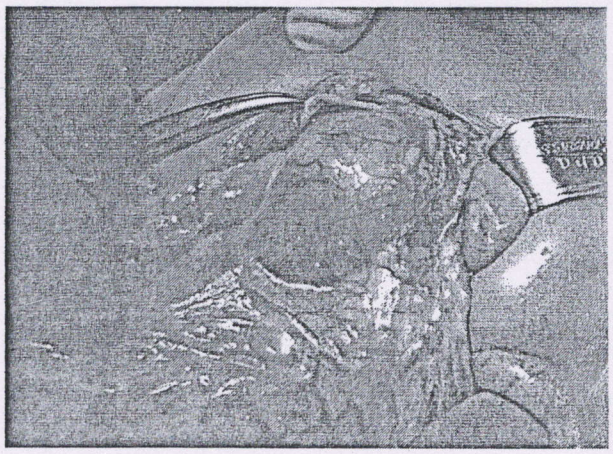

Fig. 5 Case 2—intraoperative showing capsulated mass in proximity with the intestine

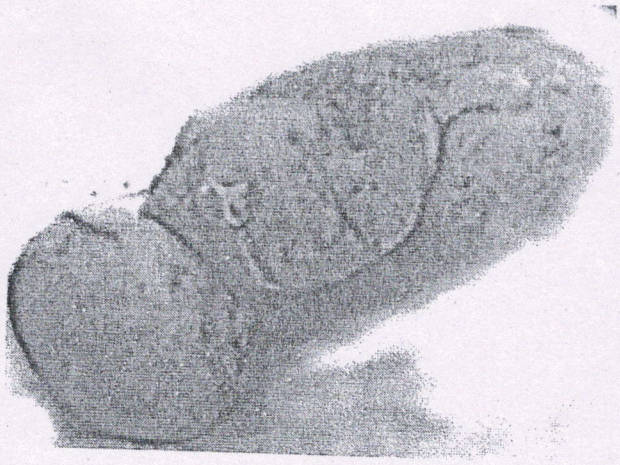

Fig. 6 Case 2—frank fetal specimen, histopathology showed presence of four vertebrae and three germ cell layers

arising from a third embryo which may have escaped the influence of its primary organizer". Occasional cases have been reported in literature where there are no vertebral bodies as it indicates dysplastic vertebral bodies which were not identified.

Conclusion

Fetus in fetu is a rare and interesting entity that typically presents as an abdominal mass in infancy or early childhood which can be diagnosed in the preoperative period. Complete excision of mass is curative and confirmatory.

References

1. Grant P, Pearn JH (1969) Foetus in Foetu. Med J Aust 1:1016–1020
2. Knox JS, Webb AJ (1975) The clinical Feature and treatment of fetus in fetu; two cases reports and review of literature. J Pediatr Surg 10:483–489
3. Thakral CL, Maji DC, Sajwani MJ (1998) Fetus in fetu: a case report and review of literature. J Pediatr Surg 33:1432–1434
4. Fedrici S, Cecarelli PL, Ferrari M, Galli G, Zanetti G, Domini R (1991) Fetus in fetu report of three cases and review of literature. Pediatric Surg Int 6:60–65
5. Tada S, Yasukochi H, Ohtaki C, Fukuta A, Takanashi R (1974) Fetus in fetu. Br J Radiol 47:146–148
6. Afshar F, King TT, Berry CL (1982) Intraventricular fetus in fetu case report. J Neurosurg 56:845–849
7. Goldstein I, Jakobi P, Groisman G, Itsokovitz-Eldor J (1996) Intracranial fetus in fetu. Am J Obstet Gyneco 175:1389–1390
8. Kimmel DL, Moyer EK, Peale AR, Wnborne LW, Gotwals JE (1950) Cerebral tumor containing five fetuses; case of fetus in fetu. Anat Rec 106:141–165
9. Tsai CH, Lin JS, Tsai FJ (1993) Intraventricular fetus in fetu: report of one case. Acta Paediatr Sin 34:143–150
10. de Lagauise P, de Napoli Cocci S, Stempfle N et al (1997) Highely differentiated teratoma and fetus in fetu: a single pathology? J of Paediatr Surg 32:115–116
11. Narsimharao KL, Mitra SK, Pathak IC (1984) Sacrococcygeal fetus in fetu. Indian Pediatr 21:820–822
12. Ouimet A, Russo P (1989) Fetus in fetu or not? J Pediatr Surg 24:926–927
13. Sada I, Shiratori H, Nakamura Y (1986) Antenatal diagnosis of fetus in fetu. Asia Oceania J Obstet Gynaecol 12:353–356
14. Sanal M, Kucukcelebi A, Abasiyanik F, Erdogan S, Kocabasoglu U (1997) Fetus in fetu and cystic rectal duplication in anewborn. Eur J of Pediatr Surg 7:120–121
15. Kakizoe T, Tahara M (1972) Fetus in fetu located in the scrotal sac of newborn infant: a case report. J Urol 107:506–508
16. Senuyz OF, Rizalar R, Celayir S, Oz F (1992) Fetus in fetu or giant epignathus protruding from the mouth. J Pediatr Surg 27:1493–1495
17. Katsuhiko Aoki, Yasunori Matsumoto, Minouri Hamazaki (2004) Fetus in fetu in mediastinum *Pediatric Radiology*. Vol 34, Number 12/December, pp 1017–1019
18. Magnus KG, Millar AJW, Sinclair-Smith C et al (1999) Intrahepatic fetus-in-fetu: a case report and review of the literature. J Pediatr Surg 34:1861–1864
19. Lewis RH (1960) Foetus in Foetu and Retroperitoneal eratoma. Arch Dis Child 36:220–226
20. Hoeffel CC, Nguyen KQ, Phan TT, Truong NH, Nguyen TS, Tran TT et al (2000) Fetus in fetu; a case report and review of literature. Pediatrics 105:1335–1344
21. Luzzatto C, Talenti E, Tregnaghi A, Fabris S, Scapinello A, Gugliemi M (1994) Double fetus in fetu: diagnostic imaging. Pediatr Radiol 24:602–603
22. Hanquinet S, Damry N, Heimann P, Deleat MH, Perlmutte N (1997) Association of a fetus in fetu and two teratoms: US and MRI. Pediatr Radiol 27:336–338
23. Khalifa NM, Maximous DW, ABD-Elsayed AA (2008) Fetus in fetu: a case report. J Med Case Reports 10(2):2
24. Heifetz SA, Alrabeeah A, St Brown B, Lau H (1988) Fetus in fetu: afetiform teratoma. Pediatr Pathol 8:215–226
25. Hopkins KL, Dickson PK, Ball TI, O'Shea PA, Abramowsky CR (1997) Fetus in fetu with malignant recurrence. J Pediatr Surg 32:1476–1479
26. Willis RA (1962) The borderland of embryology and pathology, 2nd edn. Butterworths, Washington DC, pp 442–446
27. Lee EY (1965) Foetus in Foetu. Arch Dis Child 40:689–693
28. Patankar T, Fatterpekar GM, Prasad S, Maniyar A, Mukherji SK (2000) Fetus in fetu: CT appearance—report of two cases. Radiology 214:735–737
29. Hing A, Corteville J, Foglia RP, Bliss DP Jr, Donis-Keller H, Dowton SB (1993) Fetus in fetu: molecular analysis of a fetiform mass. Am J Med Genet 47:333–341
30. Gonzalez-Crussi F (1982) Extragonadal teratomas. Atlas of tumor Pathology. 2nd ser, fasc. 18. Armed Forces Institute of Pathology, Washington DC
31. Kim OH, Shinn KS (1993) Postnatal growth of fetus in fetu. Pediatr Radiol 23:411–412
32. Du Plesis JPG, Winship WS, Kirsein JD (1974) Fetus in fetu and teratoma. S Afr Med J 48:2119–2122