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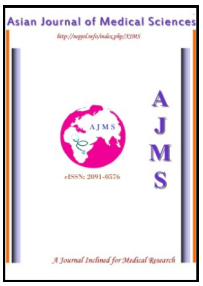

FETUS IN FETU (Pre-surgical dilemma: Review of literature and a case report)

O. P. Sharma[1*] and S. Senthil[1]

[1]Department of Radio diagnosis and Imaging, Institute of Medical Sciences, Banaras Hindu University, Varanasi 221 005; Uttar Pradesh (India).

Abstract

Fetus in fetu is an extremely rare developmental anomaly, due to disorganised embryogenesis in a diamniotic monochorionic pregnancy. Approximately 87 reports have been documented in the literature to date. We describe such an entity in two months old boy, who presented with abdominal distension and features of obstruction. Plain X ray abdomen and CT scan helped in correct pre operative diagnosis. The literature on this rare entity is reviewed, and the diagnosis and pathogenesis of the disease are discussed.

Key Words: Fetus in fetu; CT scan; Teratoma; Axial skeleton; Diamniotic monochorionic twins

1. Introduction

Fetus in fetu (FIF) was first described by Meckel in the late eighteenth century. It is an extremely rare condition, which occurs once in 500,000 deliveries. It occurs when a vertebrate fetus is enclosed within the abdomen of a normally developing fetus in a diamniotic monochorionic pregnancy.[1] Multiple theories regarding the embryogenesis of this condition have been proposed. Although abdominal masses are commonly encountered in pediatric surgery practice, the finding of a FIF causing abdominal distension is certainly unexpected.

2. Case History

Two and a half months old male infant born to 26 years healthy mother, from Chittupur village near Varanasi (Uttar Pradesh, India) presented to the paediatric out patient department with the complaints of progressive abdominal distension for two weeks, abdominal pain and vomiting for three days. The mother had uneventful antenatal period. The baby was born as third baby by full term normal vaginal delivery in hospital. He had normal developmental milestones and immunisation appropriate for age. There was no family history of twinning or any congenital anomalies. The haemogram and serum chemistry were normal.

X ray erect abdomen antero posterior and lateral views (Fig. 1) showed a soft tissue density shadow in left side of upper abdomen displacing bowel loops. A curved vertebral column like bony appearance to the left of the patient's vertebral column was noted.

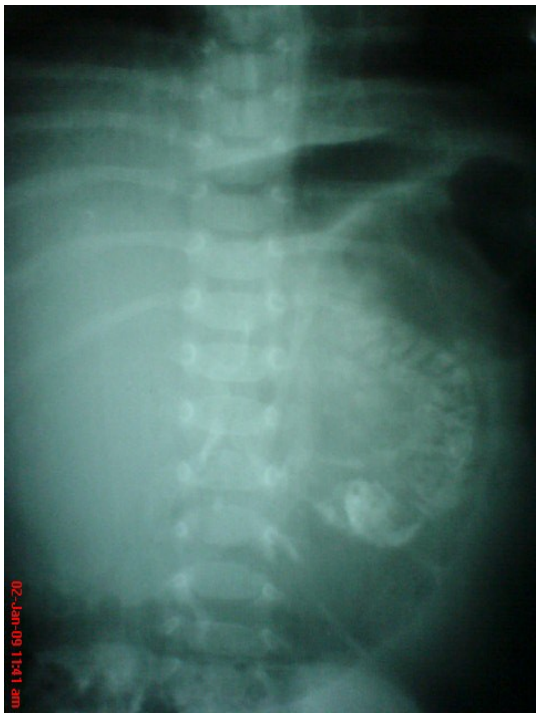

Fig.1 : Plain X ray abdomen AP view shows well formed vertebra of the fetu on the left side. Few facial bones, skull base and one femur are also seen.

***Correspondence:**

Prof. O. P. Sharma, Department of Radio diagnosis and Imaging, No. 8 / 180-R-124. Rajendra Vihar Colony, Newada, Varanasi – 221004. Uttar Pradesh (India). Contact No. 91-542-2369024 Fax.: 91-542-2369024 Email: sharmaopsonologist@indiatimes.com

Face and skull base like parts were also seen. A long bone was visualised. Ultrasonogram of the abdomen revealed complex cystic solid mass with multiple dense calcified parts predominantly occupying the retroperitoneum. Large cystic area was seen in the region of the skull. Few areas of fatty lobules showing increased echogenicity were also observed. It appeared like a retroperitoneal teratoma.

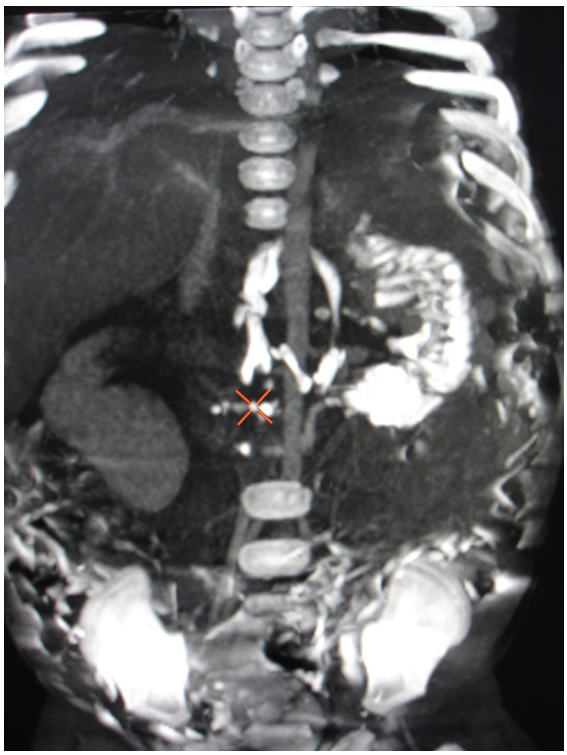

Fig. 2: Coronal MIP image from CT scan shows well developed fetus like structure with spine, neural tube, ribs and one femur. The vascular supply was from retroperitoneum.

CT scan was done to evaluate and further characterise the mass. CT showed a complex retroperitoneal mass lesion in the upper abdomen having fatty, solid and bony components. Heterogeneous enhancement and cystic areas were noted. Bony structures including complete axial vertebral skeleton, one femur, facial bones and skull base were found within the mass. Calvarial bones and brain tissue were not visualised. Three dimensional reconstruction and coronal MIP (maximum intensity projection) images (Fig-2) helped in exact anatomical reproduction of the lesion. Superior mesenteric artery was found to be the main arterial feeder supplying the mass. The left kidney was compressed. Diagnosis of fetus in fetu was made.

Emergency laparotomy was performed. Per operatively the CT scan findings were confirmed. Intra abdominal encapsulated complex mass was seen. The FIF was surrounded by amnion like fluid. Few pockets of straw coloured fluid were also found. Large heterogeneous disorganised soft tissue like mass was associated with the fetu. No definite umbilical cord structures were seen. It was adhered to the retroperitoneal structures, compressing over the left renal vessel origin. However, complete excision of the mass could be performed. Almost the whole length of the vertebral column could be identified. There were rudimentary eye ball and upper limbs having only soft tissue components. No brain parenchyma was seen.

It weighed about 600 g. Pathological evaluation showed 46 XY karyotyping of the fetus. Within the disorganised mass, histopathologically, well developed glomeruli and intestinal epithelium were found. A spinal cord with a central canal and adjacent vertebrae were observed. Fetu was acardiac. Large areas of fat were also seen. Well identifiable blood vessels, neural tissue and skeletal muscles were seen where the mass was attached with the retroperitoneum. No amnion, chorion, or villi were identified. There was no feature of malignancy at histology. The tumor markers of the patient α-FP, CEA, NSE and ß-HCG were found normal. The mass was therefore confirmed as a case of fetus-in-fetu.

Post operative period was uneventful. The patient is now on follow up. He is reviewed with ultrasonogram and serum tumor markers every three months.

3. Discussion

Fetus in fetu is detected most often in early infancy. This condition has a 2:1 male predominance.[1] A wide range of age of presentation varying from immediate new born to as old as 27 years has however been documented.[2] Despite its prevalence among infants and children, there has been reports of cases in which the anomaly had remained asymptomatic until later ages.[3] The most frequent symptoms reported are distension, palpable mass, emesis, poor feeding, jaundice, and/or dyspnea.[4]

Etiology: There have been several hypotheses on the origin of FIF such as a modified or defective process of twinning, differentiated teratoma and abnormal twin implantation prior to gastrulation with defective induction.[5] Recent reports favour defective twinning theory.

Pathogenesis: The entity defines a very rare abnormal embryogenesis of twinning where a malformed parasitic foetus is found in the body of its twin.[5] The pathogenesis

remains unknown but is thought to be because of a diamniotic, monochorionic, monozygotic twin that becomes included in its host during the process of ventral folding of the trilaminar embryonic disc. The inclusion in the sister embryo is speculated to be because of a persistent anastomosis of the vitelline circulation during development. The vitelline circulation develops into the superior mesenteric artery later in development, thereby explaining these lesions frequent appearance in the upper retroperitoneum.[4] Later, vascular accidents might have caused the heterogeneous and ill developed structures.

Criteria: To be called fetus in fetu the mass must demonstrate true organogenesis.[6] Some authors consider this entity to be an extreme form of highly organised and well differentiated teratoma, with matured organs. Willis suggested that a diagnosis of FIF be restricted to those cases where portions of an axial skeleton were present with an appropriate arrangement of organs relative to the skeletal axis. It was because of a fact of embryogenesis that the axial skeletal structures are formed during the primitive streak phase of development. For a mass to incorporate an axial skeleton, it would have to progress through the primitive streak stage of embryogenesis. In contrast, teratomas arise from the uncontrolled growth of pluripotent stem cells and develop without the overall organization imparted by embryogenesis. Therefore, the presence of an axial skeleton within a mass represents a FIF as opposed to a congenital teratoma.[4]

Karyotyping, serologic markers, and DNA restriction site mapping revealed that the FIF was actually a monochorionic, monozygotic twin of the host, thereby confirming a separate etiology for FIF as compared to teratoma.[4] Fetus-in-fetu occurs predominantly in the upper retroperitoneum. The fetus is usually single but multiple fetuses up to five have been reported.[6] Frequently vascular anastomoses with host vessels are identified. Absence of independent circulatory system could account for fetal growth retardation in all cases.[6] Our case showed feeder from superior mesenteric artery. Fetus-in-fetu does not exclusively present in the abdomen.[4] Recently a case of FIF with a rudimentary heart located in the sacrococcygeal region has also been reported.[2] Studies have shown that where intra-abdominal gonads were found, they corresponded histologically to the sex of the bearer. However, the occurrence of a dizygotic FIF is still a possibility.[5]

Pathology: On gross specimen, it is usually covered by a thin fibrous sac containing amber or straw coloured fluid along with an anencephalic fetus (100%) with limb buds (83%) and spine. The lower limbs are more developed than the upper limbs.[1] The identification of a round or tubular collection of very low density fat surrounding a central bony structure is characteristic of fetus in fetu.[1] Neural tissue and intestine were detected in half the cases. Skin, skin extensions, bone, bone marrow, fat tissue, venous structures, striated muscle and peripheral nerve sections are noted commonly.[3] Other uncommon organs reported are thyroid, parathyroid, pancreas, spleen, kidney, adrenal, testis, ovaries, urinary bladder, tongue, salivary glands, lymphnodes, trachea, lower respiratory tissue, teeth, digits and nails.[2,6,7] Histopathologically, the fetus consisting of fatty connective tissue with well identifiable blood vessels, neural tissue, salivary gland and skeletal muscles, gut with well developed mucosa, muscular coat and serosa has been found.[2]There has been reports of multiple FIF in the same mass.[1]

Investigations: Plain X-ray abdomen or the involved part will identify the axial skeleton and limb bones. The increasing use of ultrasound and CT has led to the identification of more cases. Obstetric ultrasound scan has made prenatal diagnosis of FIF possible in about 15% of reported cases.[5] CT scan will demonstrate the site, vascular supply and compressive effects of the FIF. Three dimensional reconstructions help in exact visualisation of the FIF in its anatomical position. Non-visualization of vertebral axis on radiography or CT scan does not exclude fetus in fetu as the pathologist can find it.[6] Despite the requirement of the presence of a vertebral column for diagnosis, there are reports of the cases without a vertebral column.[3]

Treatment: Treatment consists of complete resection as fetus-in-fetu may cause symptoms because of compression of adjacent intraabdominal organs.[4] Fetus in fetu is considered as a benign condition. In one case, the mass has been reported to recur as a yolk sac tumor after 4 months. This has been attributed to the presence of immature tissues in the small areas and the remnants of the capsule of the mass.[3] So complete excision has to be done always.

4. Conclusion

The requirement of an axial skeleton for the diagnosis of FIF should distinguish all cases of FIF from teratoma. But it is only present in 91% of cases.[1]Plain X ray abdomen and 3 dimensional CT image reconstruction helped us in arriving at the correct pre operative diagnosis. Complete removal is always advised, due to the remote possibility of malignant degeneration. Postoperative follow up of the patients with tumor markers and periodical ultrasound examinations is a necessary approach.

5. References

1. Iyer K.V, Vinaya K, J.O. Haller, Maximin S, Barrer-ras J, Velchek F. Multiple fetuses in fetu: imaging findings. Pediatr Radiol 2003; 33: 53-5. doi:10.1007/s00247-002-0736-1 PMid:12497241
2. Taori KB, Khurana SD, Dhomne SP, Rathi V. Fetus in fetu - a rare case. Indian J Radiol Imaging 2003;13:85-7.
3. Karaman I, Erdogan D, Ozalevli S, Karaman A, Cavusoglu YH, Aslan MK, Cakmak O. Fetus in fetu: A report of two cases. J Indian Assoc Pediatr Surg 2008;13:30-2. doi:10.4103/0971-9261.42572 PMid:20177485 PMCid:2810823
4. M. A. Escobar, J. E. Rossman, M. G. Caty. Fetus-in-fetu: report of a case and a review of the litera-ture. Journal of Pediatric Surgery 2008; 43:943-6. doi:10.1016/j.jpedsurg.2008.01.061 PMid:18485974
5. A. Rahman LO, A. Kadir AY, Rahman AG. Fetus -in -fetu in a 6-month-old. Afr J Paediatr Surg 2008;5:96 -8.doi:10.4103/0189-6725.44187 PMid:19858678
6. Phatak SV, Kolwadkar PK, Phatak MS. Fetus in Fetu: A case report. Indian J Radiol Imaging 2003;13:93-4.
7. Singh SN, Pratap A, Sinha AK, Kumar A, Lakshmi R, Shakya VC, Agrawal A. Giant retroperitoneal fetus-in-fetu: An unusual cause of respiratory distress. J Indian Assoc Pediatr Surg 2007;12:158-60. doi:10.4103/0971-9261.34961