



Case Report

FETUS IN FETU : A CASE REPORT & REVIEW OF LITERATURES

MAM RAHMAN¹, AA FAROOQ², MAH BHUIYAN³, TK CHOWDHURY⁴, HMA ROUF⁵

Abstract

Fetus-in-fetu is a rare abnormality secondary to the abnormal embryogenesis in a diamniotic, monochorionic pregnancy. It is a rare pathological condition and fewer than 200 cases have been reported in the literature. We are reporting a case in which a 15 year old girl presented with a painful lump in left upper abdomen. Preoperative imaging, exploration and macroscopic examination of the excised specimen revealed it a case of fetus-in-fetu. This case is unique in terms of age of presentation and mature fetus like external appearance.

Introduction

Fetus-in-fetu is a rare congenital condition in which a vertebrate fetus is incorporated within its twin. It was first described by Meckel in 1800¹⁻⁴ and defined by Willis in 1935 as a mass containing a vertebral axis often associated with other organs or limbs around this central axis²⁻⁷. First reported case cited in 1809 by Young⁸. This condition featuring a monozygotic, diamniotic, parasitic twin, attached by a vascular anastomosis to its host chorionic circulation. This abnormality of monozygotic diamniotic twinning results from an unequal division

of totipotent inner cell mass of the developing blastocyst as described by Lewis⁹. There is still controversy about whether fetus in fetu is a more highly differentiated teratoma^{3,4,6,7,10} or an asymmetric monozygotic diamniotic endoparasitic twin^{4,5,10-13} and dates back to 1800 AD- Meckel described fetofornity of the anomaly¹⁻⁴. Differentiation between two is possible by application of accepted criteria by Willis in 1935²⁻⁷ and Lord in 1954^{4,13,14}. Presence of whole or part of vertebral column plus appropriately situated other axial and or, appendicular bones and or, organs constitutes hallmark of fetus in fetu- well demonstrated either by imaging, macroscopic appearance or, microscopic examination of the mass.

Case report

A 15 years old girl, from a rural area admitted in Surgery department, Chittagong Medical college and Hospital, Chittagong, Bangladesh with the problems of - pain in the left upper abdomen for 5 days, lump in the same region for 5 days, vomiting for several times during the last 4 days, low grade fever & dry cough for 5 days. It is hereby noted that patient was totally unaware of this lump even 5 days before! On examination, patient was ill looking, average built, not anaemic, non-icteric & not dehydrated. Her pulse was 78/min, BP 110/80 mm hg and temperature was normal.

Abdominal examination revealed a swelling in left upper abdomen without any visible pulsation or peristalsis. On palpation there was a large non tender intra abdominal lump in left hypochondriac and left lumbar region measuring approximately 22cm X 18 cm with an ill defined margin. It was of variable consistency, bimanually palpable, had irregular surface & slightly moved with respiration. Insinuation

1. Dr. MA Mushfiqur Rahman, FCPS, MS, Assistant Professor, Department of Pediatric Surgery, Chittagong Medical College & Hospital.
2. Dr. Md. Abdullah Al Farooq, FCPS, MS, Assistant professor, Department of Pediatric Surgery, Chittagong Medical college & Hospital
3. Dr. Md. Akbar Husain Bhuiyan, MS, Assistant professor, Department of Pediatric Surgery, Chittagong Medical college & Hospital
4. Dr. Tanvir Kabir Chowdhury, MBBS, MS final part student (Pediatric Surgery), Chittagong Medical College & Hospital, Under Chittagong University.
5. Prof. Hasan Md. Abdur Rouf, FCPS, Ex Head, Department of Surgery, Chittagong Medical College & Hospital

Correspondence to : Dr. M A Mushfiqur Rahman, FCPS, MS, Assistant Professor, Department of Pediatric Surgery, Ward-11B, Chittagong Medical College & Hospital, Chittagong 4000, Bangladesh, E- mail: Piku71@yahoo.com

was possible above the lump underneath the costal cartilage and its upper margin could not be reached. There was no ascites or organomegaly. Bowel sound was present. Other systems appeared to be normal.

Several laboratory investigations and imaging were performed during her stay in the hospital.

Plain abdominal radiograph (Fig. 1) showed—large calcified mass in left hypochondrium and left lumbar region, spine & appendicular bones noted .



Fig.-1: Plain X-ray abdomen. Large calcified mass in left hypochondriac and lumbar region. (arrow-long bone, asterix-base of skull, arrowhead-spine of fetus)

Abdominal Ultrasound revealed – left upper abdominal heterogenous mass, mild left sided pelvicalyceal dilatation, bowel loops are overloaded with hard fecal matter particularly on left side, left kidney displaced down.

CXR P/A – normal

CT scan of abdomen (Fig. 2) showed- left sided abdominal mass & comment was mature cystic dermoid.

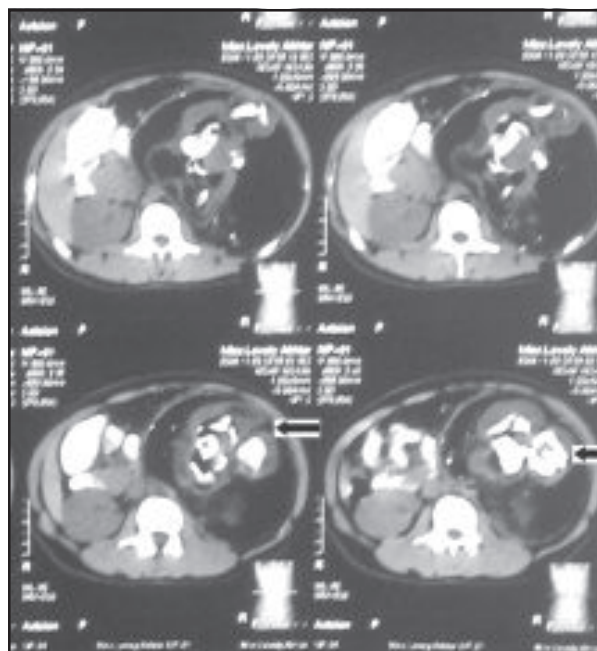


Fig.-2: Abdominal CT. Left sided abdominal mass-mature cystic dermoid (arrow) showing (?) spine, long bones and base of the skull.

Intravenous Urogram (Fig. 3) showed - Left upper quadrant mass & radiologist suggested it was a parthenogenetic homunculus fetiform teratoma. Other



Fig.-3: IVU. Suggestive of fetiform teratoma with caudally displaced left kidney (arrow).

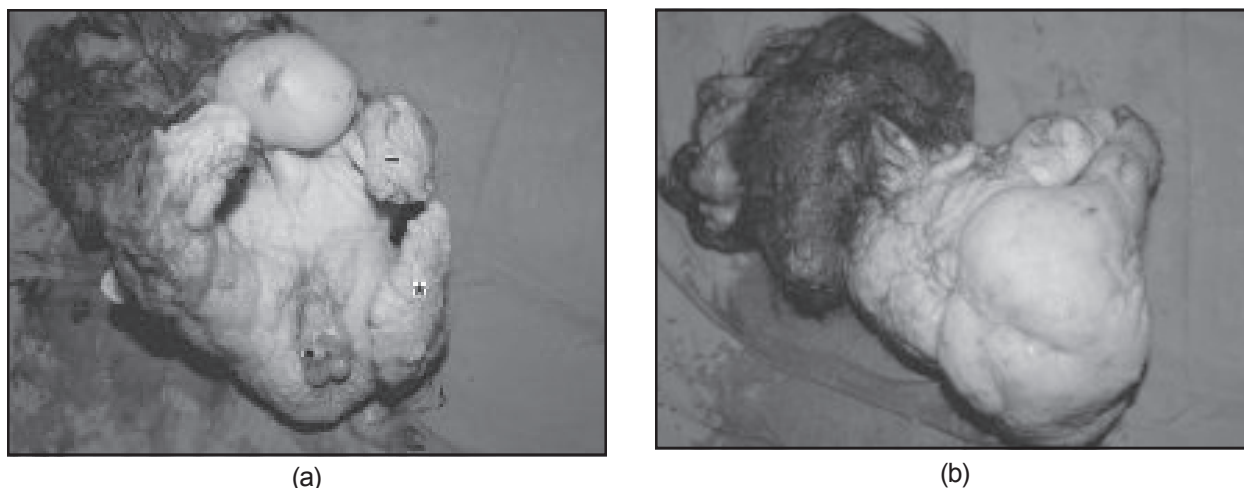


Fig-4: A and 4B . Well developed fetiform mass with visible four Limbs (asterix & arrow), exposed developing genitalia(arrowhead), anencephalic with tufts of hair, developed buttock and back.

less likely possibilities were i) fetus in fetu ii) intraabdominal pregnancy. iii) caudally displaced left kidney but urographic features appeared normal.

Blood culture showed no growth after 72 hrs of incubation. S amylase was 150.0 u/l

Pregnancy test was negative.

Other hematological tests, Urinalysis, Renal function test, Liver function tests were found normal.

After clinical, laboratory evaluation and imaging the mass was thought to represent either a fetus in fetu or a dermoid cyst.

Operative treatment was planned and a large retroperitoneal, well capsulated mass was excised that was connected to the host by a vascular pedicle close to the left crus of diaphragm, the left kidney was found to be pushed down.

On gross eye appearance, the mass measured 26×20 cm, capsule was reasonably thick containing huge amount of sebaceous material and a well developed fetiform mass with grossly visible four limbs, well developed buttock & back, developing genitalia, anencephalic with tufts of hair attached at back of the neck and fine hair over whole of the skin surface (Fig. 4A and 4B). Weight of the fetus was 1700 grams.

Discussion

Fetus in fetu was first described by Meckel in 1800 and defined by Willis in 1935 as a mass containing a vertebral axis often associated with other organs or limbs around this axis¹⁻⁷. This abnormality occurs in

1 in 500,000 live births¹⁵. Male seems to be affected more than female¹. Most of the fetus in fetu masses are recorded as being located in the

upper abdominal retroperitoneum^{1-4,6,7,11,13}. Other sites are Liver, Lesser sac, Kidney, Adrenal gland, Scrotum, Pelvis, Mesentery, Sacrococcygeal region, Cranium^{2,4,6,13}.

According to Hoeffel et al¹⁶ who have reviewed 87 case reports, in 80 percent of fetus in fetu were localized in the retroperitoneal areas but could also be found in atypical locations such as skull, scrotum, mouth and adrenal gland. Most fetus in fetu are detected in the first year as asymptomatic slow growing abdominal mass^{1,3,4,11,12}. Delayed presentation had been documented also^{4,13}. Only in 16.7% of these cases it was possible to show a preoperative diagnosis of fetus in fetu, the differential diagnosis being teratoma and meconium pseudocyst¹⁷.

Teratomas are defined as tumours containing different tissues from one or more germ cell layers with origin in pluripotent cells without systematic organization and with potential to develop into mature or malignant tissue. According to Willis the distinction between fetus in fetu and teratoma is classically based on the absence of an axial skeleton but recently some investigators pointed to the possibilities of fetus in fetu being a form of a highly differentiated teratoma, the differential diagnosis can be difficult because of similar clinical and radiological features and presence of histological examination of complex, well differentiated tissues looking as organs^{18,19}. The

diagnosis of fetus in fetu is based on confirmation of a spinal column along with the presence of complex and well differentiated tissues²⁰.

Conclusion

Histologically fetus in fetu has no malignant component but teratomas are potentially malignant^{1-4,13}. Few authors claim fetus in fetu may be malignant^{3,6} and chance of recurrence if immature components are found in histopathology³. Treatment is surgical excision^{1,3,6,13}.

References

1. Luzzatto C, Talenti E, Tregnaghi A, et al: Double fetus in fetu: Diagnostic imaging. *Pediatr Radio* 1994; 24: 602-03.
2. Al-Baghdadi R: Fetus in fetu in the liver: Case report and review of the literature. *J Pediatr Surg* 1992; 27: 1491-92.
3. Hopkins KL, Dickson PK, Ball TI, et al: Fetus in fetu with malignant recurrence. *J Pediatr Surg* 1997; 32: 1476-79.
4. Eng H-L, Chuang J-H, Lee T-Y, et al: Fetus in fetu: A case report and review of the literature. *J Pediatr Surg* 1989; 24: 296-99.
5. Willis RA: The structure of teratoma. *J Pathol Bacteriol* 1935; 40: 1-36.
6. Hanquinet S, Danny N, Heimann P, et al: Association of a fetus in fetu and two teratomas: US and MRI. *Pediatr Radio* 1997; 127: 336-38.
7. du Plessis JPG, Winship WS, Kirstein JDL: Fetus in fetu and teratoma. *SAMed J* 1974; 48: 2119-22.
8. Young GW. Case of a foetus founding the abdomen of a boy. *Med Chir Trans* 1809; 1: 234-62.
9. Lewis RH. Foetus in foetu and the retroperitoneal teratoma. *Arch Dis Child* 1961; 36: 220-6.
10. Heifetz SA, Alrabeeah A, Brown BS, et al: Fetus in fetu: A fetiform teratoma. *Pediatr Pathol* 1988; 8: 215-26.
11. Kim OH, Shinn KS: Postnatal growth of fetus in fetu. *Pediatr Radiol* 1993; 23: 411-12.
12. Cywes S, Millar AJW, Rode H, et al: Conjoined twins-The Cape Town experience. *Pediatr Surg Int* 1997; 12: 234-48.
13. Fink AM, Cuckow PM, Scott R: Case report: Fetus in fetu-Imaging, surgical and pathological findings. *Clin Rad* 1995; 50: 274-75.
14. Lord JM: Intra-abdominal foetus in fetu. *J Pathol Bacteriol*, 726
15. Thakral CL, Maji DC, Sajwani MJ. Fetus-in-fetu: a case report and review of the literature. *J Pediatr Surg* 1998;33:1432-34.
16. Hoeffel CC, Nuyen KO, Tran TS, et al:Fetus in Fetu: A case report and literature review. *Pediatrics* 2000; 105: 1335-44.
17. Chen CP,Chem SR, Lin FF: Prenatal, diagnosis, pathology and genetic study of fetus in fetu. *Prenat Diagn* 1997; 17: 13-21.
18. Griscom NT:The roentgenology of neonatal abdominal masses.*AJR* 1965; 93: 447-463.
19. Gross RE, Clatworthy HW:Twin fetuses in fetu. *J Pediatr* 1951; 38: 502-08.
20. Bassetto MA, Franceschi T, Lenotti M, et al:AFP and HCG in germ cell tumours. *Int J Biolog Markers* 1994; 9: 29-32.