Fetus -in -fetu in a 6-month-old

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ABSTRACT

Fetus-in-fetu is a malformed parasitic monozygotic diamniotic twin found inside the body of the living child or adult. We report a case of lumbar mass having superficial rudimentary phallus, labioscrotal fold, testes, pedunculated thumb-like digit and rudimentary pelvis in addition to bowel loops in a 6-month-old Nigerian girl. The mass was excised and the baby did well. We propose based on these that dizygotic parasitic foetiform twin could exist.

Key words: Dizygotic parasitic twin, differentiated teratoma, foetus in foetu

INTRODUCTION

The term fetus-in-fetu (FIF), coined by Meckel in the late 18th century, defines a very rare abnormal embryogenesis of twinning where a malformed parasitic foetus is found in the body of its twin.[1,2] Over 80% of the reported cases are retroperitoneal in location.[1,2] Cases have been reported in nearly all body parts except in the extremities.[1] We present a lumbar foetus-in-foetu with retroperitoneal extension having external rudimentary phallus and testis as well as a pedunculated well-developed thumb-like digit with nail in a 6-month-old girl.

CASE REPORT

A 6-month-old girl was brought to the hospital because of gradually increasing swelling at the lower aspect of the back since birth. She was a product of full term pregnancy delivered at home unsupervised of an unregistered, 28-year-old para7^{+0} nomadic cattle rearer. There was no history suggestive of trauma, fever, discharge or pain from the mass. She had no other deformity. There was no family history of twinning or similar anomaly.

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She had good developmental milestones and incomplete immunisation for age. The haemogram and serum chemistry were normal.

Examination revealed a healthy looking child with appropriate growth for age (7.2 kg). There was a huge left posterolateral lumbar mass, 20cm x 20cm, firm, and non-tender with 3- projections on its surface [Figure 1]:

1. Phallus, 4cm x 2cm with surrounding hyperpigmented and rugosed labioscrotal folds.
2. Pedunculated limb-like projection with dimple at

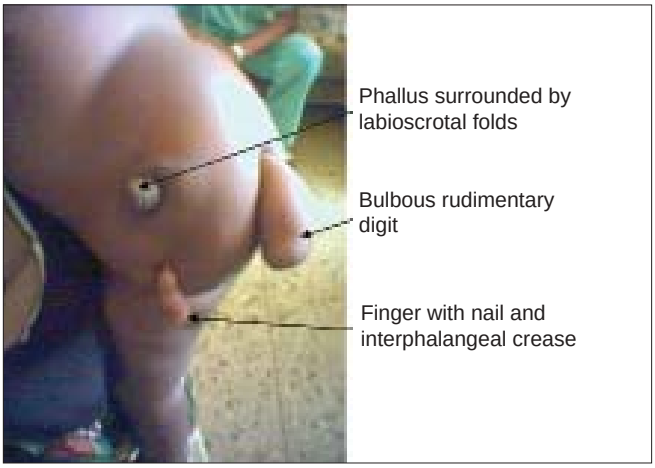


Figure 1a: Lateral photograph of the baby with FIF showing Phallus surrounded by labioscrotal folds

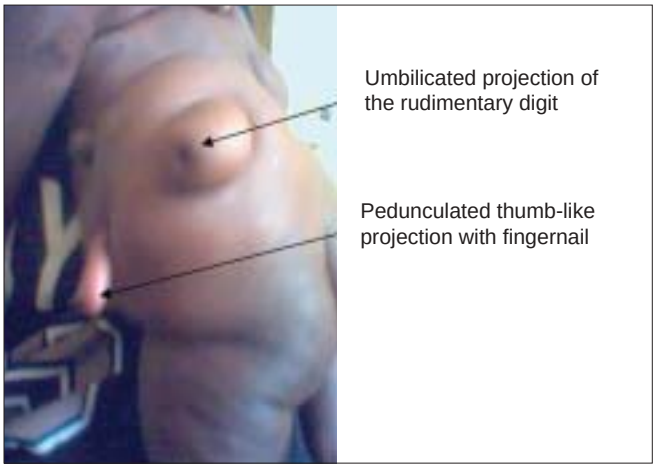


Figure 1b: Oblique photograph of the baby with FIF showing umbilicated projection of the rudimentary digit and pedunculated thumb-like projection with fingernail.

the apex (umbilication).
3. Pedunculated thumb-like lesion, firm to hard with keratinised ventral surface, interphalangeal rings and a nail

There was also a palpable deep bony hard structure fixed to the left iliac crest. There was no other palpable intra-abdominal mass or organomegaly.

X-rays revealed a left posterolateral mass of soft tissue density over the lumbar region with three pedunculated projections and two central linear calcific dense opacities appearing like a rudimentary pelvis over the iliac crest [Figures 2a, 2b]. There was associated extension of bowel loop into the mass. There was no vertebral anomaly or bony fusion between the mass and the vertebral column. The overall impression was of a parasitic twin. On excision the mass, which measured 20cm x 20cm, contained fatty deeper tissue and a central bony ring attached to the iliac crest. There was a rent in the lumbar hiatus through which the descending colon and small intestine herniated beneath the mass. These bowel loops were separated or lined by a sac. A cord of tissue anchored to an ovoid structure confirmed by histology to be gonadal tissue, and a vertebral column was also identified. The postoperative period was uneventful but the baby was lost to follow up six months later.

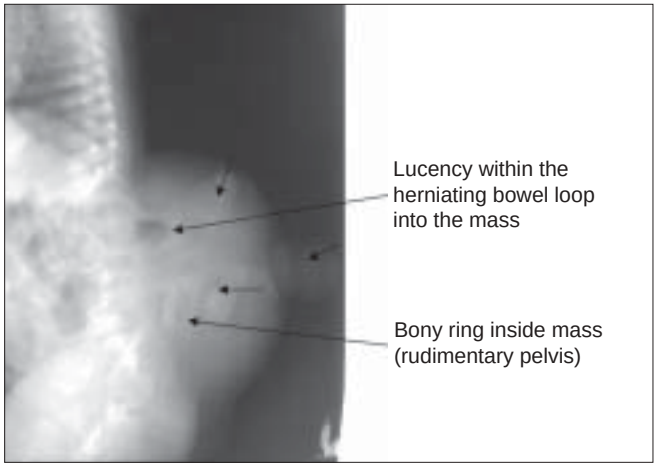


Figure 2a: Lateral radiograph of the baby with FIF showing lucency within the herniating bowel loop into the mass and Bony ring inside mass

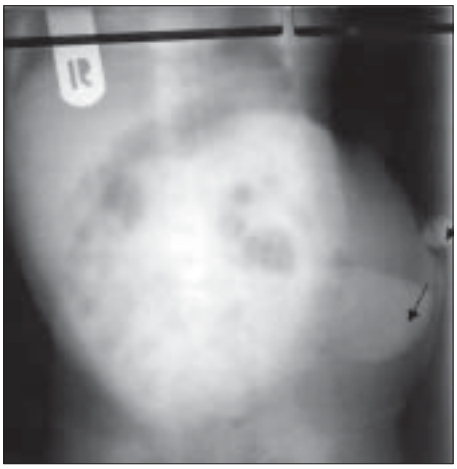


Figure 2b: Radiograph show left posterolateral mass over the lumbar area. Note the projections on its surface

DISCUSSION

Foetus-in-feotu occurs in about 1 in 500,000 deliveries with a male to female ratio of 2:1.[1-4] To date, less than 200 cases have been reported in the English literature out of which less than five cases emanated from Africa.[1] However, it remains uncertain whether this is due to under reporting.

The ages and clinical features at presentation vary depending on the site and mass effect of the homunculus.[1,2] As in the case presented, a majority of the cases present in infancy. However, in the Hoeffel *et al*.[2] review, four out of 88 (4.5%) cases are described in patients 10 years of age or older. This is unlike 12 out of 160 (7.5%) reported by Rahman *et al*.[1] The oldest case of FIF reported in both reviews was found in a 47-year-old man.[1,2,5]

Occasionally, the anomaly is asymptomatic but may present with symptoms of mass-effect related to its location such as jaundice, feeding difficulty, vomiting and dyspnoea.[1,2] Though presentation was delayed in our patient, grotesque disfigurement [Figure 1a, b] and continuous growth of the mass forced the parents to seek medical care. The retroperitoneal extension in this case conforms to the commonest location of most FIF.[1,2]

There have been several hypotheses on the origin of FIF such as a modified/defective process of twinning, differentiated teratoma and abnormal twin implantation prior to gastrulation with defective induction.[1,2,6]

The presence of external rudimentary appendages, one with umbilication and the other with fingernail in our case support the foetal inclusion theory of FIF. Studies have shown that where intra-abdominal gonads were found, they corresponded histologically to the sex of the bearer,[2] thus indicating that the foetus-in-foetu is derived from the same zygote tissue as its host. However, this is not so in our case that has an external phallus with labio-scrotal fold containing testis in a

female autosite. This rare combination of features was not found in any of the literature searched. Therefore, this case is at variance with the foetal inclusion theory of diamniotic monochorionic monozygotic twinning hypothesis of FIF. Hence, we propose based on these findings that dizygotic parasitic foetiform twin could exist and when it does occur, it may probably be superficially or externally located. However, further studies will be necessary for corroboration. Another possibility in this case is that of highly differentiated teratoma type of FIF.[7] Genetic analysis may therefore be an essential part of evaluation of foetus in foetu, which is presently out of reach in our locality and was not done.

Nearly all body organs or parts have been documented in various degrees in FIF but the identification of a vertebral column has been singled out to secure the diagnosis of FIF and its differentiation from teratoma.[1-5] The presence of vertebral column indicates that foetal development of the included twin must have advanced at least to the primitive streak stage to develop a notochord, which is the precursor of the vertebral column.[4,5] In some instances, the spinal column may not be identifiable at imaging but at histopathological examination as in our case.

The increasing use of ultrasound and computed tomography (CT) have led to the identification of more cases.[1-4] Obstetric ultrasound scan has made prenatal diagnosis of FIF possible in about 15% of reported cases.[1-4] Prenatal diagnosis was not possible in this case because the patient's mother was unregistered. Ultrasound scan of the mass was also not of extra value to the plain radiograph in our diagnosis of FIF. However, a CT scan that would have assisted in delineating the extent of this mass, and an MRI that would have supplied further information regarding tissue characterisation and vascularisation of the mass lesion were unavailable at our centre, besides their high cost. Prenatal MRI scans have been used to confirm prenatal diagnosis of an intra-abdominal FIF to aid obstetricians, neonatologists, and paediatric general surgeons in perinatal and surgical management.[1-4]

Though there was no feature of malignancy at histology in this case, we are aware that follow up is very important after the definitive surgery because malignant recurrence has been reported following resection of a case of FIF.[8]

The origin of FIF remains uncertain. The occurrence of a dizygotic FIF is still an enigma but possible.

REFERENCES

1. Rahman GA, Abdulkadir AY, Abdur-rahman LO. Fetus-in-fetu: A review article. Eur J Sci Res 2007;18:663-73.
2. Hoeffel CC, Nguyen KQ, Phan HT, Truong NH, Nguyen TS, Tran TT, *et al*. Fetus in fetu: A case report and literature review. Pediatrics 2000;105:1335-44.
3. Chua JH, Chui CH, Sai Prasad TR, Jabcobsen AS, Meenakshi A, Hwang WS. Fetus-in-fetu in the pelvis: Report of a case and literature review. Ann Acad Med Singapore 2005;34:646-9.
4. Brand A, Alves MC, Saraiva C, Loío P, Goulão J, Malta J, *et al*. Fetus in fetu-diagnostic criteria and differential diagnosis: A case report and literature review. J Pediatr Surg 2004;39:616-8.
5. 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.
6. Beaudoin S, Gouizi G, Mezzine S, Wann AR, Barbet P. Mediastinal fetus in fetu: Case report and embryological discussion. Fetal Diagn Ther 2004;19:453-5.
7. 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.
8. 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.

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