CASE REPORT

Foetus in Foetu: A Rare Prenatal Diagnosis of a Foetal Abdominal Mass

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ABSTRACT

A pregnant woman was referred to our perinatal tertiary care centre because of foetal bowel distension. Ultrasonography revealed a well-circumscribed circular mass in the foetus' abdomen. After delivery, the male neonate underwent laparotomy to excise the mass. Pathologic examination of the mass was consistent with the diagnosis of foetus in foetu. This case of foetus in foetu was differentiated from a teratoma based on the presence of a vertebral axis and appropriate arrangement of other organs and limbs relative to the vertebral axis. A multidisciplinary clinical approach may be more suitable for managing rare foetal disorders, such as this one.

Keywords: Abnormal twinning, enclosed twin, foetus in foetu, intra-abdominal foetal mass.

INTRODUCTION

Most neonatal abdominal masses are caused by benign retroperitoneal lesions such as hydronephrosis and multicystic dysplastic kidney (1). Foetus in foetu (FiF), also known as an enclosed twin, is a rare condition. It is described as a twin that 'fails to thrive' and is confined in its sibling. It is estimated to affect 1 in 500 000 live births (2). Isaacs reported that FiF was responsible for 25/534 (4.68%) of perinatal germ cell tumours (3). The first case was reported by Young in 1809 (4). Foetus in foetu is distinguished from a teratoma by the presence of a vertebral axis and, commonly, by the appropriate arrangement of other organs and limbs relative to the vertebral axis (5). In 80% of cases, the FiF was found in the retroperitoneum, while other, atypical sites include: the intracranial area (6, 7), mediastinum, lung, oropharynx, scrotum, adrenal gland, ovary, or the neck (8). Because of its malignant potential, the FiF should be completely excised by surgery. The prognosis after resection is usually benign (9).

CASE REPORT

A 31-year-old multiparous pregnant woman was referred to our perinatal tertiary care centre after a routine ultrasound scan showed that her 37-week-old foetus had bowel distension. The woman was in a non-consanguineous marriage and had no significant medical or gestational history. There was no history of twin gestation. A detailed sonogram and a follow-up ultrasound of the index pregnancy revealed no notable clinical signs.

A senior obstetrician examined the patient via ultrasound and noted that the foetal biometry was consistent with a gestational age of 34–35 weeks, there was severe oligohydramnios. There was a well-circumscribed circular mass measuring 44 × 38 × 40 mm with echoes consistent with calcification. The mass was located in the central upper abdomen, below the liver, in front of and between two foetal renal pelvises (Fig. 1A).

The male neonate was delivered via Caesarean section at 37 weeks and 1 day of gestation, because of foetal distress. The birth weight was 2740 g. The Apgar scores were 7 and 9 at 1 and 5 minutes, respectively. The mother was discharged 2 days after delivery, with no remarkable medical concerns at this time.

A physical examination of the neonate revealed that the only notable sign was mild to moderate abdominal distension. The neonate's complete blood count and arterial blood gas were normal. Ultrasound confirmed

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that the locations and structures of the liver, gall bladder, spleen and suprarenal glands were normal. The mass in the neonate's abdomen had the same features as those observed in the prenatal examination. Respiration and alimentation were noted as being normal. An experienced paediatric cardiologist reported that the neonate's cardiac system was normal. A computed tomography scan confirmed the ultrasound and physical examination findings (Fig. 1B).

The paediatric surgical team decided to perform a laparotomy to excise the mass. In open view during surgery, the mass compressed the duodenum from the posterior side (Fig. 1C). After opening the retroperitoneal space, it was possible to see the haemorrhagic structure of the mass (Fig. 1D). Excision of the mass eliminated the compression on the duodenum (Fig. 1E).

The mass was removed intact and sent for pathological and genetic examination. The mass weighed 220 g and measured 40 × 30 × 25 mm. A radiograph of the

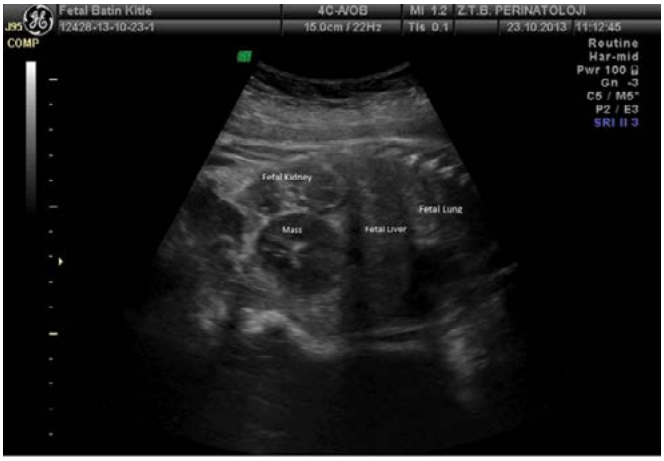

Fig. 1A. Coronal plane ultrasound view of the mass which located in the foetal abdomen.

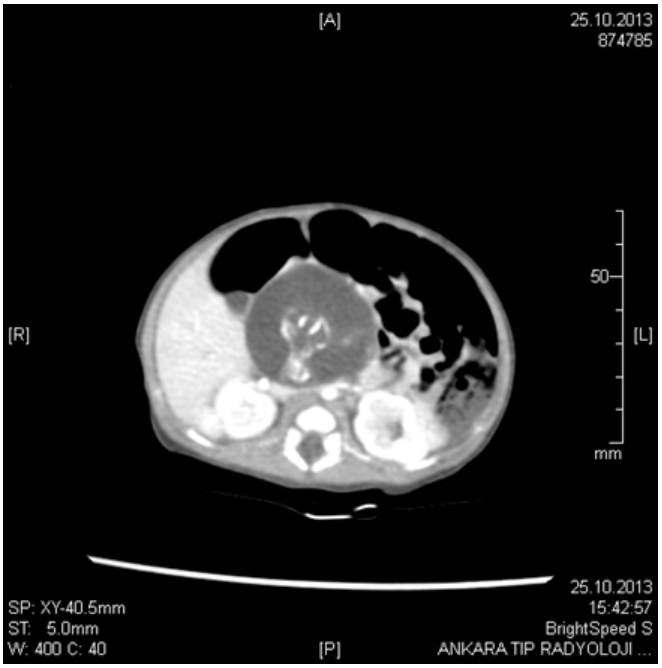

Fig. 1B. Computed tomographic view of the neonate shows the mass in the central abdomen. This view also shows the kidneys, liver and mildly dilated bowel of the neonate.

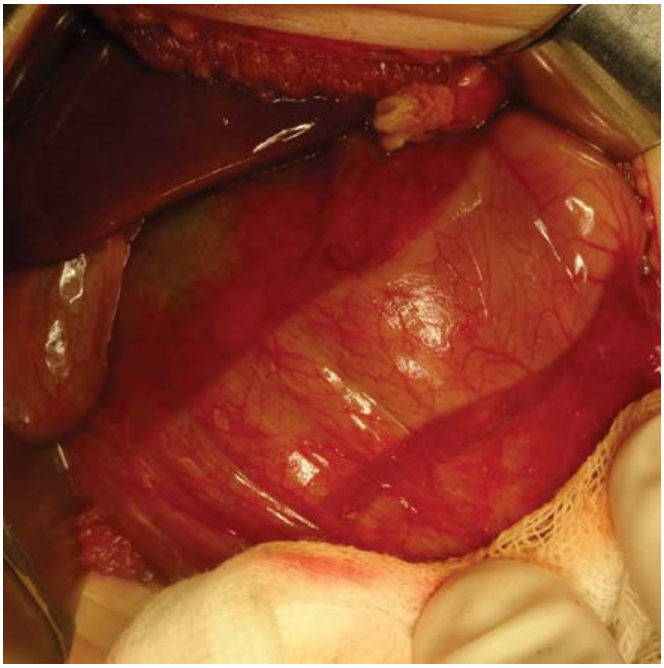

Fig. 1C. An open view of the neonate's liver, gall bladder and duodenum. The mass compresses the duodenum from the posterior side.

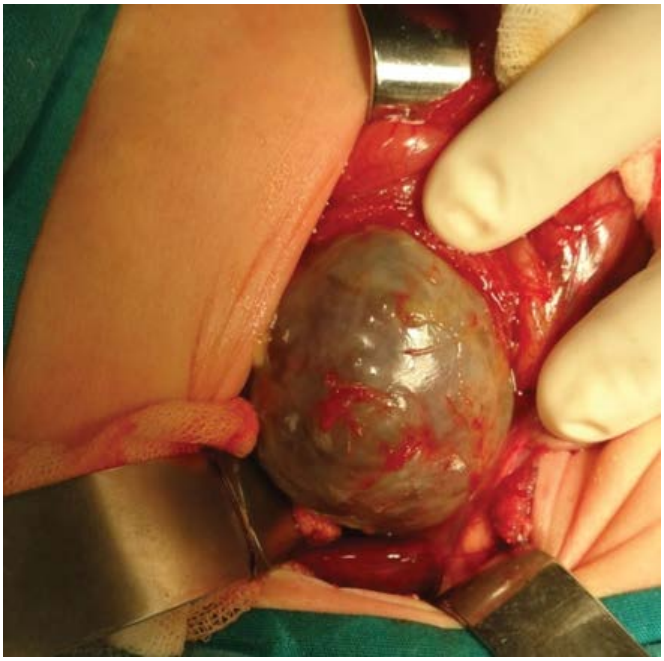

Fig. 1D. Image of the mass in the retroperitoneal space showing its congestive and haemorrhagic structure.

mass showed a linear calcification and the presence of a vertebral column and limbs (Fig. 1F). Pathologic and macroscopic examination of the mass revealed its haemorrhagic structure, including the presence of fatty tissue, irregularly arranged bone, cartilage, and a skin-covered conglomeration of recognisable rudimentary foetal parts (Fig. 1G). In addition, a rudimentary axial skeleton was found with three rudimentary limbs; the largest limb was 15 mm long and the smallest limb was 10 mm long. Microscopic examination confirmed that the mass was covered with skin and contained some osseous tissue and a rudimentary gastrointestinal tract. The final pathologic report stated that the mass was consistent with an acephalic acardiac foetus. Cytogenetic examination of the mass revealed that the karyotyping was 46 XY.

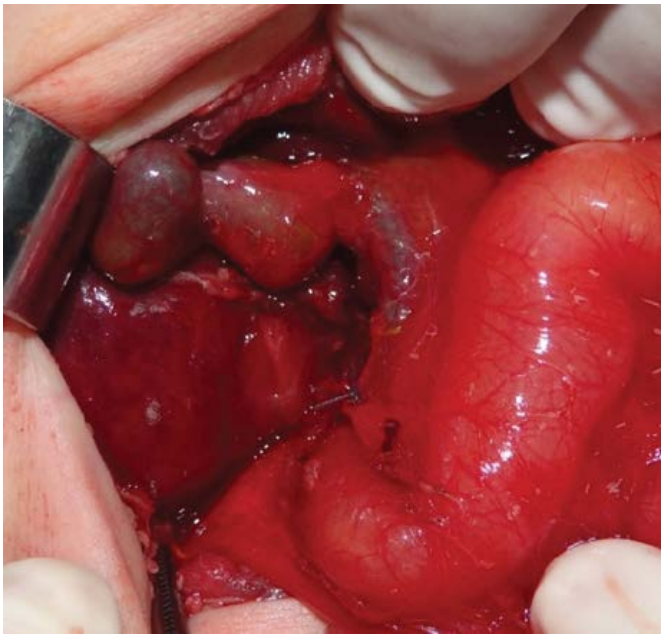

Fig. 1E. Excision of the mass eliminated the compression on the duodenum.

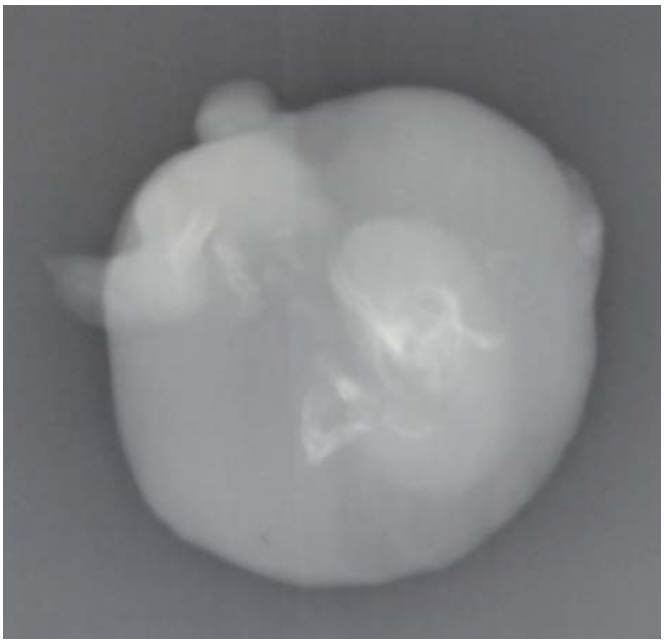

Fig. 1F. X-ray image of the mass.

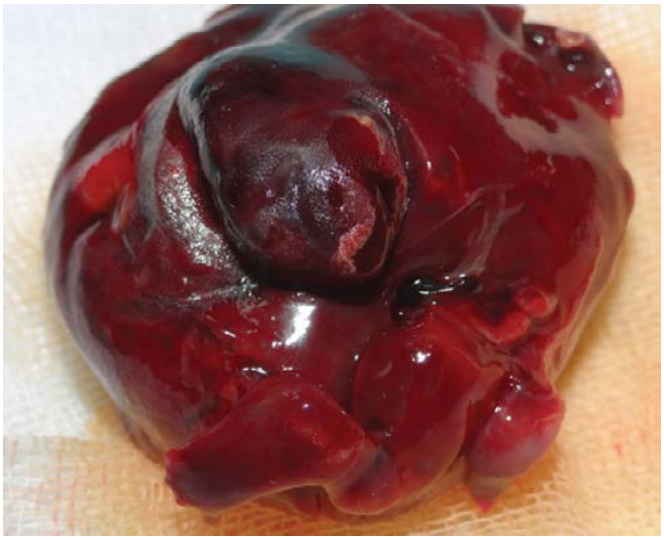

Fig. 1G. Internal view of the mass after removing its outer membrane. Structures consistent with limbs, a face and a gastrointestinal tract can be seen.

The neonate has experienced no complications at the last follow-up, 6 months after surgery.

DISCUSSION

This case of FiF was differentiated from a teratoma by the presence of a vertebral axis and the appropriate arrangement of other organs and limbs relative to the vertebral axis (5). The FiF also had a similar appearance to other cases reported in the literature.

Most authors have attributed the etiology of FiF to anomalous cleavage of monozygotic twins, where one foetus fails to thrive and is entrapped by the other foetus when the anterior abdominal wall closes around the umbilicus and linea alba (10). Therefore, the FiF must be entrapped during the second half of the first trimester or in the early phase of the second trimester.

Although FiF is more common in females (11), the gender of our neonate was male. The FiF was localised in the retroperitoneal space, the most commonly reported site.

Prenatal diagnosis of this rare condition may be difficult because of other possible diagnoses, including hydronephrosis, cystic abnormalities, solid tumours of the kidney, intestinal malformations, teratoma, choledochal cyst, neuroblastoma, adrenal haemorrhage, and infra-diaphragmatic pulmonary sequestration (12).

Ultrasound in the second trimester did not reveal a possible intra-abdominal mass in the present case, as in an earlier case reported by Huddle *et al*. (7). Therefore, FiF might not be visible until late pregnancy.

Most clinicians agree on the superiority of ultrasound imaging in everyday clinical practice, but it may be unsatisfactory in some circumstances. In the management of this case, it is possible that antepartum magnetic resonance imaging could have aided the diagnosis of FiF antenatally, or allowed us to exclude other types of intra-abdominal foetal mass from the differential diagnosis, if it had been performed. Nevertheless, pathologic examination is essential to reach the final diagnosis in such cases because all screening and imaging methods have some limitations that could hinder accurate diagnosis.

Huddle *et al*. highlighted the need for further genetic investigations, such as microarray or single nucleotide polymorphism analyses (7). It may be valuable to use these tools beyond conventional cytogenetic analysis because they can provide additional information regarding the identity of twins. Nevertheless, we think that the decision to use these tools must be clinically justified.

In conclusion, FiF is a rare clinical condition and is seldom considered in the differential diagnosis of a foetal intra-abdominal mass. If a clinician suspects a foetal abdominal mass, he/she should contact to an expert perinatologists and paediatric surgeons. Wherever possible, the foetus should be delivered in a clinic with suitable facilities, including an expert obstetric team, paediatric surgery, and neonatal intensive care unit. A multidisciplinary clinical approach may be more suitable for the management of such rare cases.

Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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