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CASE REPORT

Case report: rare congenital anomaly diagnosed by ultrasonography

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Abstract

Objectives: Report a rare congenital anomaly diagnosed by obstetric ultrasound. **Methods:** The pregnant woman suspected of have fetus with congenital anomaly was attended in the fetal medicine service of a public maternity hospital in Goiás, Brazil. A hyperechogenic nodule in the mesoabdomen of a male fetus were observed through ultrasonography. The totipotent cell mass was withdrawn one week postpartum. The anatomopathological analysis performed confirmed fetal tissue type FIF. **Results:** The ultrasonography performed at the institution detected the presence of a hyperechogenic nodule with 2cm shade in the mesoabdomen of a male fetus, performed at the 30th week of gestation. The totipotent cell mass was withdrawn during labor. It had bones and limbs under development. This pathology is due to abnormal embryogenesis. In these cases, a vertebrate fetus is enclosed within the body of another normally developed fetus and kept by a pedicle in an area filled with liquid or sebaceous material, with no placental or chorionic tissue. Intrauterine growth of FIF is initially similar to its twin, with its subsequent arrested development in a precipitate way due to the "host's" vascular domain or an inherent defect of the "parasitic" twin. The entity is rare and interesting, typically presented in childhood or early childhood. **Conclusions:** Current imaging modalities, such as pre and postnatal ultrasonography, can accurately diagnose the condition. Complete excision is curative and allows confirmation of the diagnosis.

Keywords: congenital abnormalities; ultrasonography; embryogenesis.

Introduction

Worldwide fewer than 200 cases of Fetus in Fetus (FIF) have been reported.[1] The proportion is 1 in 500,000 births, with a predominance of males. The most accepted theory is that occurs an uneven division of blastocyst, resulting in monochorionic monozygotic and diamniotic twin of different sizes contained in the host. It is a rare manifestation and the clinical manifestations are varied. The identification of the FIF can be made through ultrasonography (US) and radiography, with magnetic resonance imaging (MRI) and computerized tomography (CT) used for a more accurate diagnosis.[2] The first prenatal diagnosis of FIF occurred in 1983.[3]

The anatomopathological examination can confirm the FIF, karyotyping, serological evaluator and mapping of the DNA restriction site, after removing the mass. It occurs usually in infancy and early childhood. Usually they are anencephalic and acardiac, but there are reports of FIF with presence of heart and pulse of 108 beats / min.[4,5]

Over time, with imaging technologies evolution, the differentiation between FIF and teratoma became more accessible. Both have similarities in the location of the fetus organ compromised and usually the history of multiple gestations favors the occurrence, both cases can be found in the same fetus. Most FIF are benign and the teratoma has a more common malignant potential. Establishing the difference is important to establish pre- and post-natal management.[6]

The diagnosis made in prenatal care allows for the follow-up of complications that may occur, such as abdominal distension, intestinal obstruction, hydronephrosis, hydrops, jaundice, compression of neighboring tissues and respiratory

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difficulties. To the postnatal phase, it allows for a surgical planning and together with a multidisciplinary team, prepare the family for greater complications, as well as consent processes.[7]

In most cases, it parasites a single fetus, however, there are reports of up to five fetuses. The description of the FIF in relation to the weight, vary between 1.2 and 2000g, and this variation depends on the blood supply.[8] The earliest prenatal diagnosis in the literature is 16 weeks gestation.[9]

Case report

GAMS, 31 years, secundigravida, brown, no case of abortion, the pregnant woman and her husband were healthy and not consanguineous, with no malformations reported in the previous gestation. They did not report family malformation. Both mentioned not having the habit of smoking, of consuming alcoholic beverages, nor of making use of drugs and controlled medicines. The child's father had neurocysticercosis and used specific medication for the disease. The pregnant had no infection nor other health problems during pregnancy, but she reported the use of antiallergics.

The pregnant was referred to the fetal medicine service of a public maternity hospital in Goiás, Brazil, due to the visualization of a shadow on obstetric ultrasound. The imaging diagnosis at the institution, performed at the 30th week of gestation, detected a hyperechogenic nodule with 2 cm shade in the mesoabdomen of a male fetus. The pregnant woman presented placenta grade II (Figure 1-4). The hyperechogenic focus raised the suspicion of being a case of FIF.

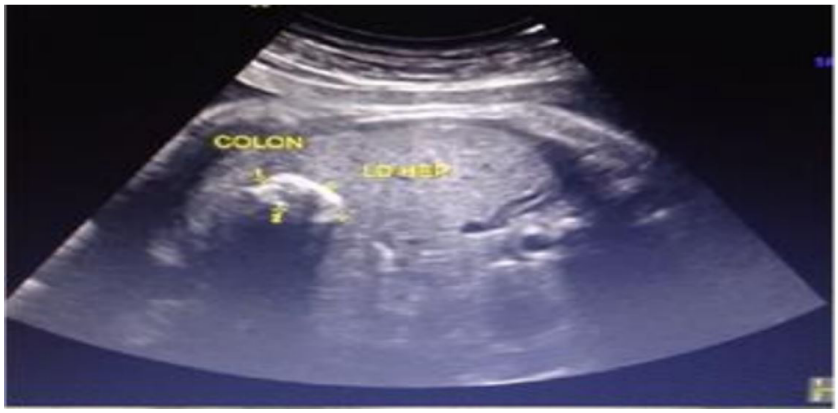

Figure 1. Hyperechogenic nodulation visualized by obstetric ultrasonography in a male fetus performed at the 30th week of gestation.

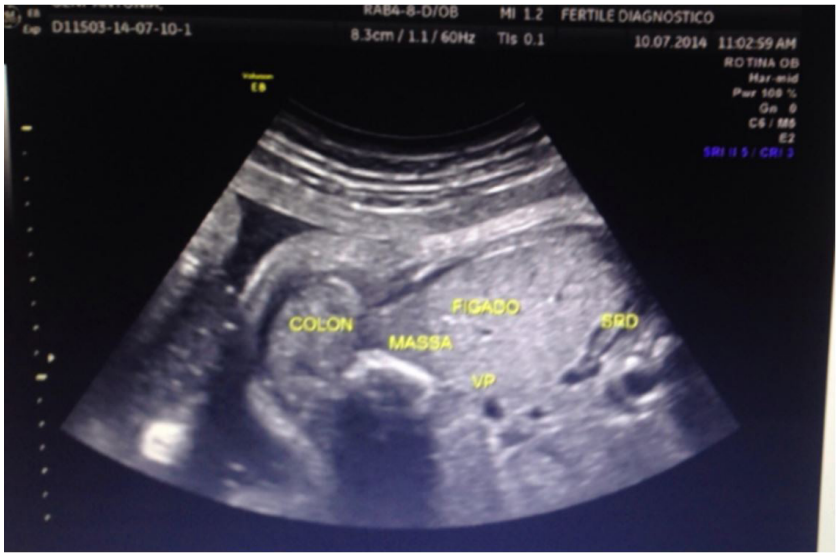

Figure 2. Hyperechogenic nodulation visualized by obstetric ultrasonography in a male fetus performed at the 30th week of gestation.

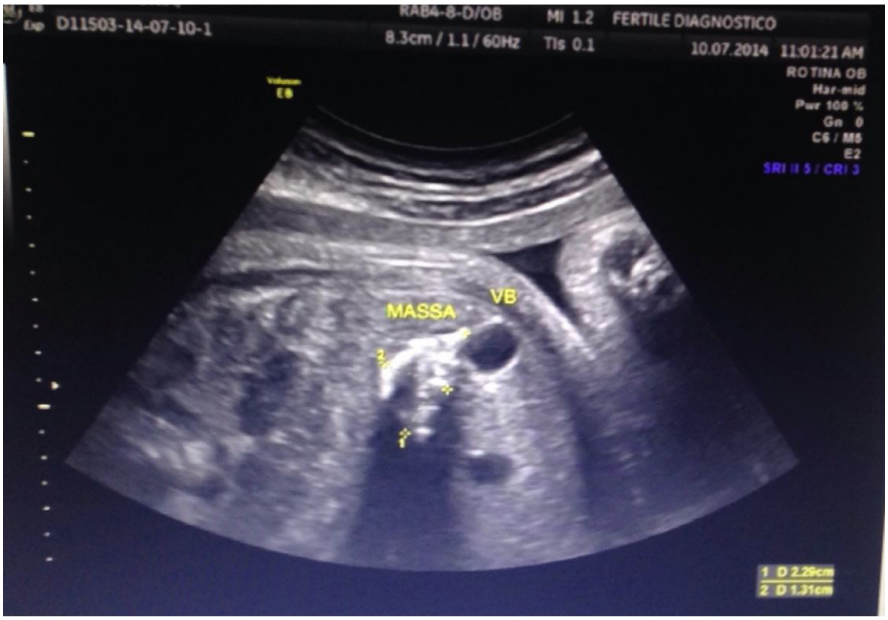

Figure 3. Hyperechogenic nodulation visualized by obstetric ultrasonography in a male fetus performed at the 30th week of gestation.

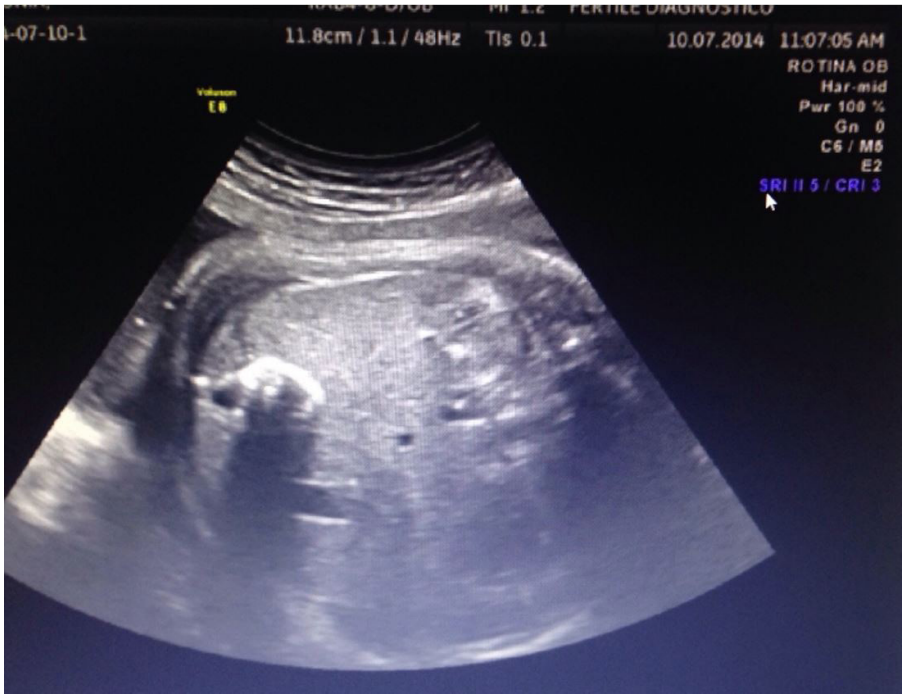

Figure 4. Hyperechogenic nodulation visualized by obstetric ultrasonography in a male fetus performed at the 30th week of gestation.

The pregnancy was conducted until the 39th week and the delivery was cesarean. The baby was born with 49 cm, weighing 3235 g, with cephalic perimeter: 35 cm, thoracic perimeter 31cm and APGAR index: 9/10. He did not present alterations in physical exams.

The totipotent cell mass was withdrawn one week postpartum. The anatomopathological analysis performed confirmed fetal tissue type FIF. It had bones and limbs under development and measured approximately 3 cm (Figures 5 and 6).

The postoperative recovery occurred without interference and the patient presented good clinical evolution, receiving discharge and general guidelines, environmental, dietary and referral to a pediatrician.

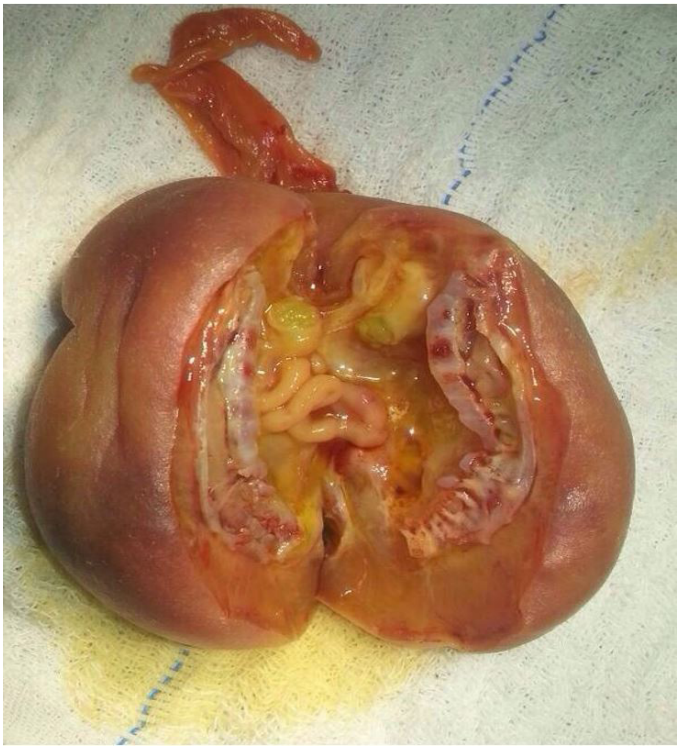

Figure 5. FIF after complete mass excision.

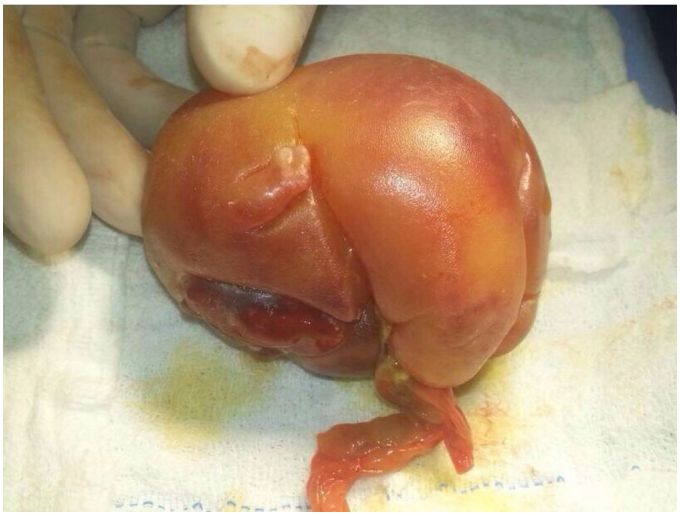

Figure 6. FIF after complete mass excision.

Discussion

The most common localization of FIF is in the retroperitoneal space, but has already been reported in the abdomen, scrotum, kidneys, adrenal, mediastinum, thorax, skull and lymph.[10,11] It is usually not a malignant condition, but it can cause abdominal distension, problems during feeding, vomiting, jaundice, renal and respiratory disorders.[12] In this case, the patient presented a stable clinical picture, without physical alterations, absence of jaundice and satisfactory suction.

There are no reports of the diagnosis of FIF in the first trimester of gestation. Most diagnoses occur in the postnatal period.[13,14] As in this report, Goette et al.[15] detected the presence of FIF in pregnancy after the second trimester through prenatal ultrasonography. However, the fetus with FIF was also affected by omphalocele, pulmonary hypertension, teratomas and complete skin resection. The surgery to remove the mass occurred normally after birth, but after 110 days,

the newborn died due to severe pulmonary dysplasia. In this case study, the infant had a slight superficial scaling, thicker skin at birth and a good clinical evolution after the surgical procedure.

Contrary to this report, Traisrisilp et al.[16] found a female fetus with intracranial fetal organ structure associated with hydrocephalus. In that case, the pathological examination confirmed the diagnosis of FIF. Despite the predominance of FIF in males in previously published cases, intracranial is predominant in females. The presence of a spine and the connection between the fetus and a large vessel is generally a characteristic of the FIF, with 84.3% confirmed with the spine, as in the case presented in this case study. However, it is possible the occurrence of fetus without the spine, differentiating it from fetiform teratoma, which has a less organized structure and has greater malignant potential.[17,18]

Prenatal detection and postnatal follow-up of FIF is very beneficial, as it can be done multidisciplinary follow-up and thus be more successful in the withdrawal. As in the case reported by Sewell et al.[6] in which the 27-year-old pregnant woman, at the 28th week of gestation, detected a white mass in the abdomen and then confirmed the FIF by ultrasonography. In our case, one week after birth, the mass was completely excised and the patient had a good evolution and hospital discharge, reinforcing the importance of the diagnosis in the gestation period. Surgical removal, in most cases, provides complete cure.[19]

The USG has become the most used method for detecting various morphological changes. This is due to its low cost, real-time access to images and wide availability. Other important markers are the serological ones, where levels of beta-HCG or Glycoprotein AFP are considered diagnostic indicators.[20]

Final considerations

Fetus in Fetus is a rare condition found most often in retroperitoneum. The use of imaging devices for prenatal diagnosis is a very useful tool for planning, together with the multidisciplinary team, for subsequent interventions and possible satisfactory resolution.

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Authors contribution

CLM - Participated in the whole process. LCM - Participated in the preparation of the text and the collection on data. FSAT - Participated in the preparation of the text and was corresponding author. SHB - Participated in the data collection and revision of the English text. LBM - Participated in the collection of data. WNA - Participated in the whole process being the study supervisor.