

Fetus– in –Fetu: CT Appearances

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Abstract

Fetus in Fetu (FIF) is a form of monozygotic diamniotic twin pregnancy where the parasitic twin installs and grows in the body of its partner.

It shows calcification in majority of cases and is distinguished by its embryological origin, its unusual location in the retroperitoneal space with presence of vertebral organization and limb buds.

This condition is a rare malformation that has some similarity with the retroperitoneal teratoma, but it is different from the latter by its fetiform aspect and the metameric segmentation of its spinal axis. The presence of a capsule covering this formation and a vascular pedicle is frequently encountered.

Keywords: Fetus; Teratoma; Vertebral organization; Computerized tomography.

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Introduction

Fetus in fetu is a condition in which fetiform calcified mass is often present in the abdomen of its host, a newborn or infant.

This entity is distinguished by its embryological origin, its unusual location in the retroperitoneal space and the presence of vertebral organization with limb buds. It is a form of monozygotic diamniotic twin pregnancy where the parasitic twin installs and grows in the body of its partner.

Case Report

An 18- day female infant with uneventful pregnancy, normal vaginal delivery and body weight of 3,6 Kg was admitted to the pediatric surgery department because of right-sided abdominal mass.

Clinical examination showed firm and fixed mass in the Rt. Upper abdomen showed no bowel or urinary complaints. Laboratory test findings were unremarkable.

Ultrasonography revealed large heterogeneous, well-defined retroperitoneal mass with calcification which was on longitudinal scan suggestive of long bones.

CT scan of the abdomen and pelvis obtained with and without intravenous contrast material revealed a large well-defined retroperitoneal mass in the right upper abdomen extending downward to the right pelvic cavity. The mass predominantly contained areas of fat and fluid that surround a central bony structure. The bony components of the mass resemble the remnants of vertebral column and long bones.

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This retroperitoneal mass displaced the bowel anteriorly; to the left side of the abdomen and displaced the right kidney anteriorly.

Based on these imaging findings a diagnosis of FIF or highly organized teratoma was made preoperatively. Surgical exploration revealed a fetus weighted about 1 Kg, with incompletely formed trunk and entirely covered by skin. Chromosomal analysis was not performed.



Figure (1): Transverse CT scan of the upper abdomen reveals right sided inhomogeneous, well defined mass that has fat, soft tissue and bony components which resemble vertebral bodies.

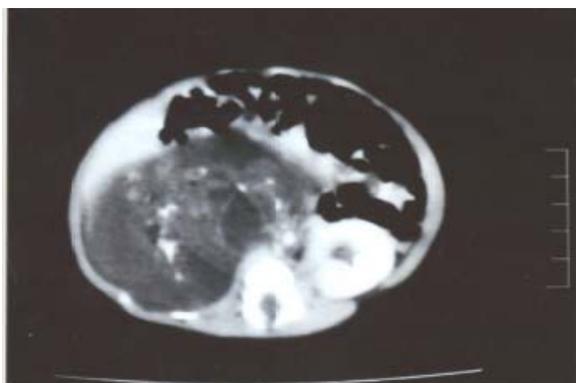


Figure (2): Transverse CT scan, with IV contrast media demonstrates the mass, displacing the right kidney and the bowel loops anteriorly and the left side.

Discussion

Fetus-in-Fetu is a very rare condition which represents a monozygotic diamniotic twin that implants itself and grows within the body of its normal karyotypically identical sibling.

It has an incidence of 1: 500 000 live births® with less than 100 reported cases in the world.

To our knowledge, this was the first case of FIF reported in Jordan. FIF originally was described by Meckel in the late 18th century.¹ In 1809, Young reported the first case of FIF. Since then sporadic cases have been reported. FIF is most commonly discovered in infancy as a retroperitoneal mass.² Which might be totally asymptomatic.³ Symptoms if present, are related to mass effect and including abdominal distention, feeding difficulties, emesis, jaundice and dyspnoea.³

In most cases and as in our case, FIF is present in the form of a mass in the upper part of the retroperitoneal space,⁴ and is commonly surrounded by encapsulated fluid.^{8,9} Other rare locations like cranial cavity, pelvis, scrotal sac, sacro-coccygeal region, mesentery and right iliac fossa are also reported.⁵ Usually, they are one in number, but some cases of more than one have been reported.⁵ There is a controversy, whether FIF represents distinct entity or a highly organized teratoma which is defined as a neoplasm, composed of multiple tissue foreign to the part in which they are located,⁷ with a slight potential for malignancy. It is difficult to make distinction between teratoma and vestigial remnants that result from abortive attempts at twinning.

As a result, some authors think that FIF is within the spectrum of abnormalities that can result from the anomalous embryogenesis in a diamniotic monochorionic pregnancy.

The spectrum includes conjoined symmetric twins; parasitic fetuses; embryonic vestigial inclusion; and organized teratoma.⁷ Thus some authors claim that FIF is only a well-differentiated organized teratoma. The presence of vertebral column as emphasized by Willis,⁶ secures the diagnosis of FIF and differentiates this entity from teratoma. Identification of the vertebral column indicates that fetal development of the included twins must have advanced at least to the primitive steak stage to develop a notochord, which is the precursor of the vertebral

column.⁶ Many imaging modalities played an important role to correctly diagnose FIF which can be made with abdominal radiographs, by identifying a vertebral column and/or specific bony structures.⁶ Nocera et al.¹⁰ initially described the CT appearances of FIF, which includes well defined, encapsulated collection of fat that surrounds a central bony structure that resemble a vertebral column or long bones. As in our case, CT findings were helpful in preoperative diagnosis of FIF.

In reviewing the literature, most case reports before 1980 showed that preoperative diagnosis was made only in 16.7% of cases because CT scan was not performed. Nowadays, CT scan has proven very helpful^{1, 11} in suggesting the preoperative diagnosis. Magnetic resonance imaging was also used in 4 cases.^{3, 12, 13} In our case, like some of the cases described in the literature, the vertebral column was detected by the pathologist. It was suspected on CT scan of the abdomen. So our case truly had a vertebral column like 91% of cases in literature. It was therefore in accordance with Willis theory⁹ that FIF is a mass containing vertebral axis often associated with other organs or limbs around this axis. However, reviewing the literature showed that in about 9% of cases of FIF there was no vertebral column, even on pathological examination.¹⁴ This has led to another definition of FIF by Gonzalez-Crussi:¹⁴ "FIF is applied to any structure in which the fetal form is in a very high development of organogenesis and to the presence of vertebral axis."¹⁴

On the contrary, teratoma is an accumulation of pluripotential cells in which there is neither organogenesis nor vertebral segmentation.¹⁵ Although the hypothesis regarding the origin of the FIF was widely accepted as the included twin theory, which was postulated as a diamniotic monochorionic monozygotic twin, some supporters of the teratoma theory have suggested that the FIF mass represent well-differentiated, highly organized teratoma.

Conclusion

To our knowledge, this is the first case of FIF reported in Jordan. The diagnosis was made preoperatively and confirmed by pathology examinations. Medical imaging plays an important role in this diagnosis and it is important to be aware that the nonvisualization of the vertebral axis does not exclude this diagnosis.

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الحمل التوأمي بشكل جنين - في - جنين، مشاهدات بواسطة التصوير المقطعي المحوسب

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الملخص

جنين-في-جنين هو شكل من أشكال الحمل التوأمي لأجنة ثنائية السلي وحيدة اللاقحة حيث يتوضع التوأم الطفيلي و ينمو في جسم الأم و تظهر تكلسات في معظم الحالات تتميز بالمنبت الجنيني متوضعا في موضع غير طبيعي (الحيز خلف البريتوان)، مع ظهور انتظام الفقرات وظهور براعم الأطراف. وتعد هذه الحالة من أندر حالات التشوهات التي تشبه الورم العجائبي (تيراتوما) خلف البريتوان، ولكنها تختلف عن السابق بمظهر الجنين والتجزؤ (التماكي = السيء) لمحور النخاع الشوكي، وكثيرا ما يصادف ظهور كبسولة تغطي هذا التشكيل وظهور سويقة وعائية (عنق وعائي رقيق).

الكلمات الدالة: جنين، الورم العجائبي، التنظيم الفقري، التصوير المقطعي بواسطة الكمبيوتر.