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Case Report

A case report of fetus in fetu with an aorta-like structure visualized by contrast-enhanced CT ☆,☆☆

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ABSTRACT

Fetus in fetu (FIF) is a rare congenital anomaly resulting from abnormal embyogenesis in monochorionic diamniotic twins and appears as a cystic mass containing fetus-like structures mainly in the retroperitoneum of infants. Although there is a theory that FIF is a highly differentiated teratoma, it is commonly distinguished from teratoma as a mass containing a vertebral axis with appropriate arrangement of limbs or other organs around this axis. Here we present a case of FIF with aorta-like structure visualized by contrast-enhanced computed tomography. A 5-day-old girl was pointed out a cystic mass in the abdomen by ultrasound examination. Abdominal contrast-enhanced computed tomography revealed a retroperitoneal cystic mass with spine- and limb-like bone structures and blood vessel-like elongated structures and it was confirmed as FIF by surgery. The presence of major vascular structures along the skeletal axis is clearly different from teratoma and suggests that it occurred as an embryo and underwent some stage of development. Our findings strongly support the monozygotic twin theory.

Introduction

Fetus in fetu (FIF) is a rare congenital anomaly with an incidence of 1 per 500,000 births. Multiple theories have been proposed with regard to the etiology of FIF. The most common theory is abnormal embyogenesis in diamniotic monochorionic twins, in which a parasitic fetus is incorporated into the body of its twin partner and grows inside of it [1–4]. Another theory is that FIF is a highly differentiated form of teratoma [1–4]. FIF was first described by Johann Friedrich in early 19th century. It was later defined by Willis

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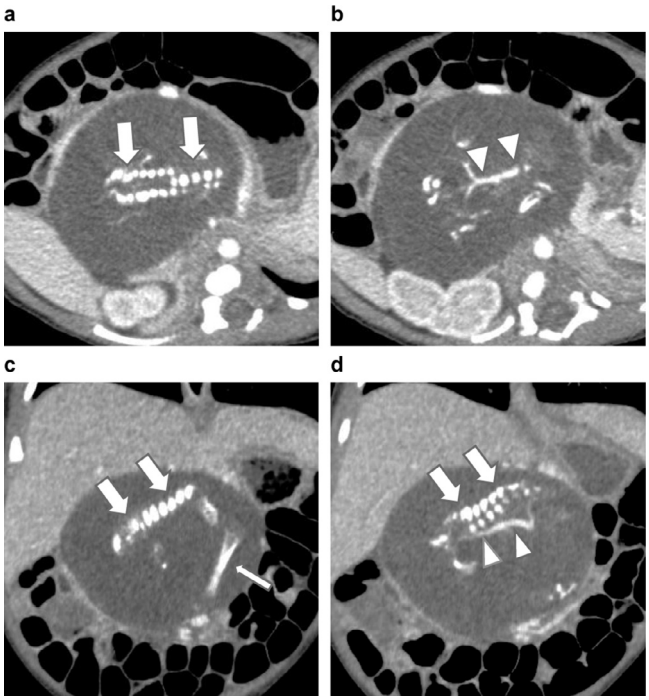

Fig. 1 – Abdominal contrast-enhanced CT on the day of visit (a, b) Axial and (c, d) coronal multiplane reformats of enhanced-contrast CT of the abdomen revealed a retroperitoneal cystic mass of approximately 6 cm. A spine-like bone structure (thick arrow), limb-like bone structure (thin arrow), and a blood vessel-like elongated structure(arrowhead) were observed within the cystic mass.

as a mass containing a vertebral axis, often with appropriate arrangement of limbs or other organs around this axis [1]. We present a case of FIF with vascular structures visualized by contrast-enhanced computed tomography (CT).

Case report

A 5-day-old girl was referred to our hospital because a cystic mass was found in the abdomen by ultrasound examination during pregnancy and after birth. Abdominal contrast-enhanced CT revealed a retroperitoneal cystic mass approximately 6 cm in diameter, extending between the celiac and superior mesenteric arteries. Within the cystic mass, spine-like bone structures, limb-like bone structures, and blood vessel-like elongated structures were observed (Fig. 1). Vascular structures, and spinal column-like and lower limb-like skeletal structures were revealed by 3D-reconstruction images (Fig. 2), in addition to structures similar to the aorta, bilateral common iliac arteries, and the umbilical arteries branching therefrom. Due to continued vomiting by duodenal compression of the mass, excision surgery was performed on day 16. The mass was located in the retroperitoneum. An umbilical cord-like structure was continuous with the cyst wall and a fetus-like component with limbs was found inside. Continuing to the spine, pelvic- and lower limb-like bones were observed along the skeletal axis. Cephalic and caudal ends were easily identifiable. A rib cage-like structure was also identified (Fig. 3). Pathological findings confirmed skin and bone tissue, such as the spine, cartilage, and skeletal muscle, on the outer surface, and it was diagnosed as FIF. Most of the tissues were necrotic, and it was difficult to identify other organs, including the vascular structure observed by contrast-enhanced CT. As inflammatory response in the infant was present before surgery, ischemic necrosis of the FIF was speculated due to blood flow impairment after birth.

Discussion

FIF is a cystic mass containing fetus-like structures found mainly in the retroperitoneum of infants. It is a rare condition with less than 200 cases reported worldwide [4].

The most common site of FIF is the retroperitoneum (72%-80%), and less frequent sites include intracranial sites, oral cavity, neck, mediastinum, thorax, liver, adrenal gland, spleen, sacrococcygeal regions, and scrotum [2–4]. FIF typically presents during infancy and early childhood; however, there are several case reports in which FIF was found during adulthood [3]. FIF is enclosed by a distinct sac and suspended by an umbilical cord-like structure containing blood vessels. A vertebral column was identified in 76%-91% and limb buds were also frequently identified in 83%-86% [1–4]. Lower limbs were more developed than upper limbs. Central nervous tissue, gastrointestinal tissue, and genitourinary tissue are often present [1–4]. Most cases of FIF are acardiac; however, there have been cases of FIF with a cardiac structure, including a case with a heart rate of 108 beats/min [4]. Blood vessels have been reported in FIF, but specific vasculature, such as the aorta, has not been reported. Although there is only one case report in which CT angiography visualized the feeding vessel of the FIF [5], there has been no report of the vascular-like structure in FIF on CT.

The pathogenesis of FIF remains controversial. The 2 most common theories are the monozygotic twin theory and teratoma theory [1–4].

The monozygotic twin theory proposes an abnormality in the development of a monochorionic diamniotic twin, suggesting that FIF becomes incorporated into the body of a host twin as a parasite [1–4]. The monochorionic diamniotic twin results from the division of the zygote 4-8 days after fertilization. If one of the blastocysts implants into the other blastocyst instead of the endometrium or is enclosed by the other, the subsequent development of an inclusion body may result in FIF [4]. The teratoma theory proposes that FIF is an extreme form of well-differentiated, highly organized teratoma with mature organs [1,6]. In the teratoma theory, highly differentiated teratomas may contain a variety of organs such as well-formed limbs, teeth, intestinal loops, a spinal cord, and brain-like tissues [6]. Several case reports suggest that certain teratomas exhibit organoid differentiation with a rudimentary heart, brain, eye ball, intestine, and limbs [4,6].

Willis et al proposed that FIF was not a well-developed teratoma and suggested that diagnosis of FIF should be restricted to cases in which parts of the axial skeleton were present with

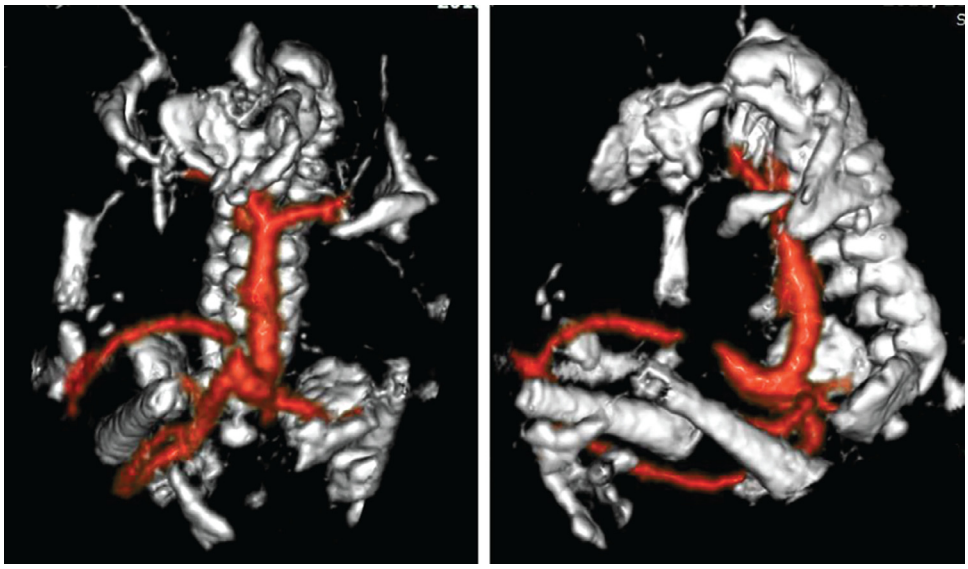

Fig. 2 – 3D reconstruction image of enhanced-contrast CT Spinal column- and lower limb-like skeletal structures and vascular structures were observed. The vascular structures resembled the aorta, bilateral common iliac arteries and umbilical arteries branching therefrom.

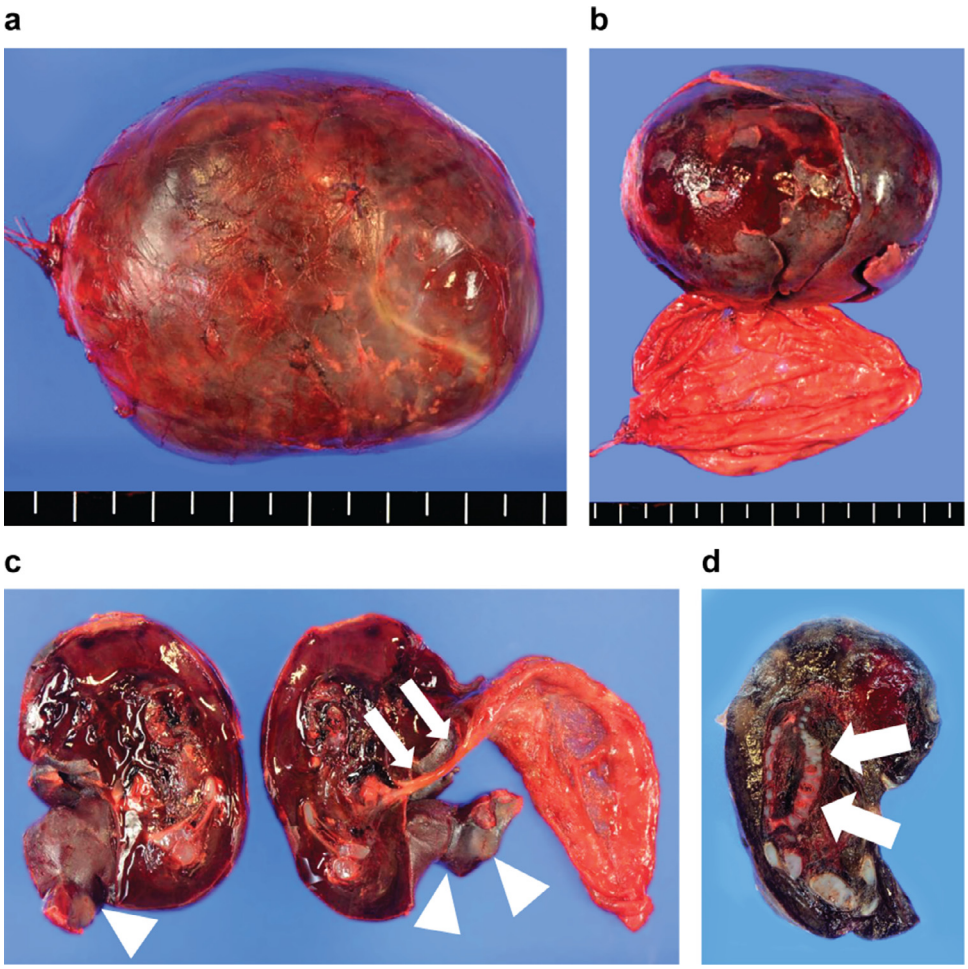

Fig. 3 – Gross photograph of the excised cystic mass (a, b) and sagittal cross section (c, d). The mass was 65 x 50 x 35 mm. A fetus-like mass with limb buds (arrowhead) was inside and connected to the cyst wall with a cord-like structure (thin arrow). Sagittal cross section revealed the spinal column (thick arrow).

an appropriate arrangement of organs relative to the skeletal axis in the body of its host, usually in the abdominal cavity [1]. The existence of the spinal column suggests that FIF occurs as one embryo and its development has progressed beyond the primitive streak stage, forming the notochord, neural tube, and spinal column, which is consistent with the former theory. The spine-, pelvic-, and lower limb-like bones were continuously arranged in our case. In addition, a vascular structure resembling an aorta, bilateral common iliac arteries, and umbilical arteries branching therefrom was identified along the skeletal axis by contrast-enhanced CT. These different organs, including a skeleton and blood vessels, were properly arranged to form a human body. The establishment of the axis suggests that the FIF in this case occurred as an embryo and underwent the initial stage of development. This is clearly different from teratoma, which demonstrates discordant congregations of pluripotent cells without systemic organization regardless of how mature the included organs are. Therefore, this case supports the monozygotic twin theory of FIF. It is important to differentiate FIF from teratoma because teratoma has independent growth ability and malignant potential. However, there are reports of retroperitoneal teratoma with malignant components after FIF removal [7] and coexistence of FIF and immature teratoma [8]. Therefore, it is recommended to follow up patients after FIF removal for a certain period of time.

Conclusion

Vascular-like structures were identified in FIF by contrast-enhanced CT. Although there are numerous opinions regarding the definition and etiology of FIF, we believe that the presence of major vascular-like structures along the skeletal axis supports the monozygotic twin theory.

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