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Case Report

Retroperitoneal fetus in fetu presenting in a male infant: A case report and literature review[☆]

Yixiao He, Master of Medicine Science, Yuzhu Ji, Master of Medicine Science, Gang Xie, Bachelor of Medicine Science, Anqun Wang, Bachelor of Medicine Science*

Department of Pathology, Mianyang Central Hospital, School of Medicine, University of Electronic Science and Technology of China, Changjia Alley, No.12, Fucheng District, Mianyang City, Sichuan Province, 621000, China

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ABSTRACT

Fetus in fetu (FIF) is a rare congenital disease caused by the abnormal development of monozygotic diamniotic twins that appears as a cystic mass containing fetus-like structures mainly in the retroperitoneum of infants. The clinical manifestations of fetus in fetu vary, but they mostly present at infancy, hence, it should be differentiated from a teratoma. Here, we report a case of an infant with fetus in fetu in the retroperitoneum. Enhanced computed tomography scans and three-dimensional images showed a huge mixed-density mass on the left side of the abdominopelvic cavity with patchy distribution of fat, intact bones, and soft tissue. The child underwent fetus in fetu resection under general anaesthesia. Histopathology confirmed that the mass contained skin, muscle, intestinal mucosa, bones and cartilage, nerves, muscles, fat, and bone marrow tissue.

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Introduction

Fetus in fetu, also known as cryptodidymus, is a rare congenital disease that occurs when a complete fetus is parasitized by one or several incomplete fetuses. It is usually caused by the abnormal development of monozygotic diamniotic twins, which is rare. Fetus in fetu can be divided into 2 types (internal and external), and it commonly occurs in the retroperitoneum and abdominal cavity. It presents as a gradually enlarging abdominal mass, and differentiated from other abdominal tumors, including teratomas, is necessary. It can be diagnosed by radiography, ultrasonography, computed tomography (CT),

and magnetic resonance imaging (MRI). Here, we report the clinical features of a retroperitoneal fetus in fetu in an infant.

Case report

Clinical presentation

A male infant aged 2 months and 10 days was brought to the hospital after his parents accidentally noted a lump in his abdomen. The abdomen was slightly hard, and a mass, approximately 10 × 6 cm in size, could be palpated around the umbilicus, which shifted when the infant changed positions. The infant was vomiting, and he was irritable when touched. Peri-

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* Corresponding author.

E-mail address: 1075468354@qq.com (A. Wang).

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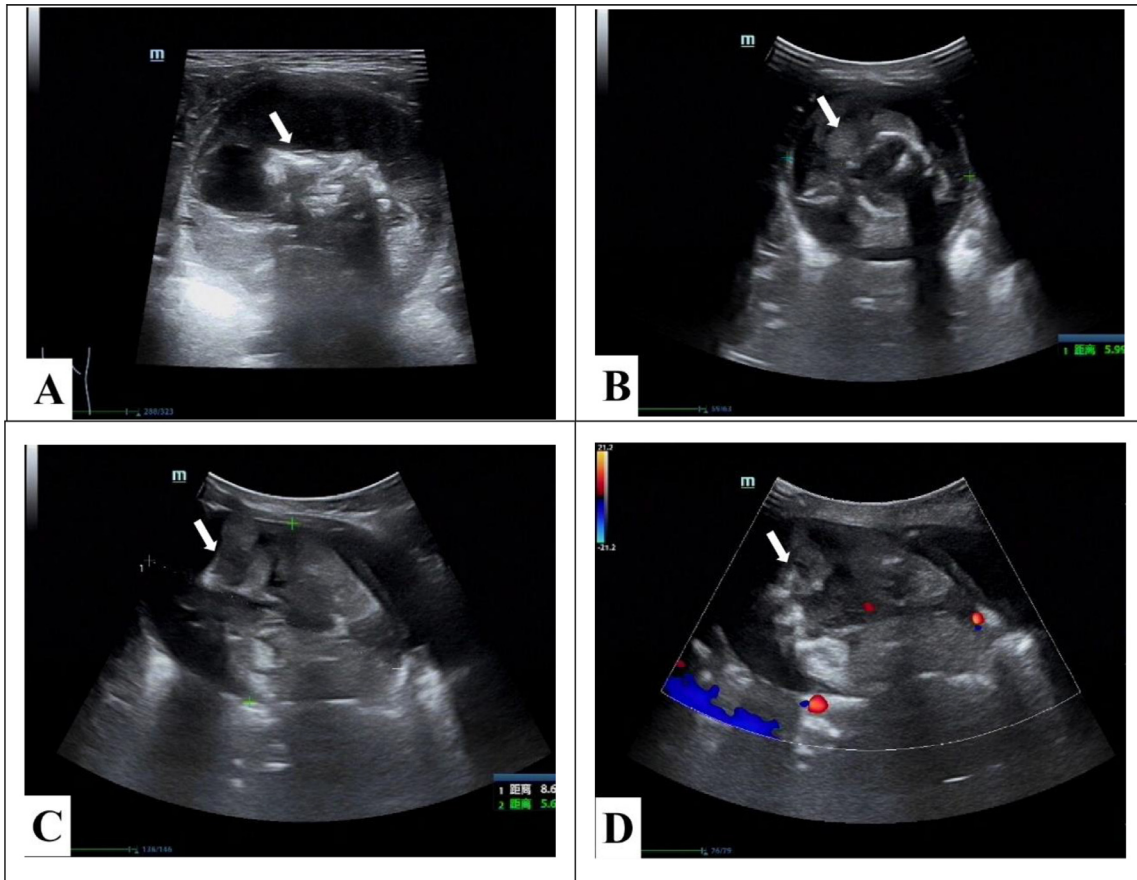


Fig. 1 – Ultrasonographic images. (A-C) Ultrasound images show a mixed-density echogenic mass in the abdominal cavity with clear boundaries and slightly strong echo. (D) Color Doppler Flow Imaging(CDFI) showed blood flow signal.

natal history revealed that the infant was delivered term via caesarean section, and there were no antenatal complications. He had a birth weight of 3.55 kg, and neonatal asphyxia or birth injuries did not occur. His parents were healthy and had a non-consanguineous relationship, and there was no history of genetic metabolic diseases in the family.

Auxiliary examinations

Ultrasonography revealed an echogenic mixed density mass in the abdominal cavity measuring $8.7 \times 6.0 \times 5.7$ cm with clear boundaries and a slightly strong echo, a teratoma was initially considered (Fig. 1). Plain and contrast enhanced CT scans and three-dimensional (3D) imaging of the whole abdomen (Fig. 2 and Fig. 3) A huge mixed-density mass was observed on the left side of the abdominopelvic cavity with patchy distribution of fat, intact bones, and soft tissue. The cross-sectional size of the mass measured approximately $6.6 \times 5.0 \times 7.6$ cm, the edge of the lesion was relatively smooth, and its soft tissue components were significantly enhanced after contrast enhancement. The adjacent rectum and bladder were displaced to the right side, while the small intestine was displaced upwards. Based on these findings, fetus in fetu was suspected.

Intraoperative findings

The mass was in the retroperitoneum, measured approximately $10 \times 6 \times 5$ cm, and closely adhered to the surround-

ing intestinal wall. After *en bloc* resection, skin, hair, a thorax and spine and long bone-like and intestinal tract-like structures could be seen. No significant abnormality was found in the rest of the abdominal cavity upon exploration (Fig. 4).

Pathological findings

An embryo-like structure measuring $8.5 \times 6 \times 4$ cm with skin attached to its surface, a few hairs on some parts of the skin and an intestine-like structure with a length of 2 cm and diameter of 0.5-1 cm was resected. Bone tissue and lacunae of the spine, skull and limbs could be seen upon dissection (Fig. 5).

The skin, muscle, intestinal mucosa, bone and cartilage tissue, nerves, muscles, fat and bone marrow tissue were confirmed with microscopy, which were consistent with a fetus in fetu (Fig. 6).

Discussion

Fetus in fetu, also known as cryptodidymus, is an extremely rare congenital disease caused by the abnormal development of monozygotic diamniotic twins, which has an incidence rate of 1 in 500,000 [1,2]. The disease was first described by Meckel [3] at the end of the 18th century, and was defined as a complete fetus parasitized by 1 or several incomplete fetuses.

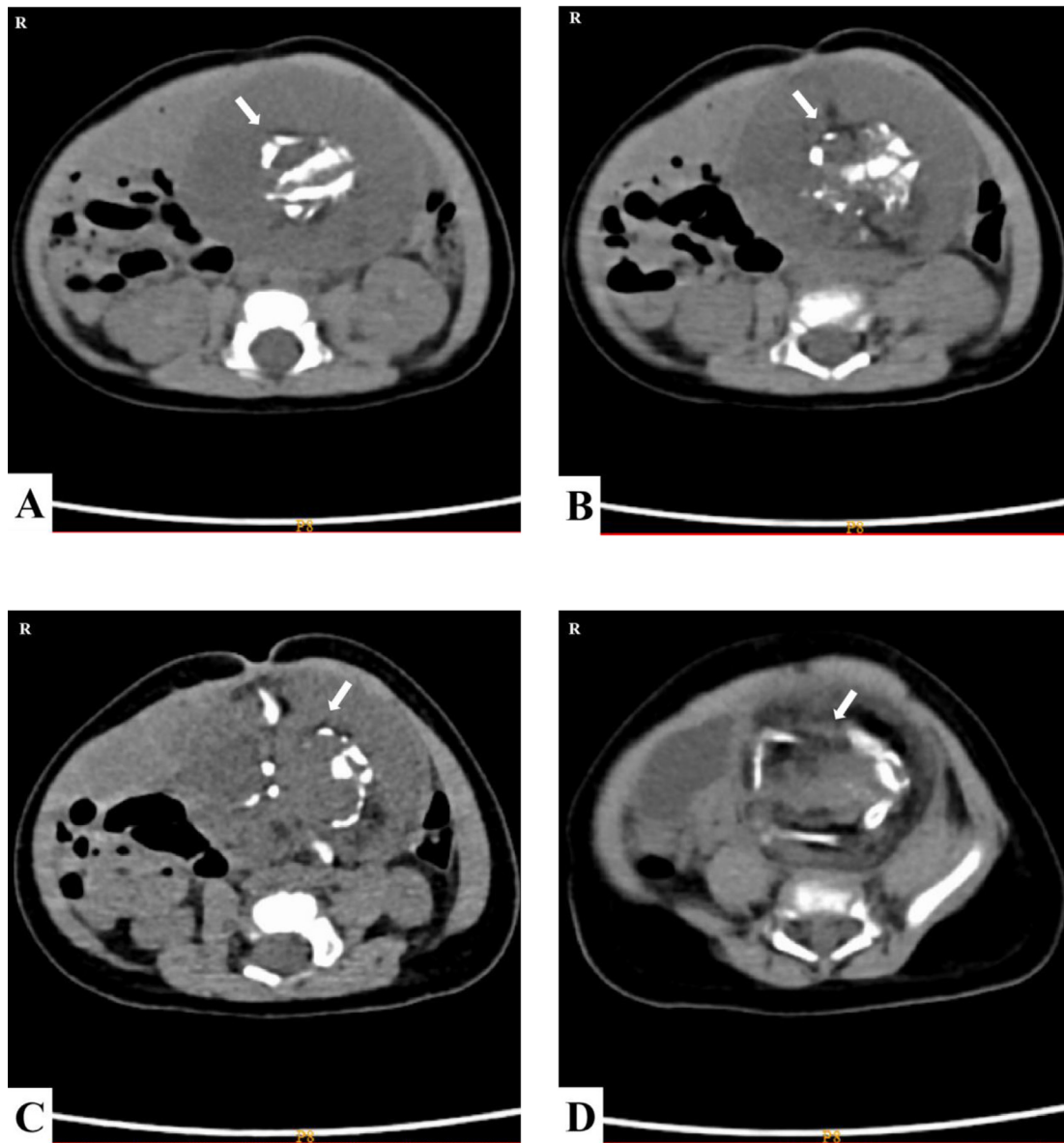


Fig. 2 – (A-D) Axial computed tomography images showing an intra-abdominal mass with fat, intact bones, and soft tissue displacing the bowel loops.

Due to the extremely low incidence rate, and there are currently no epidemiological statistics regarding the condition. If a fetus is inside the body of a larger one, it is called an internal fetus in fetu, if it is attached to the surface of a fetus, it is called an external fetus in fetu. In existing reports, it was discovered that the condition commonly occurs in infants and young children, and most cases were diagnosed before or shortly after birth. There is no gender predominance in the incidence of fetus in fetu, and patients <3 years old account for more than 90% of the cases. However, fetus in fetu can be found in adults [4] and in pregnant women [5], the oldest patient with fetus in fetu recorded in the literature was 47 years old at diagnosis [6]. Fetus in fetu can parasitize any part of the host's body; however, the abdominal cavity and retroperitoneum are the most common sites [2,7,8] followed by the sacrococcygeal

region and the abdominal wall. Other rare sites, such as the back, mouth and upper jaw, have also been reported, and very few cases have occurred in the brain [9,10], mouth [11,12], neck [13], chest [14] and scrotum [15]. Therefore, fetus in fetu lacks characteristic signs, but it can cause jaundice, hydronephrosis, intestinal obstruction, meconium peritonitis, respiratory distress and vomiting in the host. Most cases of fetus in fetu have a single parasitic twin, but cases with more than one parasitic twin have also been reported [16], the maximum number of parasitic twins reported in the literature is [11,17].

A fetus in fetu can harm their hosts, therefore, early diagnosis and evaluation are important. An early diagnosis of fetus in fetu can be made through imaging modalities. Aside from color Doppler ultrasonography, CT and MRI can visualize bones and the spinal canal in fetus in fetu, which increases the

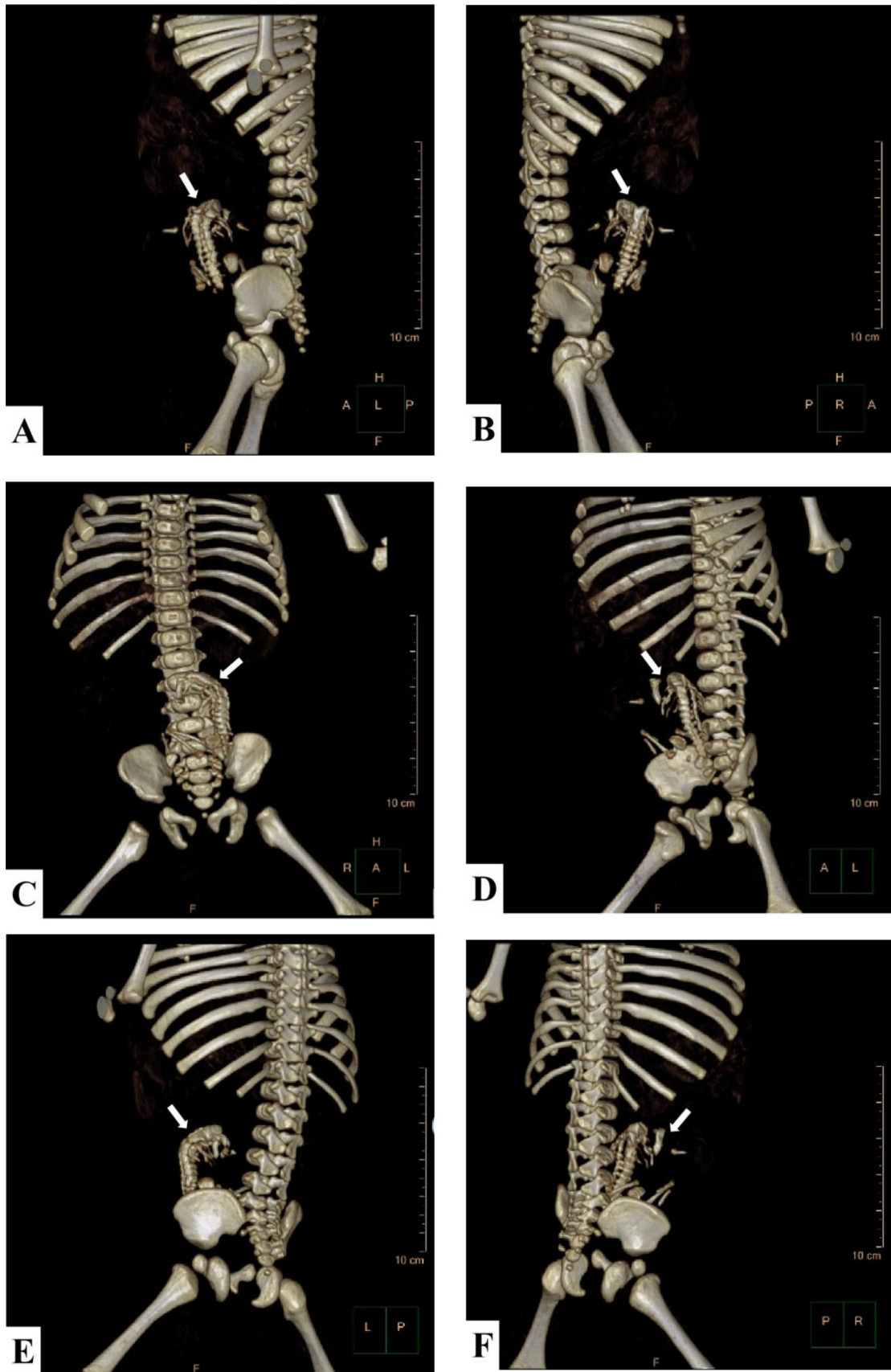


Fig. 3 – Three-dimensional reconstruction of fetus in fetu showing long bones, skull, and dysmorphic vertebrae. (A-B) sagittal view, (C-F) coronal view.

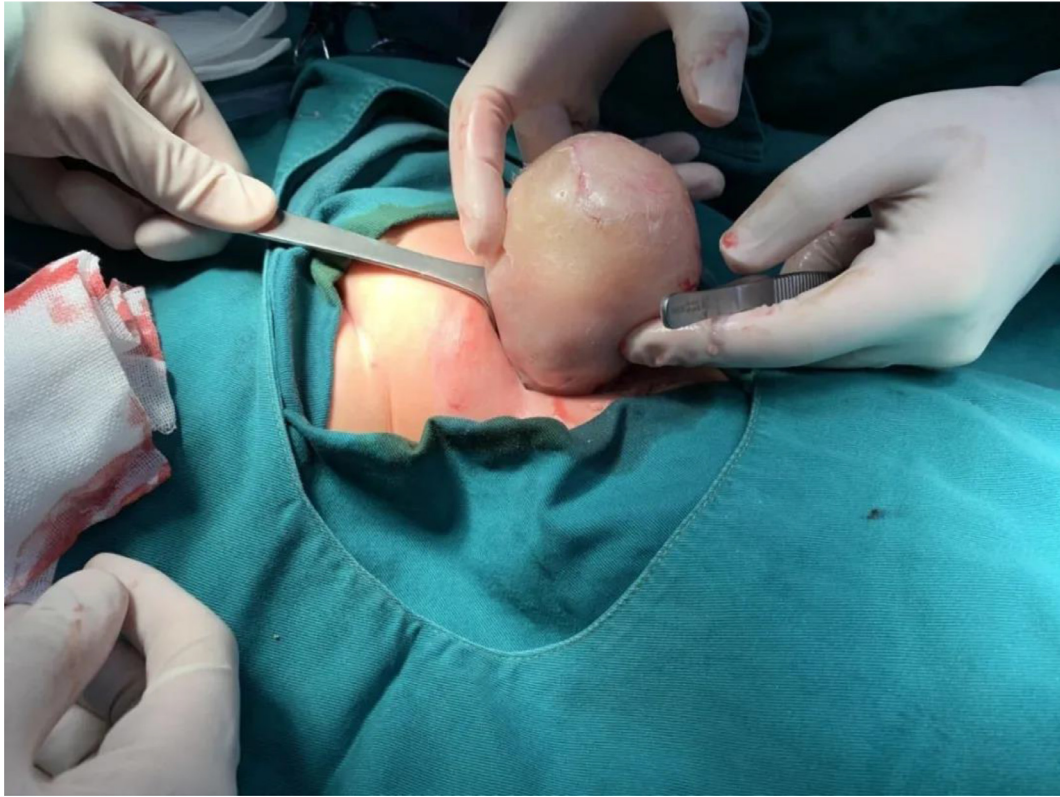


Fig. 4 – The mass was completely resected under general anaesthesia.



Fig. 5 – Gross specimen of the mass. (A-B) The appearance of the mass. (C) Upon incision, the mass exhibited long bones, skull, and dysmorphic vertebrae.

preoperative diagnosis rate. Besides imaging evaluation, post-operative pathological examination is also very important, as the presence of various well-differentiated organs, including the spine, limbs, fingers and other organs, confirms the diagnosis. Spencer suggested that a fetus in fetu should meet one or more of the following criteria: (1) separately encapsulated; (2) partially or completely covered by normal skin; (3) having recognizable anatomical structures; (4) connected to the host through large blood vessels; and (5) conjoined twins via the neural tube or gastrointestinal system. Another diagnostic method is to use molecular analysis via the isodisomy informative genetic markers of parthenogenesis on chromosomes 14 and 15, if there is no genetic difference between the

host and fetus-like mass, it can be diagnosed as a fetus in fetu [18].

Fetus in fetu must be differentiated from a teratoma [19] because the latter may have malignant transformation. A teratoma is a type of tumor produced by agamic unfertilized germ cells that mainly originate from the gonadal embryonic cells and nourished by small blood vessels, however, it is difficult to find recognizable and complete tissues in it [20,21]. A teratoma has the general characteristics of a neoplasm, whereas a fetus in fetu is an abnormal development in the differentiation of the fertilized egg. However, there are reports in the literature that state that a fetus in fetu may become malignant [1]. In addition, some scholars believe that

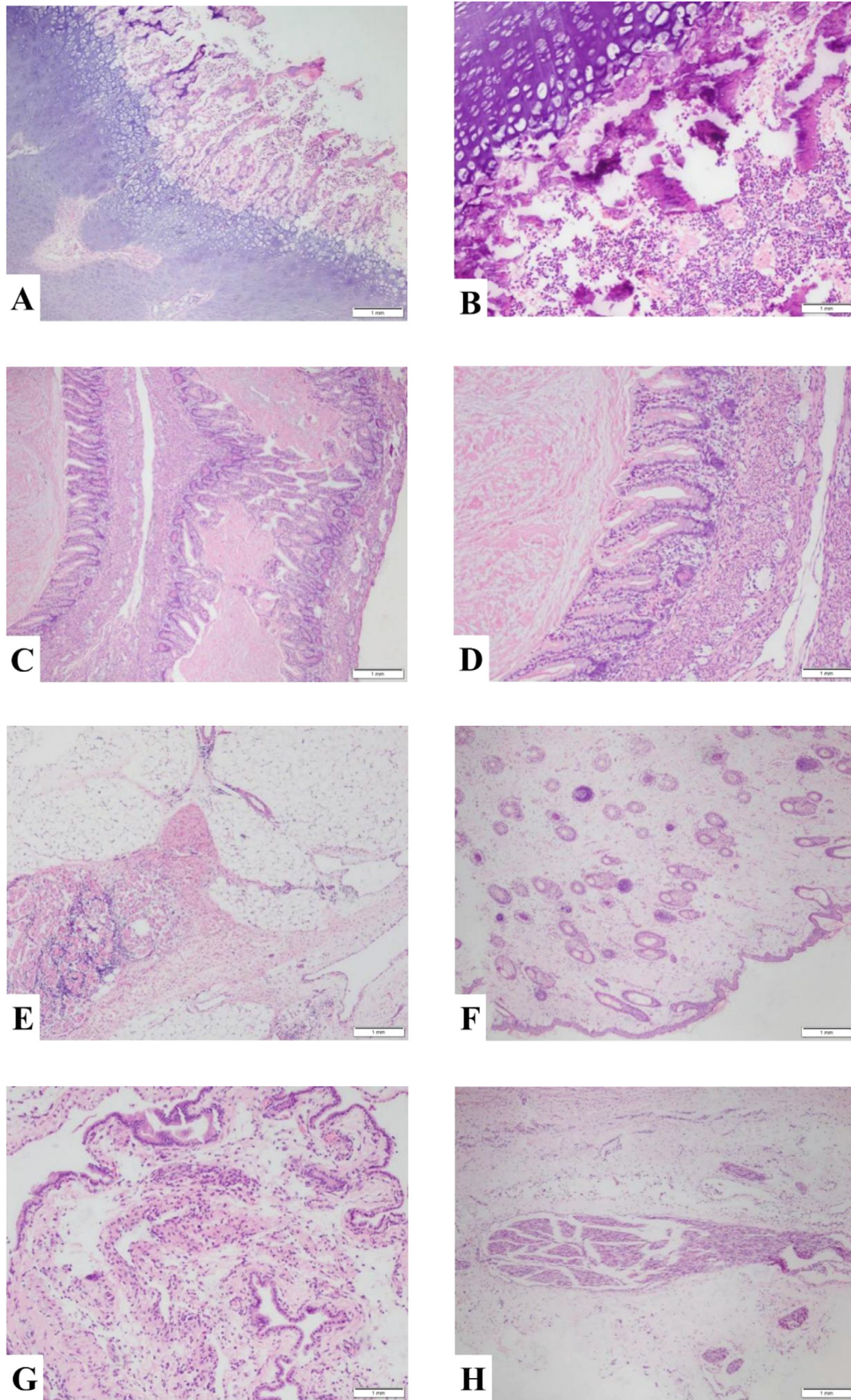


Fig. 6 – Pathology of the mass. (A-B) Bone, cartilage tissue, and bone marrow tissue (A: 40x, B: 100x); (C-D) Intestinal tissue (C: 40x, D: 100x); E: Muscle and fat (40x); F: Skin and appendages (40x); G: Mucosa (40x); H: Nerves (40x).

fetus in fetu is a highly differentiated teratoma, and both a teratoma and fetus in fetu can even occur simultaneously. Fetus in fetu also needs to be differentiated from meconium peritonitis [1], which is caused by intrauterine bowel perforation, resulting in meconium infiltration of the peritoneal cavity. The most common imaging finding in neonates with meconium peritonitis is calcification, which must be differentiated from calcification related to bones in fetus in fetu. Fetus in fetu requires surgical resection and long-term follow-up after surgery. At present, alpha-fetoprotein and β -human chorionic gonadotropin are important markers [22] that can indicate whether there is malignant transformation.

Fetus in fetu has different clinical manifestations, and most cases occur in infants and young children. The aetiology of a fetus in fetu is unclear, and it may be related to the environment, pollution, heredity, abnormal embryonic cell division and other factors. Hence, its pathogenesis remains controversial. Most scholars believe that the totipotent cell clusters in blastocysts divide into more than two or three inner cell clusters in the early stage of embryonic development. For some reasons, the development of one or several inner cell clusters is limited or discontinued, and they are enclosed in the fetus formed by other cell clusters, thus transforming into single or multiple fetus in fetu. However, this theory is not sufficient to explain all cases of fetus in fetu. Another theory is the fusion theory wherein two independent foetal sacs fuse in later stages of development [23]. Other theories include multiple pregnancies and variants of mature teratomas [24,25]. Due to its origin from the same fertilized egg as the host and its incomplete absorption by the host because of its retarded growth in the process of differentiation and development, the blood type and chromosomes of fetus in fetu are similar to that of the host [26].

Fetus in fetu has the potential to cause harm to the human body, especially in neonates. Hence, it is necessary to diagnose and distinguish it from teratoma as early as possible. All cases require surgical resection of the embryo together with the cyst so as to avoid leakage of the cystic contents, the prognosis is favourable after *en bloc* resection [27]. Long-term follow-up should be conducted after surgery. A deeper understanding of this disease can improve the accuracy of preoperative diagnosis and enable clinicians to select appropriate treatments as well as increase the cure rate of patients.

Patient Consent

The parents of the patient provided informed consent, and the study design was approved by the appropriate ethics review board.

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