Case Report
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Sacrococcygeal Fetus-in-Fetu Mimicking a Teratoma: A Rare Case with Brain Tissue and an Immature Teratoma Component[1]

기형종과 유사하게 보이는 천미골 태아 내 태아(Fetus in Fetu) : 뇌조직과 미성숙기형종 요소를 포함한 1예 보고[1]

Cherry Kim, MD[1], Hye-Kyung Yoon, MD[1], Young Ah Cho, MD[1], Jae-Yeon Hwang, MD[1], Ai-Rhan Kim, MD[2], Hee Sang Hwang, MD[3], Chong Hyun Yoon, MD[1]

Departments of [1]Radiology and Research Institute of Radiology, [2]Neonatology, [3]Pathology, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Korea

Fetus in fetu is a rare, nonviable, malformed parasitic twin, which grows within the body of its partner. It has been known as being almost always anencephalic and rarely reported to have an immature teratoma component. We report a case of a sacrococcygeal fetus-in-fetu with brain tissue seen on both imaging studies and pathologic specimens, containing an immature teratoma component on pathologic examinations. Imaging studies including plain radiography were very helpful for the correct diagnosis.

Index terms
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Corresponding author: Chong Hyun Yoon, MD
Department of Radiology and Research Institute of Radiology, Asan Medical Center, University of Ulsan College of Medicine, 88 Olympic-ro 43-gil, Songpa-gu, Seoul 138-736, Korea.
Tel. 82-2-3010-4362 Fax. 82-2-476-0090
E-mail: chyoon@amc.seoul.kr

INTRODUCTION

Fetus in fetu is a very rare congenital anomaly, which was first described by Meckel in the nineteenth century (1). It is a nonviable, malformed parasitic twin, which grows within the body of its partner. When it is located in the sacrococcygeal region, fetus in fetu can mimic sacrococcygeal teratoma. Fetus in fetu can be differentiated from mature cystic teratoma by identifying the vertebral column or other well developed organs (1).

Here, we report a rare case of a sacrococcygeal fetus-in-fetu with brain tissue, containing an immature teratoma component on pathologic examination.

CASE REPORT

A baby girl was born at 35 weeks and 2 days of gestation via elective caesarean section with a birth weight of 4020 g. Her mother was 31 years old and healthy with a history of induced abortion. There was no family history of twin pregnancy.

A fetal ultrasound at 21-week gestation revealed a cystic and solid mass in the buttock, which looked like a type II sacrococcygeal teratoma. At 33-week gestation, a shunt operation was performed for decompressing the polyhydroamnios. The mass grew larger on a follow-up ultrasound.

The baby's serum α-fetoprotein (AFP) level was 143000 ng/mL and its β-human chorionic gonadotropin (hCG) level was 68 ug/L. Plain radiography showed a huge exophytic buttock mass with amorphous calcifications (Fig. 1A) and there were at least two long bones, vertebral bodies, and phalanges-like bones in the mass on the specimen radiography (Fig. 1B).

Ultrasound revealed a large sacrococcygeal mass with extension into lower abdomen. Long bones, vertebral column (Fig.

1C), and foot-like structures were found in the upper part of the mass. Wrinkled ribbon-like structures mimicking brain cortices (Fig. 1D) were found in the exophytic lower portion of the mass. MRI showed fat tissue around the bony structures in the presacral mass and brain-like solid tissue in the buttock mass (Fig. 1E). There were prominent left superior and inferior gluteal arteries, which were thought to feed the mass. The preoperative diagnosis was a fetus in fetu based on image findings.

Mass excision was done on the 6th day of birth. The whole mass was totally removed along with the coccyx. Macroscopically, the mass was composed of a large ovoid cystic portion and a lump of soft tissue with short dysmorphic extremities (Fig. 1F). On multiple sections, the large cystic portion contained hemorrhagic serous fluid and a lump of well-organized lobulating tissue that resembled the brain. The soft tissue component was mainly composed of fibroadipose tissues that had irregular shaped ossifying cartilages. On more detailed examinations, a bunch of entangled tubular structures resembling the intestines was identified.

On microscopic examination of the brain-like tissue, it was mainly composed of astrocytes, oligodendrocytes and a few neuronal cells admixed with the epithelial cells of the respiratory and gastrointestinal tracts. It was covered with highly vascular loose connective tissue reminiscent of leptomeninges. The cyst wall generally consisted of glial tissue, fibrous connective tissue resembling dura mater, and lining epithelium simulating

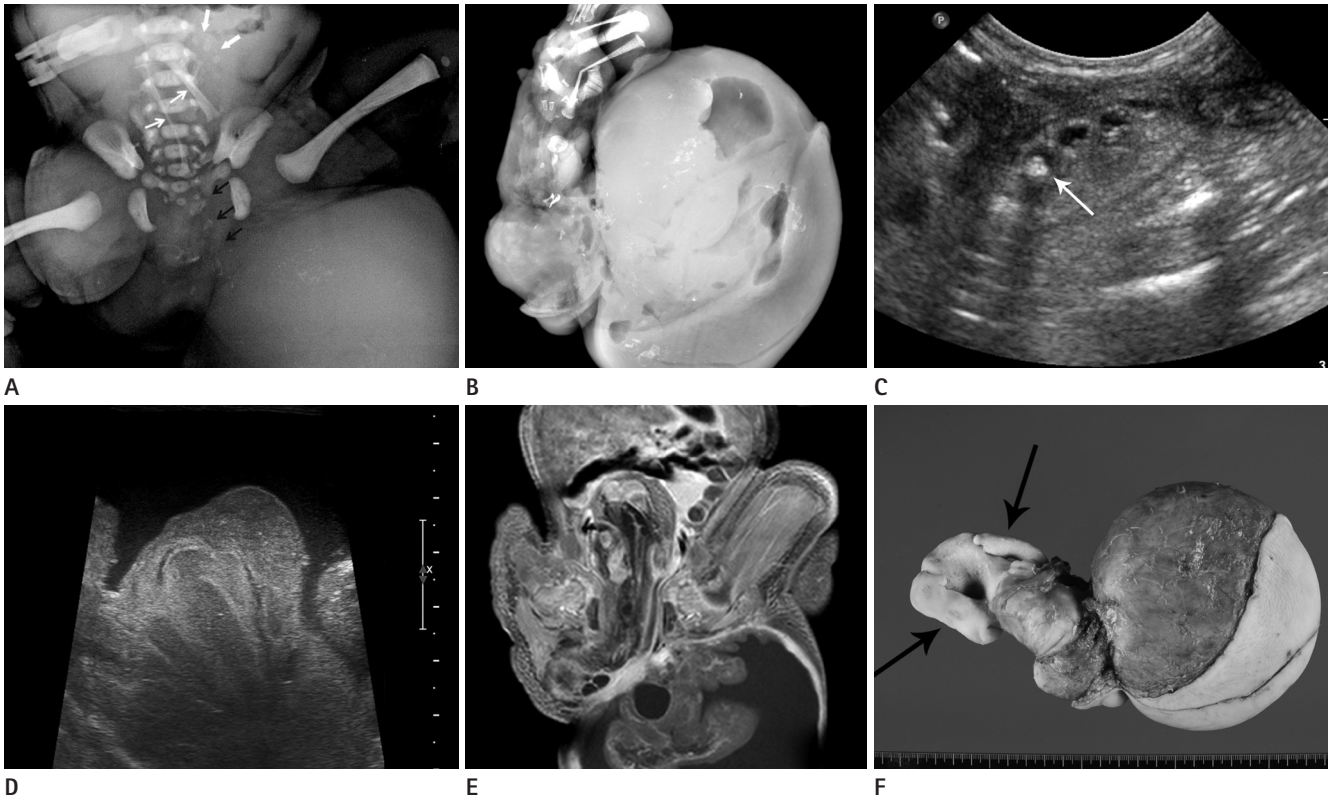

Fig. 1. A sacrococcygeal fetus in fetu in a newborn baby girl.
A. Abdominal radiograph shows a large buttock mass containing multiple amorphous calcifications. There are at least two long bones (thin white arrows), vertebral bodies (black arrows), and phalanges-like bones (thick white arrows).
B. Specimen radiograph of the excised mass reveals long bones representing the femur, tibia and fibula, short tubular bones representing the feet and hands, and a rounded ilium-like bone. The long bone was accidentally broken during manipulation of the mass.
C. The postnatal ultrasonogram shows small contiguous hypoechoic structures with a focal calcification (arrow), which looks like an underdeveloped vertebral column.
D. There are convoluted band-like structures representing the brain cortices with cystic areas, using a high resolution linear probe.
E. The T1 weighted gadolinium enhanced coronal image shows convoluted band-like structures with cystic areas in the buttock mass. It seems like brain tissue and a venticle.
F. Photographs of the gross specimen. There is a large, round head-like structure with two feet (arrows) on the other end. Feet are well developed and covered with skin.

ependyma and choroids plexus in some foci. The microscopic view of the soft tissue component showed multiple lineages of mature teratomatous tissues including skin, cartilage, bronchus, the region of the large intestine, and the pancreas. Further microscopic examinations revealed a focus of immature neuroepithelium, consistent with immature teratoma. Final pathologic diagnosis was fetus-in-fetu containing brain tissue, associated with immature teratoma.

The postoperative course was uneventful with decreased serum AFP and β-hCG levels.

DISCUSSION

Fetus in fetu is an extremely rare congenital anomaly and characterized by a parasitic monozygotic diamniotic twin: one installs and grows within the body of the other twin. To our knowledge, less than 100 cases have been reported since Wilis made the diagnostic criteria for the fetus in fetu in 1953 (1). To distinguish fetus in fetu from teratoma, Wilis defined the criteria that fetus in fetu should have the vertebral axis with surrounding organs and limbs (1). The presence of the vertebral column represents that the development of the imbedded fetus was advanced at least beyond the primitive streak stage for a notochord, which is a precursor of the vertebral column. However, the vertebral column for fetus in fetu could be underdeveloped or dysplastic, and in 9% of reported cases, there was no evidence of a vertebral axis even with a detailed pathological examination (2). Under this concept, Gonzalez-Crussi (2) proposed the modified diagnostic criteria that fetus in fetu could be defined as any structure of fetal form with a very high development of organogenesis or the presence of a vertebral axis. On the contrary, teratoma is simply an accumulation of pluripotential cells, which shows neither organogenesis nor the vertebral axis. In most cases from the literature, fetus in fetu occurs independently; but two to five fetuses in fetu cases have been reported within the literature (3-6).

The most common location of fetus in fetu is the abdominal cavity (7). It could be found anywhere in the body including the cranial cavity, oral cavity, mediastinum, lung, sacrococcygeal region, kidneys, and even the scrotum (7). On the contrary, teratomas are most commonly found in the lower retroperitoneum or sacrococcygeal region (3).

In our case, the sacrococcygeal location of the fetus in fetu might have lead us to think it was a sacrococcygeal teratoma, which is more commonly found in that location. After careful examination of the plain radiography, we could identify well formed long bones, vertebral columns and short tubular bones. Brain-like structures seen on postnatal ultrasound also suggested organogenesis within the mass, which could be a part of the fetus in fetu. In our patient, prenatal diagnosis was a type II sacrococcygeal teratoma because it might not be easy to demonstrate the spinal column or other bony structures on the fetal ultrasound.

Conventional radiographs can be very helpful in supporting the diagnosis by identifying a vertebral column as in our case (8). The CT scan is more useful to show osseous structures, especially in three dimensional reconstruction images but radiation could be problematic for newborn infants (9). We took specimen radiography using the mammography machine with low kVp and specimen CT in order to detect bony structures for the mass after it was surgically removed.

In our case, there was brain tissue with neuroglial cells within the fetus in fetu mass. According to Hoeffel et al. (6), different organs could be identified: vertebral column in 91%, limbs in 82.5%, central nervous system in 55.8%, gastrointestinal tract in 45%, vessels in 40%, and genitourinary tract in 26.5%. However, in most reported cases, the parasitic fetus was anencephalic with no brain tissue.

It has been rarely reported that the fetus in fetu is associated with the immature teratomatous component. Pourang et al. (3) reported twin fetuses in fetu with immature teratoma, but the immature teratoma was separated from the twin fetuses in fetu. The fetus in fetu has been known as a benign disease, but if there is an immature teratoma component as in our case, the possibility of a malignant transformation cannot be excluded.

Complete surgical excision is the treatment of choice and the prognosis is favorable compared to cystic teratoma (3). Nevertheless, close follow-up is mandatory because there are few chances of recurrence or malignant transformation (3).

In conclusion, we present a rare case of fetus in fetu with brain tissue containing an immature teratomatous component, mimicking sacrococcygeal teratoma in a newborn baby girl. Careful examination of plain radiography is important for the diagnosis of the fetus in fetu in order to guide further imaging studies by

identifying specific bony components within the mass.

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기형종과 유사하게 보이는 천미골 태아 내 태아(Fetus in Fetu) : 뇌조직과 미성숙기형종 요소를 포함한 1예 보고[1]

김채리[1] · 윤혜경[1] · 조영아[1] · 황재연[1] · 김애란[2] · 황희상[3] · 윤종현[1]

태아 내 태아(fetus in fetu)는 생존이 불가능한 기형적인 조직이 다른 태아의 몸 속에서 기생하는 상태를 말한다. 대부분이 무뇌 상태이며, 미성숙기형종 요소를 포함하는 경우는 매우 드물다. 저자들은 태아의 천미골 부위에 발생하여 천미골 기형종과 유사한 소견을 보였던 태아 내 태아 1예를 보고하고자 하며, 영상의학검사 및 병리조직 표본을 통해 뇌 조직과 함께 미성숙기형종 요소가 있었음을 확인하였다. 단순촬영을 포함한 영상검사가 정확한 진단을 하는 데 도움이 되었다.

울산대학교 의과대학 서울아산병원 [1]영상의학과, [2]신생아과, [3]병리과