

Fetus in fetu in the scrotal sac of newborn

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Fetus in fetu (FIF) in the scrotal sac is often overlooked in the differential diagnosis of other scrotal swellings in infants and children. Unlike teratoma, FIF is a benign disorder. Scrotal FIF is extraordinarily rare and has been reported only twice in the international literature. We describe the case of a 15-day-old neonate who presented with scrotal swelling since birth. Radiography and ultrasound revealed a FIF. On surgical removal and pathological evaluation, the anencephalic fetus had limb buds, male external genitals, and vertebral column, supporting the diagnosis of FIF. FIF in the scrotum is a very rare entity. It should be differentiated from other scrotal swellings such as teratoma because of the malignant potential of the latter. Preoperative diagnosis is

based on radiologic findings. The treatment is complete excision. *Ann Pediatr Surg* 12:68–70 © 2016 Annals of Pediatric Surgery.

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Introduction

Fetus in fetu (FIF) is characterized by enclosure of a malformed, monozygotic, diamniotic parasitic twin into the body of its normal host. It is an extremely rare condition. Of the 100 cases described in world literature, a majority occurs in the retroperitoneal space [1–3]. FIF in the scrotum is extraordinarily rare [4,5]. We describe third such case preoperatively diagnosed and successfully operated.

Case report

A 15-day-old male newborn presented with a left scrotal swelling noticed immediately after birth. The mother was a 26-year-old healthy second gravida with unremarkable family history of multiple births. The patient was delivered normally, weighed 2.5 kg, and was noted to have left scrotal swelling, which contained a 7 cm-diameter tense firm mass. The level of α -fetoprotein was 18000 IU/ml and β -human chorionic gonadotropin was normal. A plain radiograph of the pelvis showed bone densities resembling the spine, femur, and iliac bones within the enlarged left hemiscrotum (Fig. 1). The scrotal ultrasound showed a well-defined fetiform mass measuring 3 × 2 cm, which showed bone elements resembling vertebral axis and iliac bones surrounded by fluid-filled sac. Rudimentary lower extremity buds with identifiable femur bone and tiny male external genitalia were evident as a part of the mass. A tiny atrophied testis was visualized outer to the fluid-filled sac. The right hemiscrotum and right testis were unremarkable. Upon excision, the mass weighed 50 g and was enclosed by tunica albuginea-like membrane and could be easily separated from the scrotal wall. Bone structures were palpable in the mass. On incising, the mass had pinkish fetiform morphology, which at one pole had two limb-like projections and tiny male external genitalia. There were two thick-walled sacs, possible amnion, and testicular covering (Fig. 2a–c). Microscopic examination showed

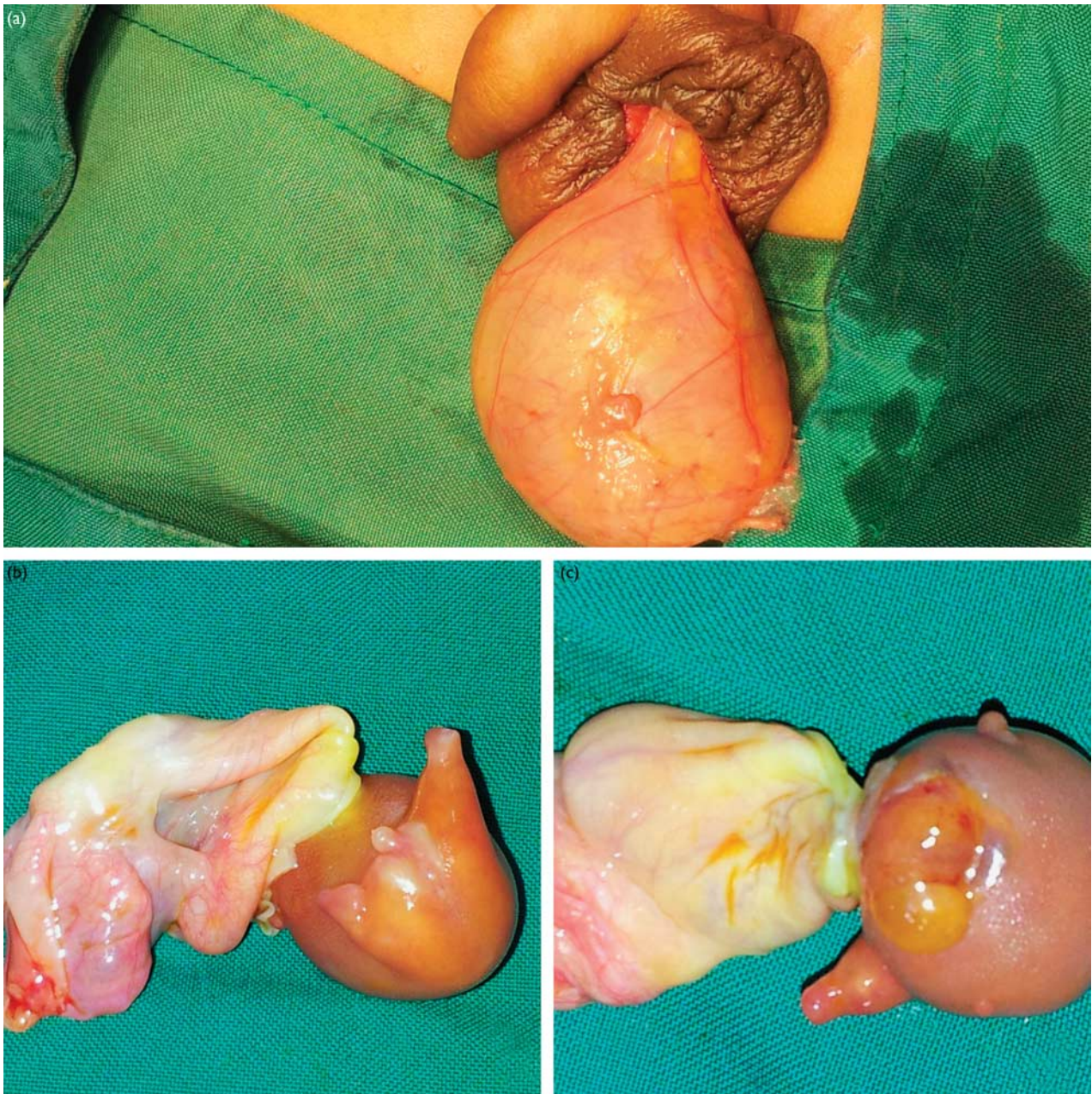
seven small spots of cartilage and bone in a line suggesting spine. These were surrounded by vascular connective tissue, skeletal muscle, and fat. Parallel to the spine, longitudinally sectioned intestinal wall was seen lined by mucosa. One pole of the mass revealed central nervous system tissue, and the other pole had a part of limb bones with soft tissue (Fig. 3a and b). The mass was covered with fetal type of skin with adenexal structures. A section from the sac showed a surface lined by cuboidal cells consistent with amnion. Another sac showed fibrous tissue and compressed infantile testicular tubules and germ cells. The patient recovered fully and the α -fetoprotein level was 6000 IU/ml postoperatively after a week.

Fig. 1



Plain radiography of the pelvis showing the vertebral column (arrow), femur, and iliac bones (arrow heads) in the left hemiscrotum.

Fig. 2



Surgical specimen showing (a) an oval mass covered with tunica in the left hemiscrotum (b) with two limb-like projections and tiny male genitalia. (c) Fetus in fetu covered with fetal skin and two thick-walled sacs, amnion, and testicular coverings.

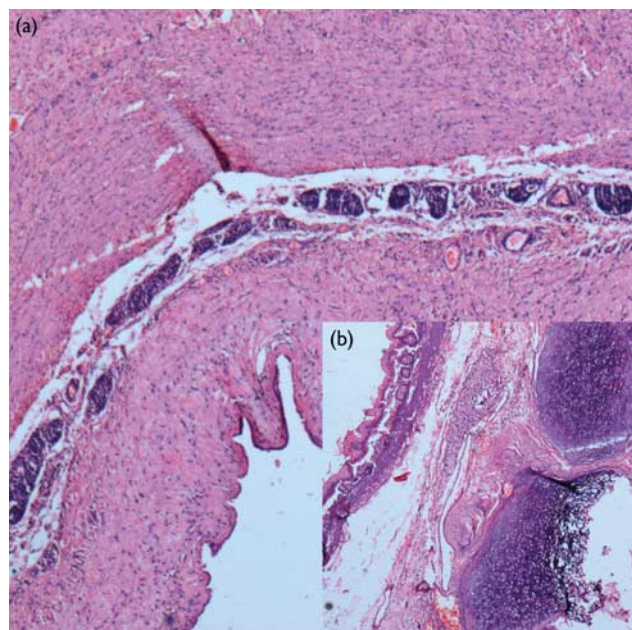
Discussion

First described by Johann Friedrich Meckel in 1800, FIF was defined by Willis [1] in 1953 as a rare condition in which a malformed parasitic twin gets enveloped in the body of its host twin. With an overall incidence of one in 500 000 live births, the majority of cases are found in infants and children [1,2,5,6]. FIF is anencephalic and acardiac and commonly located in the retroperitoneum. Rarer locations include the cranium, liver, and pelvis [2]. FIF in the scrotum is extraordinarily rare, with only two cases reported in the international literature. In 1972, Kakizoe and Tahara described a FIF in the scrotal sac of a Japanese infant and in 1999, Shin *et al.* described a similar case in a South Korean infant [4,5]. This unusual location

suggests migration of the fetus from the retroperitoneal cavity along with the normal descent of the testis to the scrotal sac through tunica vaginalis [6].

Spencer [7] suggested that a FIF must have one or more of the following conditions: (a) be enclosed within a distinct sac; (b) be partially or completely covered with normal skin; (c) have grossly recognizable anatomic parts; (d) be attached to the host by only a few relatively large blood vessels; or (e) either be located immediately adjacent to one of the sites of attachment of conjoined twins or be associated with the neural tube or the gastrointestinal system. The presence of vertebral bodies not only means that the FIF passed the primary stage of

Fig. 3



A microphotograph (a) showing testis with dark staining compressed seminiferous tubules in curved line and (b) two cartilaginous vertebrae with surrounding tissue and longitudinal section of intestine.

gastrulation but may also reflect its derivation from a primitive streak. The formation of the primitive streak normally starts during the third week, together with gastrulation that will lead to the notochord formation and subsequently to the vertebral column and segmental axis. Therefore, FIF likely arises from a zygote at a primitive-streak stage and fetiform mass develops to a certain degree in a manner similar to normal fetal development and lies close to midline commonly in the retroperitoneum [8,9]. Most of the investigators agree that the presence of the elements of the axial skeleton (vertebrae) is necessary to diagnose FIF [10,11]. Despite the fact that not more than 100 cases of FIF have been reported in the literature, the presence of a well-formed vertebral column is not documented in many cases [3]

Pathogenesis of FIF is believed to be a result of asymmetric division of totipotential cells in blastocyst or zygote in the early stage of development. As a consequence, the monozygotic twin is incorporated into his partner through anastomosis of vitelline circula-

tion [6]. Thus, it is well differentiated and encapsulated by an amnion-like membrane. Molecular analysis using an informative genetic marker for uniparental isodisomy of chromosomes 14 and 15 showed no genetic difference between the host infant and the fetiform mass [10]. Although it is a relatively benign condition, clinicians need to have a high index of suspicion and should not overlook the differential diagnosis of other scrotal swellings. Teratoma is the result of accumulation of pluripotent cells without features of organogenesis or vertebral segmentation and has a tendency to develop into mature tissue with malignant potential [10,11]. Presence of vertebral column often with appropriate arrangement of other organs or limbs around this axis differentiates FIF from teratoma.

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Conflicts of interest

There are no conflicts of interest.

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