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Case Report

Lumbar fetiform teratoma; a case report

Fazel Rahman Faizi, MD*, Najibullah Rasouly, MD, MS

Radiology department, French Medical Institute for Mothers and Children (FMIC), Kabul, Afghanistan

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ABSTRACT

Fetiform teratoma is a rare highly developed mature teratoma with organoid differentiation although it is not as developed as fetus in fetus which is the only differential diagnosis of this entity. It should be distinguished from the fetus in fetu by absence of the axial skeleton. A 6-month-old baby girl with a lumbar lump underwent ultrasonography, CT-scan and surgery. On imaging, a sac-like structure was noted in the lumbar region containing bowel segments, mesentery-like structure and few lymph nodes. The patient underwent surgery. The entire sac with its content were completely excised. Very few cases of fetiform teratoma have been reported in English literature however to the best of our knowledge lumbar fetiform teratoma has not been reported up to now. Highly developed teratoma is a diagnostic dilemma as it resembles fetus in fetu. The absence of a spinal axis differentiates this entity from the fetus in fetu.

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Background

Sacrococcygeal teratoma is a rare neoplasm with an incidence of 1 in 40,000 newborns. It is divided into 3 categories; classical teratoma, fetiform teratoma, and malignant teratoma. Occasionally teratoma may develop high grade of differentiation and organization resembling fetiform structure [1,2]. The only differential diagnosis for highly developed teratoma is fetus in fetu, an extremely rare entity with an incidence of 1 in 500,000 newborns. It is differentiated from fetiform teratoma by the presence of an axial skeleton. From the point of embryology, fetus in fetu goes through stages of primitive streak that will result in vertebral formation. This process is considered too advanced for fetiform teratoma to develop [2,3].

Case report

A 6-month-old baby girl with a slowly growing mass in the lumbar region (Fig. 1) was referred to the radiology department of French Medical Institute for Mothers and Children. The mass was soft and nontender. No neurological disorder was reported by the referring clinician. A small aperture was noted separately and inferiorly from the lump near the gluteal cleft. The mother stated normal vaginal delivery. No prenatal ultrasound was performed.

On ultrasonography, a sac-like structure was noted in the lumbar region containing hypoechoic tubular structures with perceptible walls in keeping with bowel segments (Fig. 2). There were multiple hypoechoic lobulated structures within the sac representing conglomerated lymph nodes (Fig. 3). No

^{*} Corresponding author. E-mail address: fazelf2016@gmail.com (F.R. Faizi).



Fig. 1 – A large lump is noted at the lumbar region. It was mobile and nontender. An aperture is noted below the mass near the gluteal cleft.

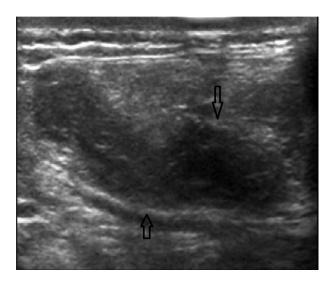


Fig. 2 – Ultrasonography with linear transducer reveals a hypoechoic tubular structure with a perceptible walls (arrows) in keeping with bowel loops.

echogenic focus with posterior acoustic shadowing was noted inside the sac to suggest calcifications or bony components.

CT of the spine was advised for further clarification and evaluation of the intraspinal extension of the lesion. It revealed a well-defined sac in the lumbar region containing bowel segments, mesentery-like structure, and lymph nodes. No intraspinal or intra-abdominal communication was detected (Figs. 4 and 5). The associated finding was spina bifida at L5/S1 level with a fluid-attenuated sac extruding through the bony defect posteriorly in keeping with meningocele (Fig. 6). The differential diagnosis is neuroenteric cyst however it is usually located in the ventral aspect of the dorsal spine and seldom associated with spina bifida. Widening of

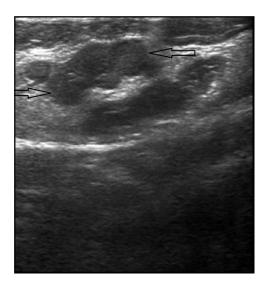


Fig. 3 – Ultrasonography with linear transducer also demonstrates a hypoechoic lobulated structure within the mass representing conglomerated lymph nodes (arrows).

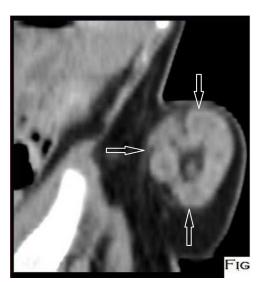


Fig. 4 – Postcontrast selected sagittal image from the lumbar region demonstrates a sac-like structure containing bowel loops (arrows). No intraspinal or intraabdominal communication is noted.

the spinal canal was also noted (Fig. 7). No sign of Chiari II malformation was noted on subsequent brain ultrasound.

The patient underwent surgery. Surgical exploration revealed bowel segments attached to a mesentery-like structure originating from the teratoid tissue (Figs. 8 and 9). The entire sac, contents and overlying rudimentary skin tissue were completely excised (Figs. 10 and 11). Meanwhile, meningocele which was located inferiorly and separately from the teratoma was also repaired.

Histopathology is recommended whenever there is an unrecognized or underdeveloped structure within the *fetiform teratoma*. In this case scenario, highly developed and organized

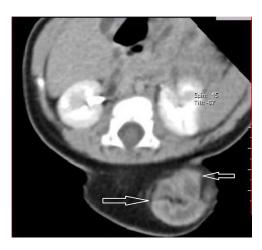


Fig. 5 – Postcontrast selected axial image from the lumbar region demonstrates a sac-like structure containing bowel loops (arrows). No intraspinal or intra-abdominal communication is noted.



Fig. 6 – Postcontrast selected axial image at a lower level demonstrates spina bifida with dural sac extruding through bony defect posteriorly in keeping with meningocele (arrow).

bowel loops with multiple lymph nodes were seen attached to a mesentery-like structure that originated from the teratoid tissue w. Therefore, histopathology workup was not recommended.

Discussion

Teratoma is a type of neoplasm containing 1 or more germ cell layers. It can affect individuals at any age, any location

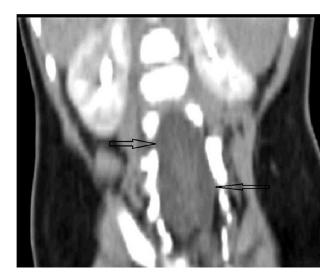


Fig. 7 – Postcontrast selected coronal image from the lumbar region demonstrates widening of the spinal canal (arrows).

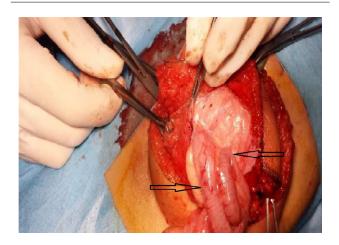


Fig. 8 – After excision of the skin, bowel segments were seen attached to a mesentery-like structure originating from the teratoid tissue (arrows).

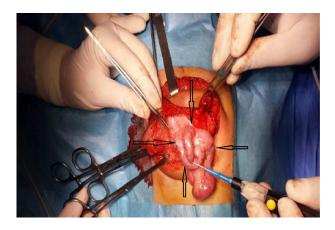


Fig. 9 – After excision of the skin, bowel segments were seen attached to a mesentery-like structure originating from the teratoid tissue (arrows).



Fig. 10 – The entire sac along with its content and overlying rudimentary skin were completely excised.



Fig. 11 – The entire sac along with its content and overlying rudimentary skin were completely excised.

with variable clinical presentation. Sacrococcygeal teratoma is the most common teratoma in neonatal and infancy period [4]. It is classified in 3 categories; classical type, fetiform teratoma, and malignant teratoma. On rare occasion, teratoma demonstrates high degree of differentiation and organization resembling malformed fetus, so called fetiform teratoma [1,2]. As in our case, highly organized bowel loops with lymph nodes was attached to mesentery-like structure in the lumbar region. This entity needs to be distinguished from the fetus in fetu.

Fetus in fetu is a vertebrate fetus enclosed in the body of normally developing fetus. Initially, the growth of fetus in fetu is comparable in the uterus with its twin however, it is halted due to vascular dominance of the host twin or inherent defect of the parasitic twin. Some authors suggest that teratoma and fetus in fetu are part of the same spectrum however, others believe that fetus in fetu is a distinct entity that is distinguished from the teratoma by the presence of vertebral column. The notochord of fetus in fetu goes through stages of primitive streak beyond the formation of vertebral column. In 9 % of the fetus in fetu, vertebral axis cannot be detected on imaging this is likely that spinal cord and vertebral bodies cannot be visualized in underdeveloped or dysplastic spinal cord [5].

Radiography, ultrasonography, CT-scan and MRI may be used for evaluation of these entities however CT-scan is a valuable test in diagnosis and pre-operative planning of the lesion particularly among pediatric population. It gives detailed anatomy of the tumor and extension of the lesion with adjacent structures [1,5]. Complete surgical excision is the definitive treatment of this entity [1].

Conclusion

Fetiform teratoma is a rare mature teratoma with high degree of differentiation and organization. It is a diagnostic challenge, as it resembles fetus in fetu. The absence of vertebral axis differentiates this entity from the fetus in fetu.

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