Case Report

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Fetal Vesicoallantoic Cyst and Intraabdominal Defects: An Unusual Case and Review of the Literature

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Abstract

We present an unusual and, until now, unreported case of vesicoallantoic cyst associated with multiple malformations in a fetus. A differential diagnosis is discussed, including the hypothesis of a genetic disorder.

Keywords: Fetus; Prenatal diagnosis; Vesicoallantoic cyst

Introduction

Allantoic cysts are rare fetal conditions. Their etiology is unclear but seems to be related to a persistent communication between the bladder and the umbilical cord. The urachus, an embryologic fibrous remnant of the allantois, could remain patent, resulting in a vesico-allantoic cyst.[1] The incidence of a patent urachus is around 3 for 1,000,000 births with a higher prevalence in male fetuses.[1] Allantoic cysts are generally isolated but have been reported to be associated with aneuploidies[2] and other defects such as Vacterl association[2] and posterior urethral valve.[3]

Here we report a case of fetal vesico-allantoic cyst associated with a complex multiple defects syndrome including anal imperforation, left colon atresia, rectum connection into the umbilical cord. In addition, there was uterine agenesis and a right iliac artery headed into the umbilical cord. At the best of our knowledge, this complex syndrome has not yet been reported previously in the literature and a differential diagnosis is discussed. After a discussion of the results, the patient gave us her consent for publication.

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Case presentation

A 33-year-old (gravida 4, para 1), Caucasian woman was referred at 11 week's gestation for an abnormal abdominal mass. The patient had a personal medical history of congenital urogenital malformations including anal imperforation, recto-vaginal fistula, vesicoureteral reflux with ureteral duplication, urethral stenosis, and neurogenic bladder requiring serial catheterization.

Ultrasound examination at 13^{+6} week's gestation showed a 35 mm × 30 mm diameter hypoechogenic mass, in close proximity to the abdominal insertion of the umbilical cord. Umbilical vessels identified using Color Doppler were located around the cyst, (Fig. 1A).

At 15^{+1} week's gestation, the cyst was enlarged and connected with the bladder at ultrasound examination (Fig. 1B). Amniocentesis was performed to exclude aneuploidy and a normal 46, XX karyotype was found. Cystic cavity, measuring 33 mm × 7 mm, was sampled and fluid analysis was compatible with fetal urine with a beta 2-microglobuline concentration higher than 4 mg/L and normal natrium (128 mmol/L) and calcium (9.1 mg/dL).

At 17^{+2} week's gestation, the eutrophic fetus had normal kidneys but a poorly visualized bladder. The cyst was enlarged, measuring 37 mm × 60 mm. Amniotic fluid index was low, estimated to 30 mm.

At 19^{+1} week's gestation, an intrauterine fetal death was diagnosed and induction of labor started. Pathological examination of the fetus showed measurements matching for 17 weeks. Multiple congenital anomalies were found including one low-set left ear, an umbilical cord cyst communicating with the bladder. There was anal imperforation, a left colonic atresia with sigmoid and rectal dilatation. Both bladder and rectum ended inside the umbilical cord. A cloaca could not be ruled out as an uterine agenesis was found. Finally, the right iliac artery was not connected to the lower limb but into the umbilical cord. Microscopic examination was not contributive because of an advanced degree of autolysis.

Discussion

During pregnancy, the incidence of umbilical cord cysts ranges between 0.4% and 3.4%.[2–11] he presence of multiple cord cysts could be related to aneuploidy while

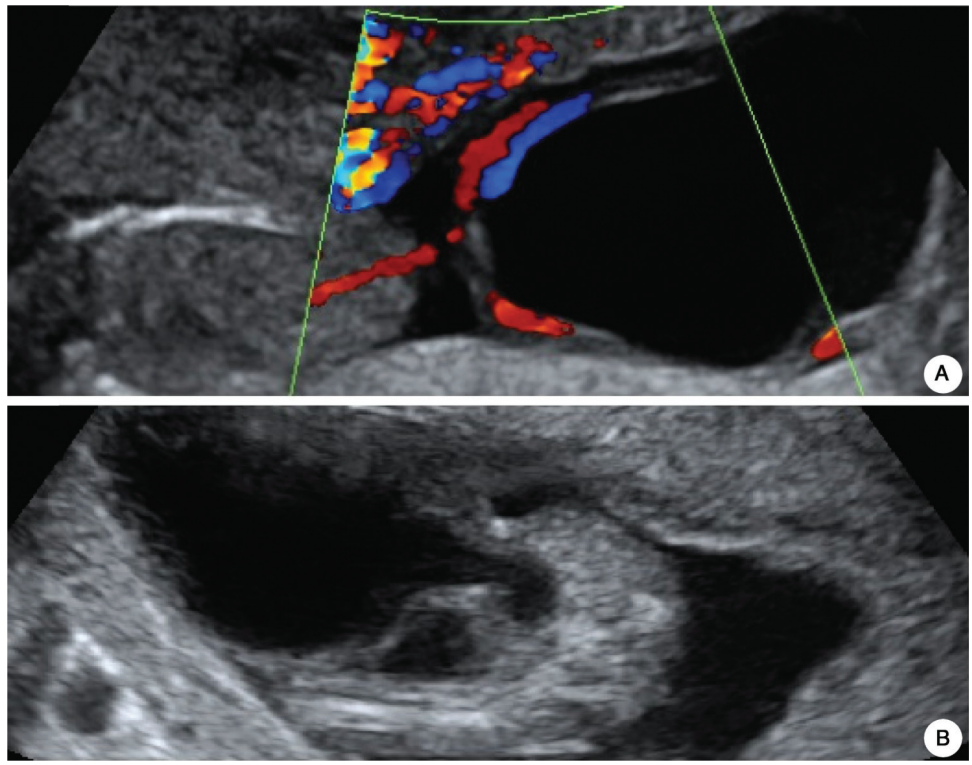

Figure 1. Ultrasound imaging of the pregnant woman. A Umbilical vessels identified using Color Doppler located around the cyst. B Connection between the cyst and the bladder at 15^{+1} week.

isolated cysts diagnosed at the first trimester are generally transient and do not interfere with the pregnancy outcome.[3] Umbilical cord cysts may be either pseudocysts or true cysts.

Pseudocysts are rare and carry a poor prognosis due to their association with fetal chromosomal defects, mainly trisomy 13 and 18 and with congenital anomalies such as omphalocele.[3,4]

True cysts originate from embryonic remnants such as the allantois or the omphalo-mesenteric duct. They are epithelial lined and are located nearby the fetal insertion of the umbilical cord.[3] A failure of the omphalo-mesenteric duct obliteration can lead to umbilical-enteric fistula, umbilical polyp, Meckel diverticulum or umbilical omphalocele.[1] Allantoic cysts are associated with urachal anomalies responsible for persistent communication between the bladder and the umbilical cord.[3] Vesicoallantoic cysts may be isolated associated with omphalocele,[3] pelvic organ prolapse,[4] obstructive uropathy,[2] and in one case, with multiple congenital anomalies including single heart ventricle, hydronephrosis, anal atresia, and club foot.[5]

There is no similar cases to ours in the literature. The association of a large bladder with an umbilical cord cyst at the end of the first trimester has been reported featuring an urachal abnormality.[6] Yoo *et al.* described a case of vesicoallantoic cyst with a spontaneous drainage, suggesting either a cyst rupture or a persisting communication between the cyst and the bladder.[7] In our patient, bladder was poorly seen at 16 weeks.

A familial genetic cause was suspected on the basis of the maternal medical history and the presence of a presacral tumor in the maternal grandmother. Considering these data, a Currarino triad was discussed. This syndrome, defined as an association of presacral mass, sacrum abnormality, and anorectal malformation, had an autosomal dominant pattern of inheritance with a reduced penetrance and a variable expressivity. Mutations in the homeobox gene *HLXB9*, located in 7q36, were identified in almost all cases of familial Currarino triad.[8] A sequencing study of the gene was performed but no mutation was identified in this family.

Differential diagnosis for this fetus include VACTERL association as it has been already reported to be associated with a vesicoallantoid cyst.[9] Fraser syndrome, a rare autosomal recessive disorder characterized by cryptoph-thalmos, syndactyly, and abnormal genitalia was excluded in the absence of eyes and extremities abnormalities.[10] The outcome of a prenatal vesicoallantoic cyst may vary from patent urachus and bladder dystrophy.[11]

In conclusion, this antenatal observation associated with a specific family history could not be related to any already published syndrome nor explained by cytogenetic and molecular biology existing analyses. We emphasize that a careful ultrasound examination should be carried out in all cases of umbilical cord cysts in order to exclude associated anomalies. In our patient, abnormal iliac artery insertion and complex genitourinary malformations could not be evidenced during the early second trimester by US. Genetic counseling is an essential step aiming to exclude rare syndromes such as Currarino triad and Fraser syndrome.

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Author Contributions

Mathieu Delvaux and Corinne Hubinont did the clinical follow-up of the case report, collected the data, and wrote the paper; Yves Sznajer participatedinthe management of the case and review of the literature; Etienne Marbaix did all the pathological analysis and reviewed the paper; Jean Marc Biard did the ultrasound clinical follow up and reviewed the paper.

Conflicts of Interest

None.

Editor Note

Corinne Hubinont is an Editorial Board Member of *Maternal Fetal Medicine*. The article was subject to the journal's standard procedures, with peer review handled independently of this editor and their research groups.

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