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Patent urachus or bladder exstrophy occulta? A case of prenatally disappeared umbilical cord cyst

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ABSTRACT

We are reporting an umbilical cyst detected at early trimester which mimicking bladder exstrophy occulta. A 3-cm umbilical cord cyst and a slight ventrally located urinary bladder beneath the cord insertion site was detected at 14th gestational weeks, which decreased in size and disappeared at 28th week. A term female neonate born with a 2-cm defect over the base of the umbilical cord, revealed a patent urachal fistula, and a part of the herniated urinary bladder. Detection of a vanished umbilical cord cyst has to keep aware of, making an immediate definite diagnosis and management of urachal anomaly.

Introduction

Cysts of the umbilical cord may persist through the entire pregnancy or may spontaneously disappear. These cysts can be simple or multiple, and are classified as true cysts with an epithelial lining and pseudocysts without epithelial lining depending on origin entity, but they have almost similar appearance on antenatal ultrasound, making challenges in prenatal differential diagnosis and consultation.[1,2] We present a case of patent urachus which mimicking bladder exstrophy occulta that showed a vanished umbilical cord cyst in late trimester as a sole abnormal prenatal ultrasound finding.

Case report

A 25-year-old patient was referred to our hospital at 14 weeks of gestation with a chief concern of a 3-cm cystic lesion near the base of the umbilical cord. The patient had no significant obstetric, medical, or family history. Detailed ultrasound examination showed a singleton fetus consistent with gestational age, normal appearing kidneys and urinary bladder, and normal amniotic fluid volume. She had a normal noninvasive prenatal screening test report for Down syndrome, and fetal karyotyping or MRI imaging were declined. A repeat ultrasound at 22 weeks of gestation showed that the mass was measuring approximately $48 \times 45\ mm^2$ in size (Fig. 1). The fetal abdomen appeared intact except for a slight ventrally located urinary bladder just beneath the cord insertion (Fig. 2). Both umbilical arteries entered normally in the fetal pelvis and ran alongside the fluid-filled fetal bladder. By the 28th week of gestation, the cord cyst decreased in size and disappeared. The patient went into spontaneous labor at 39 weeks of gestation and delivered a female infant weighing 2970 g. Immediately after birth, the insertion base of the umbilical cord in the neonate developed a slight budging peanut-size, pink-color mass was noted in a 2-cm defect. A patent urachal fistula was noted with continuous urine flow, and a part of the bladder herniated out of the fistula (Fig. 3).

Primary repair by excision of the urachal fistula and repair of the urinary bladder and abdominal wall defect with umbilicoplasty were successfully conducted. The neonate had no associated urogenital or skeletal anomalies. She has been regularly followed-up and developed well for a year after the repair surgery.

Discussion

An umbilical cord cyst can be diagnosed in the first trimester, with an incidence of approximately 1%–3%. If it persists during pregnancy, there is a possibility of developing associated malformations. The umbilical cord cyst shows a male predominance of 2:1.[1,2]

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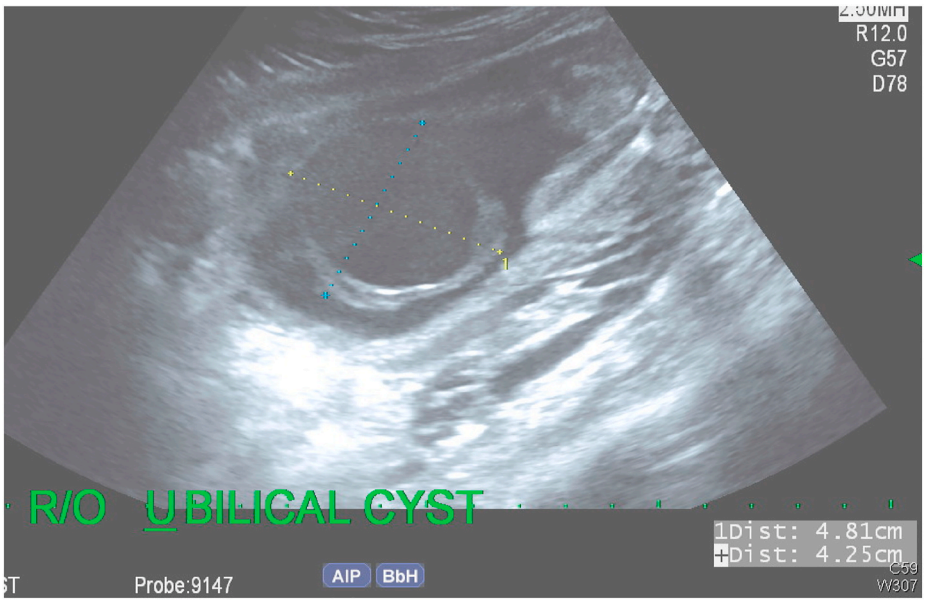

Fig. 1. Ultrasound at 22 weeks of gestation presented a 4.8 × 4.5 cm single umbilical cyst near the fetal pole.

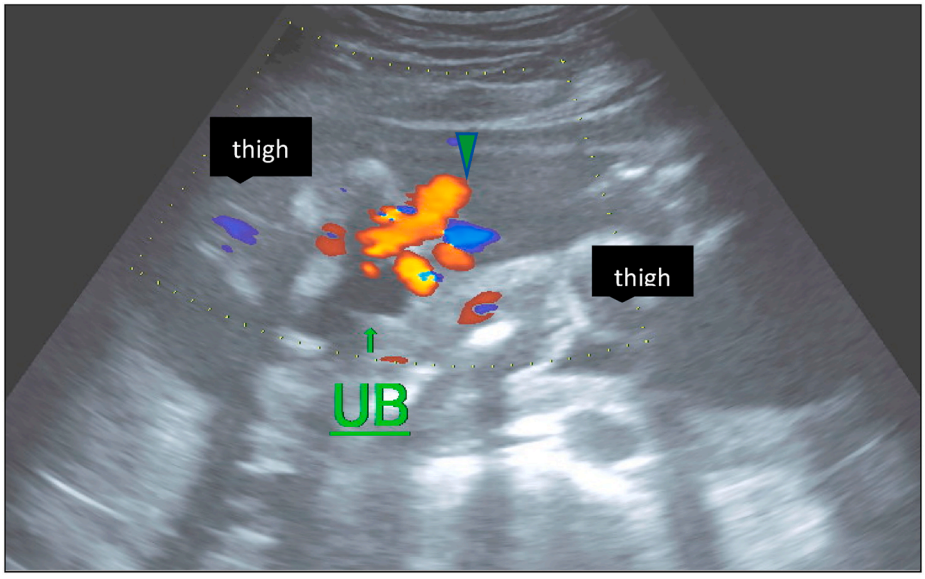

Fig. 2. Ventrally located urinary bladder (green arrow; UB: urinary bladder) beneath the umbilical cord insertion (arrow head) at 22 weeks of gestation. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

True umbilical cord cysts are derived from embryological remnants of either the allantois or omphalomesenteric duct, have no epithelial lining, and represent localized edema of the Wharton's jelly. Pseudocysts are more common than true cysts. Cases showing a high association between allantois cysts and omphalocele, between patent urachus and hydronephrosis, and among omphalomesenteric duct cysts have been reported. In addition, chromosomal or structural abnormalities have also been found.[1]

Umbilical cord cysts may persist through the entire pregnancy or may spontaneously disappear, as observed in our case report. As those detected in the second and third trimester are associated with fetal abdominal wall defects and chromosomal anomalies in up to 50% of cases.[1,2] Therefore, early karyotyping for the later cases may have a substantial impact on the management of pregnancy and delivery.

The urachus forms intraembryonic communication between the allantois and the cloaca, undergoes lumen obliteration in the fifth month of gestation because the bladder stretches on descending into the pelvis. Urachal remnants result from incomplete closure of this vesico-allantois channel (1). In this case, the ultrasound examination of the urinary bladder showed that the umbilical arteries ran alongside the fluid-filled fetal bladder throughout the prenatal period, which caused a misinterpretation of a simple umbilical cyst. After birth, the neonate had normal level of umbilical cord insertion but a 2-cm umbilical defect, with a small herniated mass having an everted urinary mucosal surface. The mass grew and further herniated, because of crying of the neonate, led to more urine leakage. The diagnosis of a patent urachus is confirmed.

Bladder exstrophy is a rare congenital anomaly, requiring prompt surgical intervention in a tertiary center after delivery. It occurs in 1 per 25000–50000 births with a 2:1 male to female ratio. Bladder exstrophy is easily recognizable at birth but is rarely diagnosed prenatally. The most common antenatal ultrasound finding suggesting bladder exstrophy is an absent fetal bladder, normal amount of amniotic fluid,

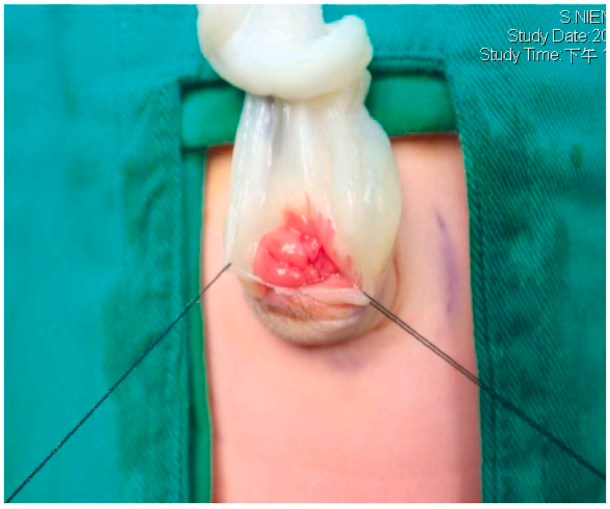

Fig. 3. A defect approximately 2-cm in size, with the urinary bladder herniated out of the fistula at the umbilicus. Urachal fistula is excised and the urinary bladder and the abdominal wall defect repaired.

normal appearing kidneys, with an anterior protruding mass at the lower fetal abdominal wall.[3,4] Some minor findings include low insertion of the umbilical cord, widening of the iliac crests, ambiguous external genitalia, and other skeletal anomalies. In occasional reports, the cystic mass in the lower fetal abdomen showed dynamic changes of filling and emptying, further demonstrating bladder exstrophy.[5] The initial diagnosis of exstrophy variants can be difficult because their presentation can be confusing (Fig. 2). The postnatal clinical features we presented were not compatible with bladder exstrophy occulta because the newborn had no pubic diastasis, short colon, vesico-colonic fistula, and other urologic or skeletal abnormalities.

Conclusions

In conclusion, when a diagnosis of umbilical cord cyst is made, detail ultrasound scans should be conducted until the time of delivery, despite the cord cyst spontaneously disappearing. Clinicians at perinatal care should manage carefully for detecting any associated rare diseases.

Authors' contributions

CWC and ASC: Management of the case and preparing the manuscript. ASC, KJC and JYL: Management of the case and critical appraisal and review of the manuscript. All authors read and approved the final manuscript.

Ethics approval and consent to participate

Ethics approval of this study was given by the Institutional Review Board of Chang Gung Memorial Hospital (202001829B0).

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Declaration of competing interest

The authors declare that there is no conflict of interest regarding the publication of this article.

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