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Allantoic Cyst Prior to Transient Megacystis in Patent Urachus: A Paradoxal Association?

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Abstract

Patent urachus is a rare umbilical anomaly with an incidence of 1 to 2.5 per 100,000 deliveries. The urachus is a thin fibromuscular tubular structure that represents the intra-abdominal portion of allantois extending from umbilicus to the vertex of the bladder. Under normal conditions, the urachus is not seen in utero, since the obliteration of its lumen usually occurs at six weeks of gestational age. An anomaly in allantois involution can result in patent urachus, umbilical urachal sinus, vesicourachal diverticulum, urachal cyst, or alternating sinus and should always be included in the differential diagnosis of midabdominal cysts. Before the advent of prenatal sonography, this condition was always diagnosed in early neonatal period. The authors report a rare case of a patent urachus suspected in the first trimester ultrasound.

Case Report

A 36 year old, primigravida, without medical relevant history, presented to our hospital for the first trimester sonographic screening at 13 weeks of gestational age. Transabdominal ultrasound revealed a singleton pregnancy, a fetus with a crown-rump length of 67 mm with normal nuchal translucency thickness and an anechoic proximal umbilical cord cyst close to the abdominal insertion (Figure 1). Color flow imaging showed both umbilical arteries surrounding the cyst (Figure 1), confirming that was an allantoic cyst. Two weeks later, a follow-up scan revealed megacystis with a longitudinal diameter of 22.6 mm, two umbilical cord cysts with 18.5 and 12.7 mm of diameter (Figure 2), and a communication between the proximal umbilical cyst and the bladder (Figure 2). Fetal karyotype was normal (46,XY) and no other anomalies were detected. All the findings and prognosis were explained to the parents who opted for a second opinion. Contradictory information was provided and they returned to our center for pregnancy termination. The good prognosis associated to the findings was once again explained and the couple decided to continue the pregnancy with follow up in our hospital. Subsequent ultrasound evaluations revealed an increase in size of the umbilical cord cysts at 16 weeks scan (28 and 27 mm), Wharton jelly edema and at 23 weeks, fetal bladder was not visualized, suggesting prolapsed bladder (Figure 3).

The pregnancy progressed normally with no further complications. A cesarean section delivery was performed at 38 weeks and male neonate weighing 3500 g with Apgar scores of 9 and 10 at 1 and 5 minutes, respectively, was delivered. Clinical evaluation of newborn was consistent with prenatal findings, except for the bladder prolapse. On day 2, laparotomic resection of the patent urachus was performed

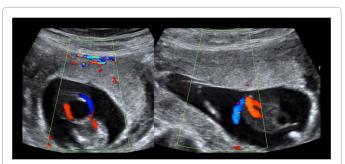


Figure 1: Allantoic cyst-umbilical cord cyst close to the fetal abdominal insertion at 13 weeks (left image). Visualization of two umbilical arteries with empty bladder (rightimage).

without complications. Follow-up was uneventful and the infant is now two-years-old.

Discussion

The authors report a case of patent urachus suspected in first trimester ultrasound, where the prominent finding was an umbilical cord cyst [1-5]. The prevalence, in the first trimester, is 0.4-3.4% and should lead to the suspicion of patent urachus [2,6]. Color flow imaging is used to identify the umbilical vessels surrounding the umbilical cyst, confirming that it is an allantoic cyst and not a pseudocyst [7,8]. Prenatal ultrasound findings of patent urachus consist in anechoic proximal cord cyst communicating with the bladder [5].

In this case, the characteristic findings were reported in the 15 weeks scan with an associated megacystis. Few cases have been reported of megacystis appearance after the umbilical cyst [6,9], what refutes the possible explanation of urachus patency has a result of megacystis [9]. Possibly the spontaneous resolution of megacystis is explain by the temporary malfunction of bladder smooth muscle due to autonomic innervation immaturity [10,11]. The evolution of a patent urachus may result in cyst rupture and/or bladder protrusion in the second trimester, an event which likely occurred in our patient [5].



Figure 2: Follow-up scan at 15 weeks. Bladder longitudinal diameter of 22.6 mm (left image). Umbilical cord cysts with 18.5 and 12.7 mm of diameter (middle image). Ultrasound shows the channel communicating from the umbilicus to the bladder on the right image.

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Figure 3: 23 weeks ultrasound. Bladder was not visible suggesting a prolapsed bladder

The differential diagnosis of this entity includes omphalocele and bladder extrophy. Karyotyping is not mandatory when this is an isolated finding, because of the lack of association between urachal anomalies and aneuploidy [12]. In this case, it was performed after parents' request.

Early postnatal surgery is usually required [5]. Laparoscopic surgical correction of patent urachus is the goldstandard but due to the lack of experience in our hospital, a laparotomic approach was performed. Both methods have equal efficiency, however laparoscopic approach is associated with less morbidity and short length of hospital stay.

In prenatal ultrasound evaluation, misdiagnosis or incorrect diagnosis can provide contradictory information and different prognosis to the parents.

Conclusion

The presence of an umbilical cord cyst in the first trimester is an ultrasound finding consistent with patent urachus. Evaluation of characteristic ultrasound findings associated with a patent urachus is important for adequate prenatal counseling. The differential diagnosis includes other fetal abdominal wall anomalies that can have severe implications in neonatal prognosis.

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