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A case of giant urachal cyst in a neonate☆

Soichi Nakada [a,*], Koji Kumori [a], Yoko Mizota [a], Shuichi Ishibashi [a], Seiji Yano [a], Showa Aoki [b], Yoshitsugu Tajima [a]

[a] Department of Digestive and General Surgery, Shimane University Faculty of Medicine, 89-1 Enya-cho, Izumo, Shimane 693-8501, Japan
[b] Department of Obstetrics and Gynecology, Matsue Red Cross Hospital, Shimane, Japan

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ABSTRACT

We report a case of giant urachal cyst in a neonate presenting characteristic features on fetal ultrasonography. A 28-year-old woman of 21 weeks gestation was referred to our hospital for evaluation of a cystic mass in the lower portion of the fetal abdomen. On fetal ultrasonography, at 23 weeks gestation, the two umbilical arteries in the base of the umbilical cord became separated from each other due to the fetal cyst. On fetal MRI, at 25 weeks gestation, the cyst was depicted as a unilocular serous cyst, independent of the surrounding organs such as the kidney, gallbladder and intestines. The cyst had no communication with the bladder or umbilical cord. A male infant was delivered at term. Although the infant was thriving and remained asymptomatic, an enhanced CT examination 13 days after birth showed the cyst was still present without any decrease in size. The infant underwent surgery to make a definite diagnosis and to prevent future complications. The cyst was excised without any complication and finally diagnosed as a urachal cyst based on morphological and pathological findings.

Recent advances in fetal ultrasound and magnetic resonance (MR) imaging studies have made it possible to detect a patent urachus prenatally, based on the presence of an allantoic cyst communicating with the bladder [1,2]. However, there have been few reports on prenatal detection of urachal anomalies other than patent urachi [3,4]. We present a case of giant urachal cyst in a neonate that presented characteristic findings on fetal ultrasonography.

1. Case report

A 28-year-old woman, gravida 0 para 0, at 21 weeks gestation was referred to our hospital for further examination of a cystic mass in the lower portion of the fetal abdomen detected by fetal ultrasonography. At 23 weeks gestation, a fetal ultrasonography demonstrated that the two umbilical arteries in the base of the umbilical cord became separated from each other due to the fetal cyst (Fig. 1). On fetal MRI, at 25 weeks gestation, the cyst was depicted as a unilocular serous cyst (Fig. 2). The cyst was clearly separated from the surrounding organs, such as the kidney, gallbladder and intestines, and showed no communication with the bladder or umbilical cord. Umbilical cord swelling was not detected, and no other abnormalities were seen.

At 40 weeks and 2 days of gestation, a 2956 g live male with Apgar scores of 8 and 8 at 1 and 5 min, respectively, was born by spontaneous vaginal delivery. On physical examination, the infant showed no signs of gastrointestinal obstruction or respiratory symptoms. Examination of the umbilicus was normal. There was a mobile, palpable mass in his lower medial abdomen. No other abnormal findings were noted. The infant had no clinical symptoms and was discharged from the hospital at 5 days after birth.

A follow-up contrast-enhanced CT examination, 13 days after birth, showed that the cystic mass was still present with no decrease in size. It was a unilocular cyst, 54 × 47 × 45 mm in size, without solid components or calcification (Fig. 3). Surgical resection was thus recommended to make a definite diagnosis and to prevent future complications.

At first, the infant underwent diagnostic laparoscopy following a percutaneous cyst puncture with aspiration of 60 ml of serous clear liquid under general anesthesia 15 days after birth. A 5-mm port was introduced into the lower medial abdomen using the open method. Laparoscopic exploration showed a cystic structure, possibly a urachal anomaly, in the lower medial portion of the peritoneal cavity that was attached to the underside of the anterior abdominal wall just below and caudal to the umbilicus. We thus

* Corresponding author. Tel.: +81 853 20 2232; fax: +81 853 20 2229.
E-mail addresses: soichin@med.shimane-u.ac.jp, soichinakada18@yahoo.co.jp (S. Nakada).

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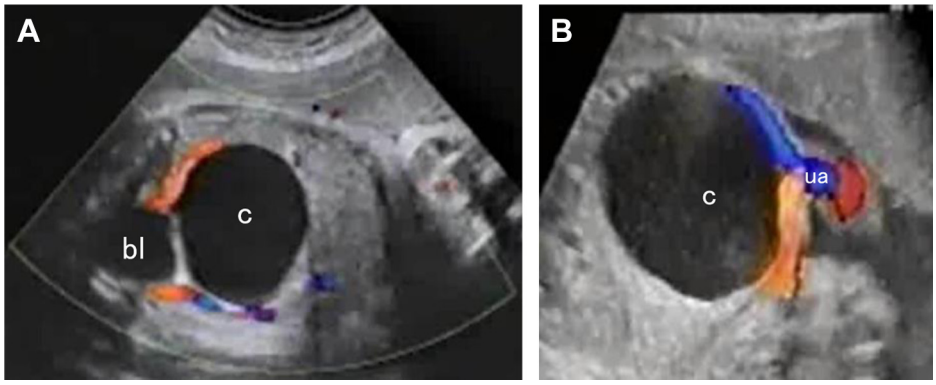

Fig. 1. Coronal color Doppler US images (A, B) demonstrate the cyst (c) separating the two umbilical arteries (ua) at the base of the umbilical cord. bl: bladder.

extended the wound to convert to a laparotomy and the cystic structure was excised. The cyst was connected to the dome of the bladder with a tiny fibrous strand, which had no luminal structure, and to the umbilicus with the median umbilical ligaments.

Histologically, the cystic wall was composed of fibrous tissue and partially covered with the urothelium, and was compatible with the diagnosis of a urachal cyst.

The patient's postoperative course was uneventful. The infant was discharged from the hospital on postoperative day 5 and he has been doing well without recurrence 18 months after surgery.

2. Discussion

Urachal anomalies are due to failure of complete obliteration of the urachus during gestation. The urachus is a fibrous remnant of the allantois. Between the 4th and 6th weeks of gestation, the allantois and the superior end of the presumptive bladder undergo regression. From the 4th to 5th months of gestation, the allantois and the constricted bladder apex become a narrow epithelial fibromuscular strand, i.e., the urachus or median umbilical ligament, which runs through the subperitoneal fat from the bladder to the umbilicus [5]. Incomplete obliteration of the urachal lumen results in four major types of abnormalities: patent urachus, urachal cyst, urachal sinus, and vesicourachal diverticulum. The most common urachal abnormalities are urachal cysts (45%), followed by urachal sinus (37%) and patent urachus (16%). Urachal diverticuli are extremely rare [6].

Some recent reports have described the utility of fetal ultrasonography in making a prenatal diagnosis of patent urachus, in which the detection of an allantoic cyst, a type of umbilical cord cyst, provides an important clue suggesting patent urachus. Characteristics of allantoic cysts are as follows: 1) the cyst is commonly detected in the proximal part of the umbilical cord, near its attachment to the anterior abdominal wall; 2) the cyst is usually found between the two umbilical arteries [7]. When the cyst communicates with the bladder, a diagnosis of patent urachus is made [1,2]. If the allantoic cyst suddenly disappears as gestation progresses, patent urachus with bladder prolapse is suspected [1].

On the other hand, characteristic findings of urachal cyst include a unilocular serous cyst located inside the lower middle portion of the anterior abdominal wall that is in intimate juxtaposition to the umbilical vein insertion superiorly and to the bladder inferiorly on

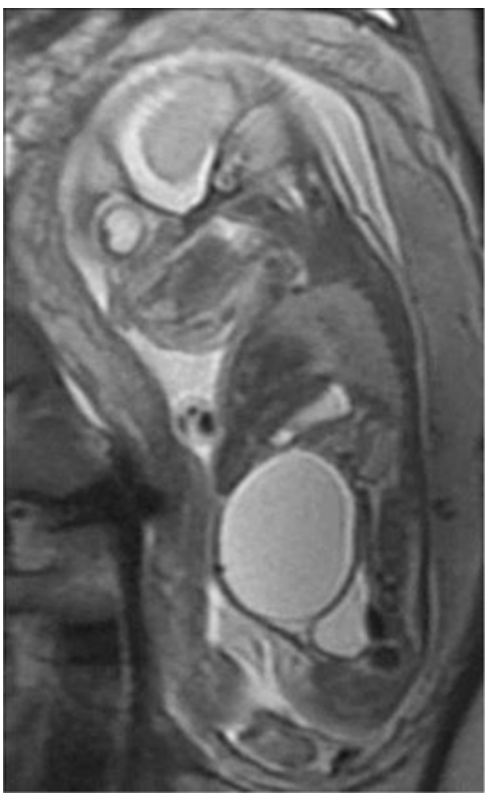

Fig. 2. A sagittal T2-W image demonstrates a unilocular serous cyst without any communication with the bladder or umbilical cord.

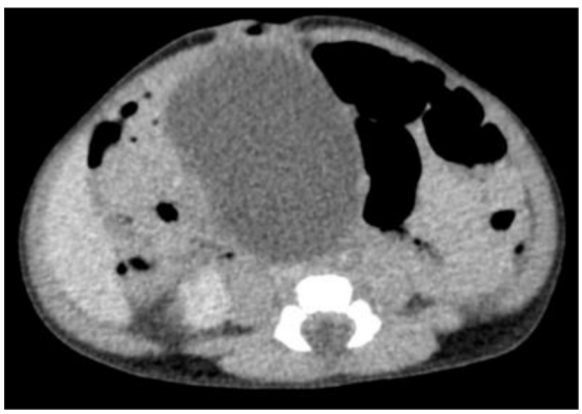

Fig. 3. A contrast-enhanced CT demonstrates a unilocular cyst, 54 × 47 × 45 mm in size, with no solid components or calcification.

fetal ultrasonography [3,4]. In the present case, fetal ultrasonography demonstrated a giant unilocular serous cyst located inside the lower middle portion of the anterior abdominal wall, in which the cyst separated the two umbilical arteries to the base of the umbilical cord. A unilocular cyst located between the two umbilical arteries looks like an allantoic cyst, which was seen in the present case. The location of the urachal cyst in our case was similar to those reported previously. This finding is ontogenetically consistent because the urachus is derived from the allantois. On the other hand, lower abdominal cystic masses, except for urachal cysts, may not separate the two umbilical arteries because they instead push these vessels forward from the rear. When an abdominal cystic mass is detected on fetal ultrasonography, confirmation of the positional relation between the cystic mass and the umbilical arteries is therefore useful to distinguish a urachal cyst from other cystic lesions.

The treatment of incidental urachal cysts remains controversial. Galanti et al. stated that spontaneous resolution of urachal remnants is possible in patients younger than 6 months of age, whereas those with symptoms or failure in resolution after 6 months should undergo urachal remnant excision [8]. On the other hand, any urachal cyst, even if incidental or asymptomatic, should be removed as soon as possible after achieving an accurate diagnosis because infected urachal cysts can cause peritonitis [9].

3. Conclusion

A fetal ultrasonography demonstrating a unilocular serous cyst that separates the two umbilical arteries may be an important clue to the diagnosis of a urachal cyst. Prenatal diagnosis may allow appropriate counseling and planned neonatal surgery.

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