Urology Case Reports 49 (2023) 102440

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr

Pediatrics

Prenatally diagnosed giant umbilical cord resulted from patent urachus in a monochorionic diamniotic twin with selective intrauterine growth restriction

Satoko Matsuyama, Fumi Matsumoto *, Futoshi Matsui, Koji Yazawa

Department of Urology, Osaka Women's and Children's Hospital, Osaka, Japan

ARTICLE INFO

Keywords:
Giant umbilical cord
Patent urachus
Prenatal diagnosis
Monochorionic diamniotic twins

ABSTRACT

A long diffuse giant umbilical cord (GUC), caused by umbilical cord edema associated with a patent urachus, is an extremely rare anomaly. While patients with diffuse GUC appear to experience no significant complications and a good prognosis, little is known about their etiology and prenatal course. Here, we report the first case of prenatally diagnosed diffuse GUC resulted from patent urachus in a monochorionic diamniotic twin with selective intrauterine growth restriction. This case indicates that GUC is epigenetic and unrelated to multiple births.

1. Introduction

A giant umbilical cord (GUC), defined as a cord diameter of >5cm, is a rare anomaly. It may present as either localized or diffuse enlargement of the umbilical cord. Differential diagnosis of a localized GUC includes various conditions, such as umbilical pseudocysts, umbilical hernias, omphalo-mesentric duct cysts, vascular anomalies, abdominal wall defect, bladder/cloaca exstrophy, and urachal anomalies. On the other hand, a diffuse GUC is uniformly caused by umbilical cord edema associated with patent urachus.[1] Through an open urachus, retrograde voided hypotonic fetal urine is absorbed into the Wharton's jelly, resulting in massive swelling of the umbilical cord. Although patients with diffuse GUC appear to experience no significant complications and a good prognosis, they are limited to scattered case reports. Herein, we report a case of prenatally diagnosed diffuse GUC resulted from patent urachus in a monochorionic diamniotic (MD) twin with selective intrauterine growth restriction (sIUGR). Discordant GUC in MD twins has not been reported previously.

2. Case presentation

A 26-year-old primigravida was referred to our hospital for a survey at 24 weeks of gestation because of a twin pregnancy with discordant fetal growth and anomalies. Her medical and obstetric history was unremarkable. Discordant fetal growth of MD twins was first detected by ultrasonography at 22 weeks of gestation. At referral, the estimated fetal weight (EFW) of the smaller twin was 530g (−1.55 SD), and the discordant score was 31%. Doppler sonography revealed that the end-diastolic flow of the umbilical artery was positive in both twins.

Ultrasonography revealed an umbilical cord cyst close to the fetal abdominal insertion of the smaller twin at 23 weeks of gestation (Fig. 1A). The umbilical cord cyst first grew to 40.1 × 49.7mm at 30 weeks of gestation (Fig. 1B), and spontaneously shrank to 19 × 14mm at 33 weeks of gestation (Fig. 1C). Subsequently, the umbilical cord diameter of the smaller twin gradually increased. Doppler sonography revealed an intermittent absence of end-diastolic velocity in the umbilical artery of the smaller twin. As an enlarged heart and tricuspid regurgitation were observed in the other twin, the smaller twin was delivered first by scheduled cesarean section at 34 weeks and 0 days of gestation. Her birth weight was 1,522g (−1.7 SD). Apgar scores were 8 and 9 after 1 and 5 min, respectively. The umbilical cord was entirely edematous and swollen, with a length of >39cm and a maximum diameter of 9 cm (Fig. 2A). A small reddish mass, suspected to be prolapsed urachal mucosa, was noted at the base of the umbilical cord (Fig. 2B). The creatinine level, measured by puncture of Wharton's jelly in the umbilical cord was 0.8mg/dl. The diameter of the umbilical cord of the second largest twin was 2cm, within the normal range. Surgical repair was performed on the first day of life. Under general anesthesia, the umbilical cord was removed after ligation of the 3 umbilical vessels surrounded by edematous Wharton's jelly. The prolapsed urachal

* Corresponding author. Department of Urology, Osaka Women's and Children's Hospital, 840 Murodocho, Izumi, Osaka, 594-1101, Japan.
E-mail address: fumim@wch.opho.jp (F. Matsumoto).

https://doi.org/10.1016/j.eucr.2023.102440
Received 20 March 2023; Received in revised form 10 May 2023; Accepted 13 May 2023
Available online 20 May 2023

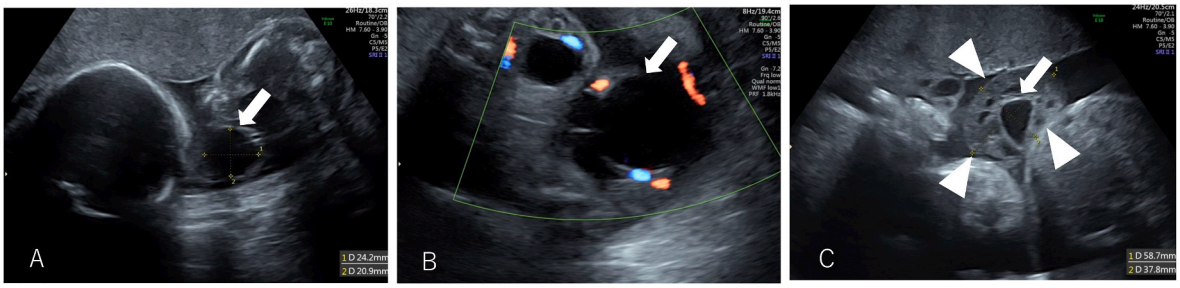

Fig. 1. Ultrasonography revealed an umbilical cord cyst (arrows) close to the fetal abdominal insertion of the smaller twin at 23 weeks of gestation (A). The umbilical cord cyst grew to 40.1 × 49.7mm at 30 weeks of gestation (B), and spontaneously shrank to 19 × 14mm at 33 weeks of gestation. Subsequently, the diameter of the umbilical cord (arrowheads) gradually increased (C).

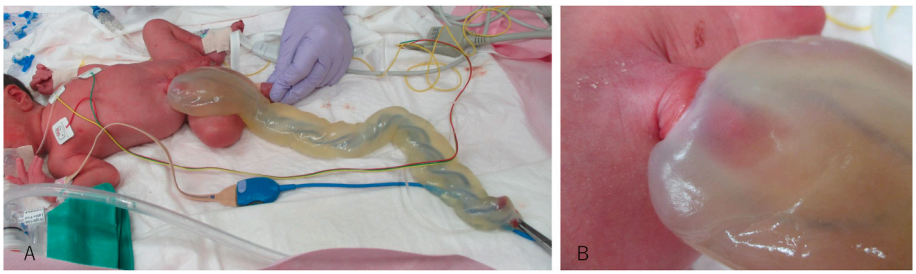

Fig. 2. The umbilical cord was entirely edematous and swollen, with a length of >39cm and a maximum diameter of 9 cm (A). A small reddish mass, suspected to be prolapsed urachal mucosa, was noted at the base of the umbilical cord (B).

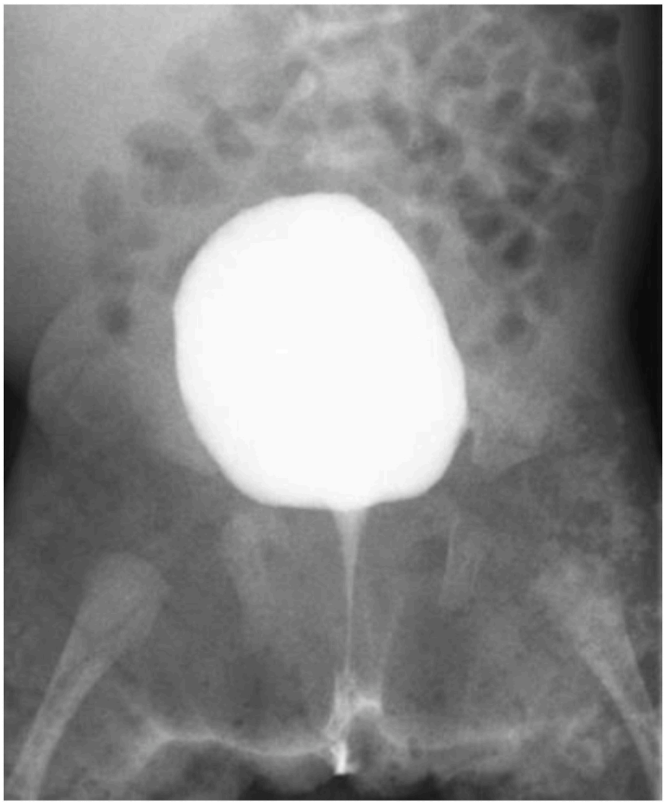

Fig. 3. Postoperative voiding cystourethrography showed no obvious abnormalities.

mucosa was noted at the base of the umbilical cord. Patency to the bladder was not evident on gross examination, and no abnormalities other than the urachal remanent were identified. The prolapsed urachal mucosa and remnants were excised. A histopathological examination confirmed this diagnosis. Postoperative voiding cystourethrography showed no obvious abnormalities, such as urethral stricture or vesicoureteral reflux (Fig. 3). The patient was healthy and had no urological symptoms at the 5-year follow-up.

3. Discussion

Urachal anomalies result from the failed obliteration of the urachus during the second trimester of pregnancy. The type of urachal anomaly was classified according to the persistent segment of the urachus. Patent urachus, a persistent communication between the bladder and umbilicus, is a rare malformation with an incidence of 3 per 1,000,000 live births. It is three times more common in males than females.[2] It is usually identified by urinary leakage from the umbilicus after birth. In some cases, a patent urachus can be prenatally diagnosed. Some reports have described patent urachus appearing as a cyst located at the base of the umbilical cord in early pregnancy.[1,3] As gestation progresses, the cyst becomes larger and spontaneously ruptures. When the cyst ruptures outside the umbilical cord, the bladder may prolapse through the patent urachus.[3] On the other hand, when the cyst ruptures inside the umbilical cord, retrograde voided fetal urine may form umbilical cord pseudocysts or be absorbed into the Wharton' jerry, resulting in GUC as same as the present case.

sIUGR has been defined as the EFW of one twin <10th percentile and an EFW discordance ≧25%. sIUGR accounts for 8.7% of MD twin pregnancies.[4] The pathophysiology of sIUGR in MD twin pregnancies relies not only on unequal placental sharing but also on blood flow interchange through placental anastomoses. In this case, worsening of arterial blood flow in the umbilical cord of the affected twin was observed later in pregnancy. The exact relationship between sIUGR and GUC remains unknown. Although cases of prenatally diagnosed localized GUC often have chromosomal abnormalities, and amniocentesis might be performed,[5] no reports have described severe complications in cases with diffuse GUC. The arterial flow of the umbilical cord of prenatally detected diffuse GUC <30cm in length was normal in all cases.[1]

Moreover, discordant umbilical cord in MD twins indicated the possibility of an epigenetic cause of GUC in our cases.

Surgery was performed in almost all GUC cases in the early postnatal period, and no obvious complications, including lower urinary tract dysfunction, were observed after surgery. Knowledge and proper diagnosis of this rare condition may help provide perinatal support to the parents and prevent unnecessary medical interventions.

4. Conclusion

We report the first case of prenatally diagnosed diffuse GUC resulted from patent urachus in a MD twin with sIUGR. This case indicates that GUC is epigenetic and unrelated to multiple births.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Credit author statement

Satoko Matsuyama: Investigation, Data curation, Visualization, Writing- Original draft preparation. Fumi Matsumoto: Writing- Reviewing and Editing. Futoshi Matsui: Writing- Reviewing and Editing. Koji Yazawa: Writing- Reviewing and Editing.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared ton influence the work reported in this paper.

References

1. Schaefer IM, Männer J, Faber R, Loertzer H, Füzesi L, Seeliger S. Giant umbilical cord edema caused by retrograde micturition through an open patent urachus. *Pediatr Dev Pathol*. 2010 Sep-Oct;13(5):404–407. https://doi.org/10.2350/09-10-0731-CR.1. Epub 2010 Jan 19.
2. Nix JT, Menville JG, Albert M, Wendt DL. Congenital patent urachus. *J Urol*. 1958 Feb;79(2):264–273. https://doi.org/10.1016/s0022-5347(17)66268-2.
3. Matsui F, Matsumoto F, Shimada K. Prenatally diagnosed patent urachus with bladder prolapse. *J Pediatr Surg*. 2007 Dec;42(12):e7–e10. https://doi.org/10.1016/j.jpedsurg.2007.07.055.
4. Curado J, Sileo F, Bhide A, Thilaganathan B, Khalil A. Early- and late-onset selective fetal growth restriction in monochorionic diamniotic twin pregnancy: natural history and diagnostic criteria. *Ultrasound Obstet Gynecol*. 2020 May;55(5):661–666. https://doi.org/10.1002/uog.20849. Epub 2020 Apr 3.
5. Haac B, Garcia A, Moore J. Urachal remnant presenting as a giant, cystic umbilical cord. *Pediatr Surg Int*. 2017 Mar;33(3):393–395. https://doi.org/10.1007/s00383-016-4040-9. Epub 2017 Jan.