Journal of Perinatology (2010) 30, 500–502

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IMAGING CASE REPORT

Intermittent 'bulge' in the umbilical cord

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Journal of Perinatology (2010) **30,** 500–502; doi:10.1038/jp.2010.34

Swelling at the base of the umbilicus noted at birth can be an omphalocele, a knot, polyp, cyst, cord hemangioma or a hematoma. An omphalocele is a defect in the ventral abdominal wall with herniation of the abdominal viscera through the widened umbilical ring. The defect is covered by a sac consisting of amnion and lined by peritoneum. The incidence of omphalocele is 2.5 per 10 000 live births.[1] The viscus within an omphalocele is usually the small intestine or the liver. Occasionally a urachal cyst or diverticulum may be found. We present the case of a herniated bladder (urachal remnant) into the umbilicus presenting as an intermittent protuberance at the base of the umbilical cord.

Case presentation

A 3430 g male infant was delivered by a 32-year-old G2P1 mother at term gestation by cesarean section under spinal anesthesia for breech presentation. The pregnancy was remarkable for detection of an umbilical cord 'cyst' and a three-vessel cord by the obstetrician during a routine 18-week prenatal ultrasound. The cyst was not visualized on subsequent ultrasound examinations. After birth, the nurse practitioner noted a swelling at the base of umbilicus at birth and clamped the cord 6 inches from the insertion. An omphalocele was suspected and an orogastric tube was introduced for gastric decompression. The baby was transferred to the regional perinatal center.

On arrival at the neonatal intensive care unit, physical examination revealed a small and reducible swelling at the base of the umbilicus (Figure 1), which increased in size with crying. The anus was patent with no external genitourinary abnormalities. The baby voided and passed meconium. Subsequently, the swelling was less prominent.

Ultrasound of the umbilical cord identified a herniated urinary bladder, but no bowel loops extending into the protuberant umbilicus (Figure 2a). Abdominal ultrasound was normal except for increased echogeneticity in the gall bladder, and a normally located urinary bladder with a wide apex protruding into the umbilical cord (Figure 2b).

The baby underwent primary surgical repair on the second day of life that included umbilical exploration with excision of the distended urachal remnant from the dome of the bladder. A Foley catheter (C.R. Bard Inc., Covington, GA, USA) was left in place to decompress the bladder for the next 72 h. A cystourethogram

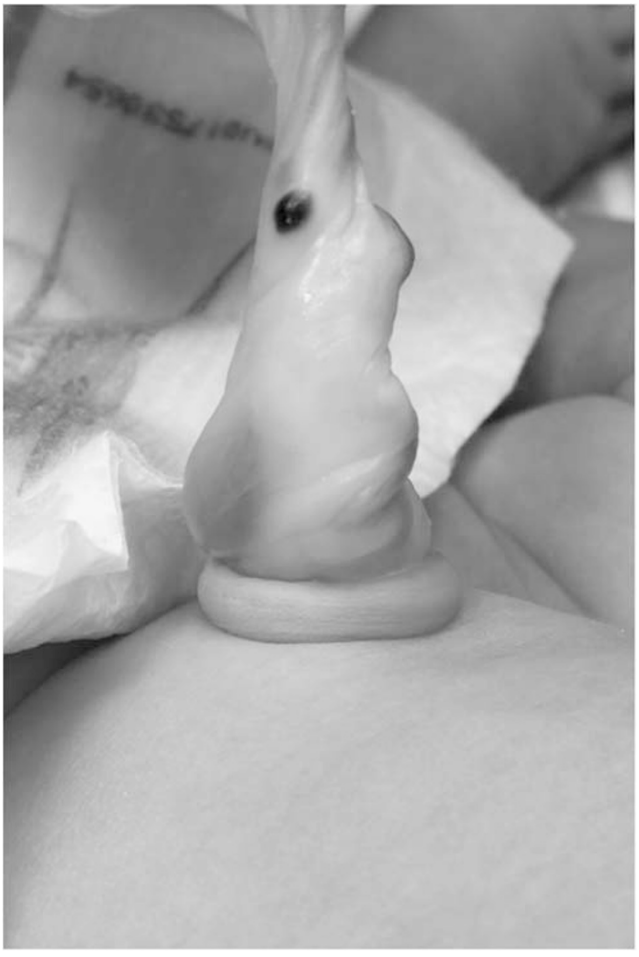

Figure 1 Umbilical cord with a small protuberance above the insertion. This protuberance increased in size with crying and decreased in size after voiding.

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Received 19 October 2009; revised 21 December 2009; accepted 17 January 2010

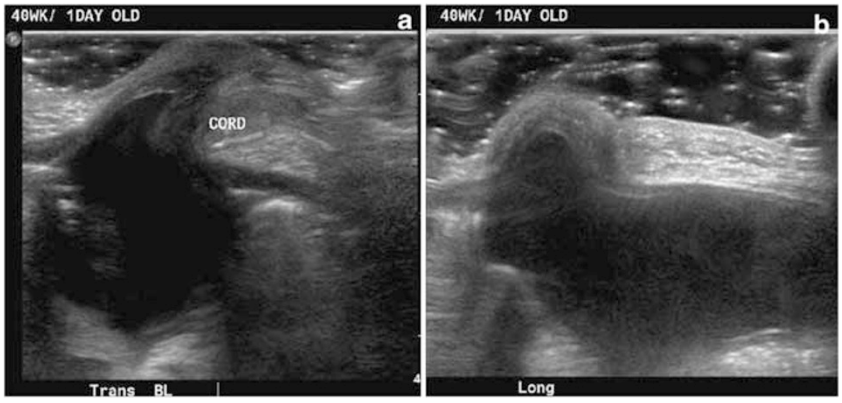

Figure 2 Ultrasound of the abdomen and umbilical cord. Herniation of the upper part of the bladder into the umbilical cord is seen in the transverse (**a**) and longitudinal (**b**) axis.

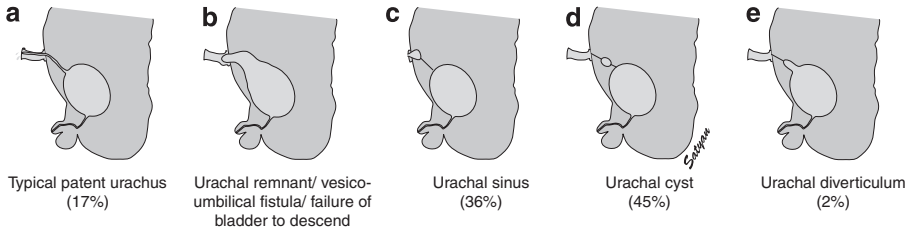

Figure 3 Anomalies associated with vesico-urachal remnants and their relative frequency based on a review of six case series in children (Table 1). (**a**) Typical patent urachus is a communication between the bladder and the umbilical cord and presents with umbilical drainage. (**b**) A urachal remnant with herniation of bladder into the umbilicus is a rare lesion as seen in our patient. The exact incidence of this lesion is not known. (**c**) An external urachal sinus presents with umbilical discharge but does not communicate with the urinary bladder. (**d**) Urachal cysts and (**e**) diverticula are often detected by an abdominal ultrasound during work-up of abdominal pain.

performed before the discharge showed no evidence of a diverticulum or leak. Histopathology of the excised mass showed a transitional epithelial lining suggestive of a urachal remnant.

Discussion

The urachus closes by desquamation of its epithelial mucosa into a solid cord between the 4th and 5th month of gestation.[2] Urachal anomalies result from a failure of fibrosis and involution of the urachus during fetal development.[3] A variety of clinical urachal anomalies exist (Figure 3).[4] Congenital patent urachus is a rare anomaly occurring in only 3 of more than 1 000 000 admissions to a large pediatric center.[5,6] Patent urachus may present with local cord enlargement, edema,[7] delayed sloughing[8] or with periumbilical discharge.[5] This condition should be differentiated from that of omphalitis, umbilical granuloma, patent omphalomesenteric duct, infected umbilical vessel or external urachal sinus (Figure 3c).[5] Analysis of the periumbilical fluid for creatinine and urea is useful.[5] A urachal remnant with vesicoumbilical fistula representing failure of the bladder to descend (Figure 3b) or herniation of bladder into the umbilical cord is rare and can present as a localized umbilical swelling[5] as seen in our patient. If this mass is not suspected at birth, it may be inadvertently injured while clamping the cord or during umbilical catheterization.[9] This injury may result in persistent umbilical drainage or urinary ascites. Urachal cyst (Figure 3d) and diverticulum (Figure 3e) do not present in the newborn period and are often detected during a work-up of abdominal pain or urinary tract infection. Complete excision of the patent urachus and with a cuff of bladder is recommended. If urachal remnants are not removed in childhood and they are observed, exposure to chronic urinary stasis, infection and inflammation in the remnant may predispose the patient to potentially lethal malignancy.[10]

The relative frequency of these lesions in six case series of 184 children with urachal anomalies is shown in Table 1. The precise incidence of an urachal remnant herniating into the umbilical cord (Figure 3b) is not described in the literature. These numbers differ from those quoted in a standard textbook of urology (patent urachus—50%, umbilical–urachal sinus—15%, urachal cyst—30% and vesicourachal diverticulum—3–5%).[11]

In our patient with a urachal remnant, the umbilical swelling was detected at birth, was not traumatized, and underwent successful surgical repair. We present this patient with this rare anomaly to emphasize the importance of physical examination at birth.

Table 1 Relative frequency of various urachal anomalies in children

Reference	*Total number of patients*	*Patent urachus (%)*	*Urachal sinus (%)*	*Urachal cyst (%)*	*Urachal diverticulum (%)*
Mesrobian *et al.*[12]	21	2 (10%)	9 (43%)	9 (43%)	1 (4%)
Cilento *et al.*[13]	45	7 (15%)	22 (49%)	16 (36%)	0
Yiee *et al.*[14]	31	7 (23%)	5 (16%)	19 (61%)	0
Choi *et al.*[15]	21	6 (29%)	4 (19%)	10 (47%)	1 (5%)
Ashley *et al.*[10]—children only	46	6 (13%)	14 (30%)	25 (54%)	1 (2%)
Huang *et al.*[16]	20	4 (20%)	13 (65%)	3 (15%)	0
Total	184	32 (17%)	67 (36%)	82 (45%)	3 (2%)

Conflict of interest

The authors declare no conflict of interest.

References

1 Baird PA, MacDonald EC. An epidemiologic study of congenital malformations of the anterior abdominal wall in more than half a million consecutive live births. *Am J Hum Genet* 1981; **33**: 470–478.

2 Caldamone AA. Anomalies of the bladder and cloaca. In: Gillenwater JY, Grayhack JT, Howards SS, Duckett JW (eds). *Adult and Pediatric Urology*. Vol. 2. Year Book Medical Publishers: Chicago, IL, 1987, pp 1809–1835.

3 Austin PF. Umbilical and urachal anomalies. In: Wilcox DT, Godbole PP, Koyle MA (eds). *Pediatric Urology—Surgical Complications and Management*. Wiley-Blackwell: UK, 2008, p 92.

4 Bauer SB, Retik AB. Urachal anomalies and related umbilical disorders. *Urol Clin North Am* 1978; **5**: 195–211.

5 Gearhart JP. Exstrophy, epispadias, and other bladder anomalies. In: Walsh PC, Retik AB, Vaughan Jr ED, Wein AJ (eds). *Campbell's Urology*. 8th edn, Vol. 3. Saunders: Philadelphia, 2002, pp 2136–2196.

6 Nix JT, Menville JG, Albert M, Wendt DL. Congenital patent urachus. *J Urol* 1958; **79**: 264–273.

7 Schiesser M, Lapaire O, Holzgreve W, Tercanli S. Umbilical cord edema associated with patent urachus. *Ultrasound Obstet Gynecol* 2003; **22**: 646–647.

8 Razvi S, Murphy R, Shlasko E, Cunningham-Rundles C. Delayed separation of the umbilical cord attributable to urachal anomalies. *Pediatrics* 2001; **108**: 493–494.

9 Mattei P. Urachal remnant perforation during umbilical vein catheterization in a newborn. *J Pediatr Surg* 2007; **42**: 722–724.

10 Ashley RA, Inman BA, Routh JC, Rohlinger AL, Husmann DA, Kramer SA. Urachal anomalies: a longitudinal study of urachal remnants in children and adults. *J Urol* 2007; **178**: 1615–1618.

11 Frimberger D, Kropp BP. Bladder anomalies in children. In: Wein AJ, Kavoussi LR, Novick AC, Partin AW, Peters CA (eds). *Campbell-Walsh Urology*, Vol. 4. Saunders: Philadelphia, 2007, pp 3576–3579.

12 Mesrobian HG, Zacharias A, Balcom AH, Cohen RD. Ten years of experience with isolated urachal anomalies in children. *J Urol* 1997; **158**: 1316–1318.

13 Cilento Jr BG, Bauer SB, Retik AB, Peters CA, Atala A. Urachal anomalies: defining the best diagnostic modality. *Urology* 1998; **52**: 120–122.

14 Yiee JH, Garcia N, Baker LA, Barber R, Snodgrass WT, Wilcox DT. A diagnostic algorithm for urachal anomalies. *J Ped Urol* 2007; **3**: 500–504.

15 Choi YJ, Kim JM, Ahn SY, Oh JT, Han SW, Lee JS. Urachal anomalies in children: a single center experience. *Yonsei Med J* 2006; **47**: 782–786.

16 Huang CS, Luo CC, Chao HC, Chen HM, Chu SM. Urachal anomalies in children: experience at one institution. *Chang Gung Med J* 2003; **26**: 412–416.