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CASE REPORT

Bladder prolapse through a patent urachus presenting as an umbilical mass in the newborn: characteristic prenatal sonographic findings and the diagnostic benefit of postnatal cystography

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Abstract

Bladder prolapse through a patent urachus is rare. We present a newborn with an unusual exophytic, erythematous umbilical mass. Voiding cystography readily demonstrated continuity of the bladder dome with the umbilical mass, confirming bladder prolapse through a patent urachus. The diagnosis of bladder prolapse was rapidly made in a second newborn with similar imaging and clinical findings and confirmed by cystography. We discuss the embryology of this condition including the association with a vesico-allantoic cyst in utero. Pre- and postnatal images are presented. The use of cystography in diagnosis is emphasized.

Keywords Allantoic cyst · Bladder prolapse · Cystography · Neonate · Patent urachus · Vesico-allantoic cyst · Voiding cystourethrogram

Introduction

The urachus is the intra-abdominal embryological remnant of the allantois that extends from the bladder dome to the umbilicus [1]. Abnormalities of urachal involution occur in approximately 1.6% of children, with a 33% rate of regression in children under the age of 1 year [2]. Complete failure of involution makes up 1.5% of all urachal anomalies and results in a patent urachus with communication between the bladder and umbilicus [3]. Rarely, the bladder dome may evert and prolapse through a patent urachus presenting as an exophytic umbilical mass [3–5]. We report a case of bladder prolapse through a patent urachus. Characteristic clinical and imaging findings allowed prompt diagnosis in a second newborn with the same abnormality.

Case presentation

A male neonate was delivered by cesarean section. Prenatal ultrasound performed at 12 weeks' gestation revealed an enlarging bladder, adjacent to a 1.4-cm umbilical cord cyst. The cyst was noted to lie between the two umbilical arteries (Fig. 1). By 14 weeks of gestation, the umbilical cord cyst measured up to 6.4 cm and was contiguous with the bladder, compatible with a vesico-allantoic cyst (Fig. 1). At 28 weeks' gestation, the umbilical cyst was no longer seen, and the bladder was identified outside the fetus, raising the possibility of exstrophy (Fig. 2). This appearance persisted until delivery at 38 weeks. No associated abnormalities were present. Amniotic fluid volume and fetal karyotype screening were normal.

At birth, the infant exhibited a 4×3-cm, tubular, erythematous umbilical mass just below the umbilical cord (Fig. 3). The abdominal wall was intact. Normal male genitalia were identified. A bladder catheter was placed via the urethra without difficulty. The umbilical mass was thought to be a urachal or omphalomesenteric duct remnant. On ultrasound, the bladder

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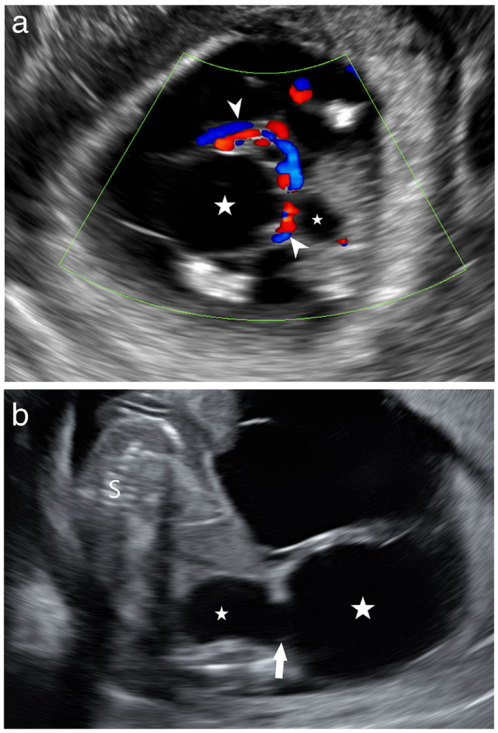

Fig. 1 Prenatal ultrasound at 12 weeks' and 14 weeks' gestation. **a** A transverse image with Doppler from a fetal ultrasound at 12 weeks' gestation. The larger cyst (*large star*) represents an umbilical cord cyst (allantoic cyst) and the small cyst (*small star*) represents the fetal bladder. These are joined by a patent urachus forming a vesico-allantoic cyst. The umbilical vessels (*arrowheads*) are draped around the umbilical cord cyst. **b** A longitudinal image with Doppler from a fetal ultrasound at 14 weeks' gestation. The open channel (*arrow*) connecting the fetal bladder (*small star*) and the allantoic cyst (*large star*) is seen to better advantage and represents a patent urachus. *S* fetal spine

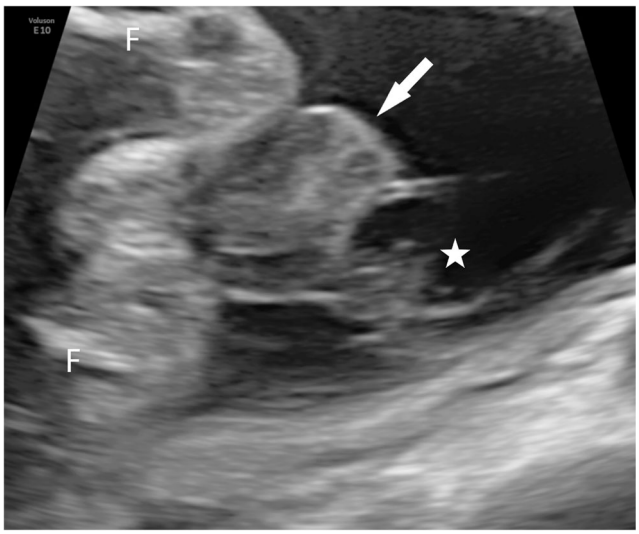

Fig. 2 A transverse image from a fetal ultrasound at 28 weeks' gestation at the level of the umbilical insertion. The bladder is no longer seen in the fetal pelvis. A mass (*arrow*) is present outside the abdominal wall and adjacent to the umbilical cord and represents an everted and herniated bladder. Note that the umbilical cord cyst (*star*) is significantly smaller. *F* femur

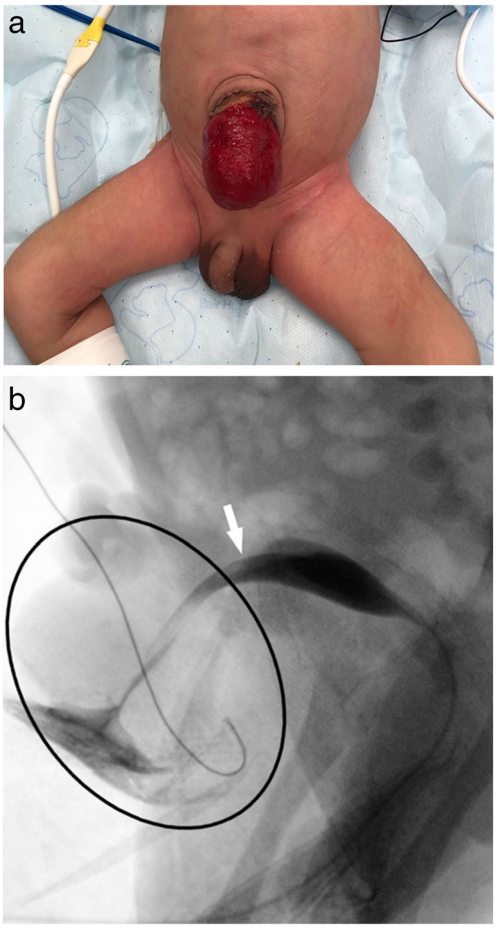

Fig. 3 Postnatal findings. **a** A photograph of a newborn boy demonstrates a large, red, pedunculated umbilical mass representing bladder prolapse through a patent urachus. **b** A lateral image from a voiding cystourethrogram demonstrates the free flow of contrast from the bladder dome (*arrow*) through a patent urachus to the anterior abdominal wall. The soft-tissue mass surrounding the patent urachal channel represents everted bladder mucosa (*oval*). The bladder dome is pulled superiorly and anteriorly toward the umbilical mass

was smooth, thin-walled and contained a catheter that was oriented toward the umbilicus. This prompted a voiding cystourethrogram (VCUG) that demonstrated a bladder dome that was pulled superiorly and anteriorly, and free flow of contrast from the bladder through the umbilical mass to the skin surface (Fig. 3). The diagnosis of bladder prolapse through a patent urachus was made. There was no urethral abnormality and no abnormalities of the pelvic bones. The infant underwent resection of the urachus, closure of the bladder dome defect, and umbilicoplasty. Postoperative VCUG demonstrated an intact bladder and normal voiding.

Discussion

The allantois is an extension of the posterior wall of the yolk sac and has both an extra-abdominal and intra-abdominal

component. The extra-abdominal component spans the umbilical cord and extends to the most inferior portion of the umbilical ring, a fibrous structure surrounding the umbilicus. The umbilical ring contains the umbilical vessels, vitelline duct, vitelline vessels and, transiently, the midgut [1]. The intra-abdominal component of the allantois extends from the umbilicus to the urogenital sinus, which forms the bladder and urethra. As the bladder enlarges and descends into the pelvis, the intra-abdominal portion of the allantois involutes and becomes the urachus. The urachus closes between the 16th and 18th weeks of gestation, becoming the median umbilical ligament, a fibrotic cord that extends from the bladder dome to the umbilicus [1, 6].

Complete patency of the urachus results in an open channel between the bladder dome and umbilicus. It presents after birth with leakage of urine from the umbilicus. Other urachal anomalies result from partial failure of urachal involution and include the urachal cyst (the most common urachal anomaly), urachal sinus and urachal diverticulum; these can present later in life as incidental findings [2].

Bladder prolapse through a patent urachus presenting as an exophytic umbilical mass at birth is unusual but has been reported [3–5]. The rarity of this condition may delay the recognition of the umbilical mass as prolapsed bladder and potentially lead to an error in diagnosis. As in previous reports of infants with bladder prolapse through a patent urachus, the cases we present herein exhibited a concurrent umbilical cord cyst in utero, located at the base of the umbilical cord adjacent to and continuous with the bladder. This has been referred to as an allantoic cyst or vesico-allantoic cyst. Vesico-allantoic cysts are described on prenatal ultrasound as bilobed cystic structures with one "lobe" of the cyst representing the umbilical cord cyst and the other representing the fetal bladder. A patent urachus connects the two cystic structures [5] (Figs. 1 and 4). Vesico-allantoic cysts characteristically splay the umbilical vessels (Fig. 1). It is theorized that intrauterine rupture of the umbilical portion of the cyst allows the bladder dome to evert and prolapse through the patent urachus (Fig. 4) [4]. Indeed, as in the case we present and in other reported cases, disappearance and presumed rupture of an umbilical cyst in a fetus, reported between 25 weeks and 31 weeks of gestation, occurs at the same time the bladder is identified outside the fetal abdominal wall [3–5]. The etiology of the umbilical cord cyst rupture is unknown but may be related to pressure from fetal urination or osmolality differences between the fetal urine and the umbilical cord stroma [3, 7].

Bladder prolapse through a patent urachus presents in the newborn as a red, fleshy mass at the inferior aspect of the base of the umbilical cord. The mass represents the exposed, prolapsed bladder mucosa [3–5] (Figs. 3 and 5). The caudal location of the mass relative to the umbilical cord reflects the relationship of the fetal allantois and urachus to the umbilical ring. Observing urine or a bladder catheter coming from the umbilical mass confirms bladder prolapse via a patent urachus [4]. Bladder prolapse may be distinguished from bladder exstrophy by in utero visualization of the bladder until the latter half of the second trimester, by the absence of pubic diastasis, and by the lack of anomalies of genitalia (epispadias in males and a bifid clitoris in females) [1]. Distinguishing bladder prolapse from an umbilical hernia may be done

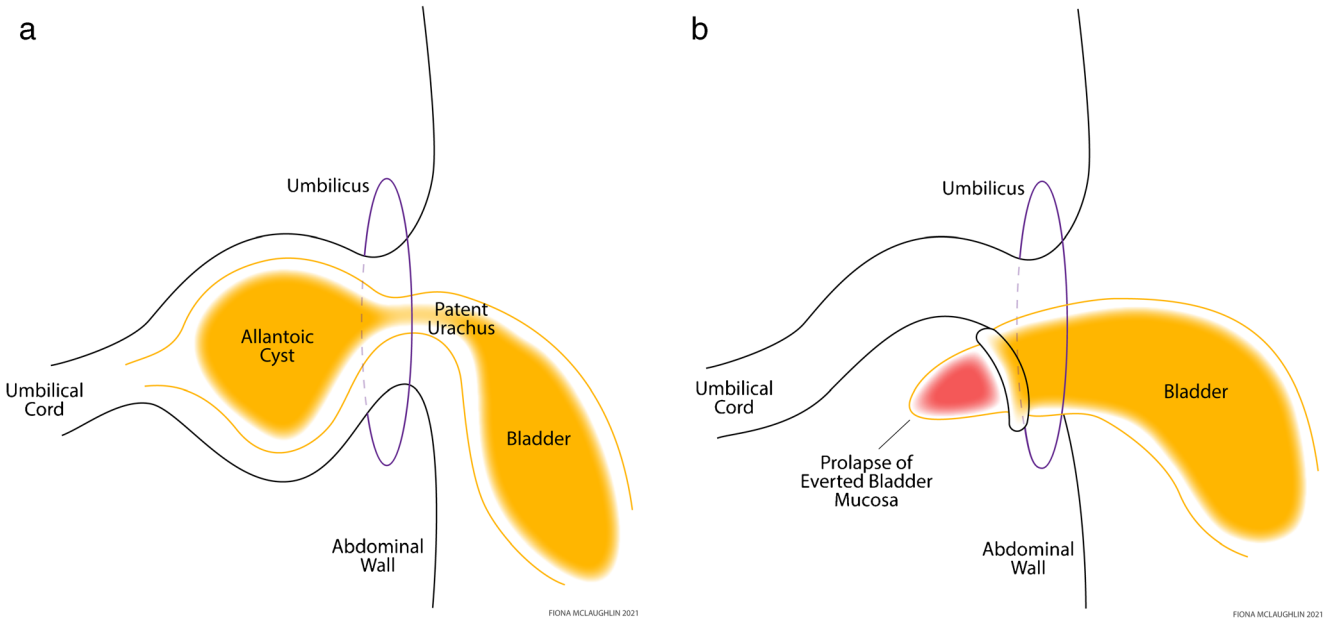

Fig. 4 Illustrations demonstrate the association of a vesico-allantoic cyst and bladder prolapse through a patent urachus. **a** In utero, an allantoic (umbilical) cyst communicates with the bladder via a patent urachus forming a vesico-allantoic cyst. The allantoic cyst enlarges during the second trimester, and disappears during the second trimester or early third trimester likely secondary to rupture. **b** At birth, an exophytic mass inferior to the insertion of the umbilical cord represents the bladder, which has everted and herniated through the patent urachus

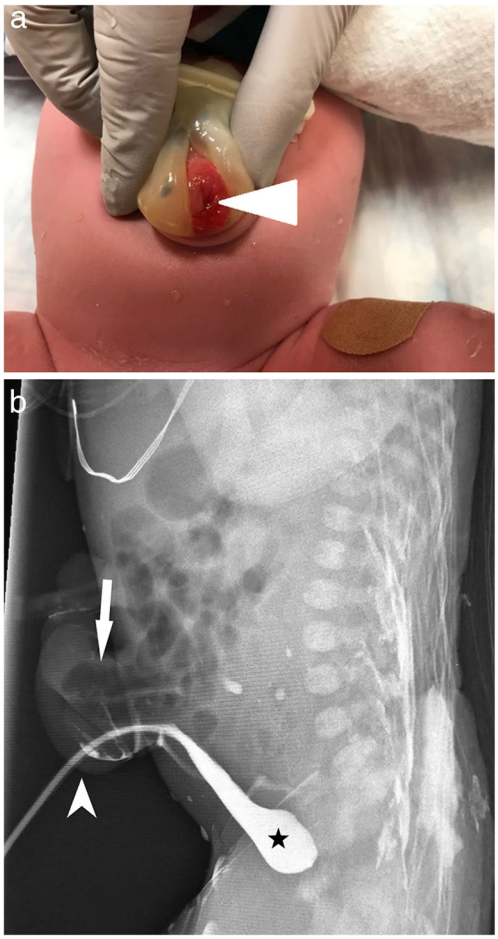

Fig. 5 A second male neonate with a prenatal history of umbilical cord cyst presented with a 1-cm exophytic stoma-like red mass inferior to the umbilicus. **a** A photograph demonstrates the exophytic mass. A central lumen (*arrowhead*) is evident. This mass represents everted bladder mucosa herniated through a patent urachus. Note the intact periumbilical skin, the lack of an abdominal wall defect and the location of the mass at the base of the umbilicus. **b** A lateral image from a cystogram performed through the central lumen of the umbilical mass (*arrowhead*) shows contrast filling the bladder (*star*). Continuity of the umbilical mass with the bladder confirmed the diagnosis of prolapsed bladder through a patent urachus. Note the bowel containing umbilical hernia (*arrow*). There is external spillage of contrast onto the infant's back. A renal ultrasound was normal. At 2 days of age, the infant underwent urachal resection and umbilicoplasty. A follow-up renal and bladder ultrasound at 1 month of age was normal

clinically. In contrast to an umbilical hernia, which is secondary to incomplete closure of the abdominal wall fascia and is covered by skin, bladder prolapse occurs below the umbilical insertion and has no skin covering [3, 8].

In infants in whom the diagnosis is unclear, contrast studies demonstrating continuity of the bladder and the anterior abdominal wall mass are diagnostic. VCUG findings include a bladder directed toward the umbilicus, a normal bladder neck and urethra and free flow of contrast from the bladder lumen through the umbilical mass (Fig. 3) [3]. Similarly, an antegrade contrast study through the central orifice of the umbilical mass, when identified, will reveal communication with the bladder (Fig. 5). Additional imaging is unnecessary, as no known significant associated abnormalities have been reported. Bladder function after repair is normal.

Other causes of umbilical masses, including isolated umbilical cord cyst, pseudocyst of the cord secondary to mucoid degeneration of Wharton's jelly, and omphalomesenteric duct cyst, will not demonstrate communication with the bladder [1].

Conclusion

We present an infant with an unusual umbilical mass secondary to bladder prolapse through a patent urachus. Identifying characteristic clinical and imaging features allowed for rapid diagnosis in another infant born with the same condition (Fig. 5). Characteristic findings on prenatal ultrasound include a vesico-allantoic cyst that disappears in the late second trimester or early third trimester concomitant with the identification of a mass outside the fetal abdomen. Diagnosis is rapidly made with VCUG or retrograde cystography via the umbilical orifice. At birth, the umbilical mass lies below the cord insertion. Pediatric radiologists, urologists and surgeons should be aware of this unusual entity as it is easily repaired and has no known significant associated abnormalities and no sequelae.

Declarations

Conflicts of interest Dr. Mark C. Liszewski is an unpaid member of the Carestream Health Medical Advisory Board and receives grant support from Carestream Health for an unrelated study. Dr. Liszewski has received travel and meal support from Carestream Health.

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