

The umbilical mass: a rare neonatal anomaly

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Abstract Umbilical anomalies are a rare presentation in the pediatric patient. The differential diagnosis includes anomalies resulting from urachal and vitelline duct derivatives such as urachal sinus, urachal cyst, urachal diverticulum, patent urachus, herniated Meckel's diverticulum, umbilico-enteric fistula, or umbilical polyp. In this article, a case presentation of an umbilical anomaly along with the differential diagnosis and management options are discussed. Based upon this review of the literature, the authors propose a management algorithm for treating children with umbilical anomalies.

Keywords Umbilical cord · Umbilicus · Congenital anomalies · Urachus · Vitelline duct · Persistent urachus · Persistent vitelline duct

Case report

An 8-day-old male was referred to the pediatric surgery clinic for the evaluation of an “enormous umbilical cord.” The patient's perinatal course was uncomplicated, and he was delivered vaginally at 36 weeks gestation. The child was noted to have an umbilical cord of normal length and size, however, distal to the skin segment was a markedly dilated portion covered by amnion. One day following delivery, the patient was

discharged home. He had been tolerating formula feedings with normal bowel and bladder function. On physical exam in our clinic, the patient had a 10 cm long by 2.5 cm in diameter soft umbilical stump with irreducible contents; the cord clamp was still in place (Fig. 1). The initial ultrasound of the mass was non-diagnostic, so a non-contrast CT of the abdomen was obtained. The CT scan demonstrated possible herniation of bowel into the umbilical stump (Fig. 2).

The child underwent emergent umbilical stump exploration. The skin was incised circumferentially at the skin-amnion junction; two umbilical arteries, a single umbilical vein, and a persistent urachal remnant were identified (Fig. 3). To rule out the possibility of a persistent omphalomesenteric duct remnant within the umbilicus, an infraumbilical curvilinear incision was fashioned in a skin crease. To determine patency of the urachal remnant, a Foley catheter was inserted and saline infused. The bladder, when distended, did not appear to communicate with the urachal remnant. The urachal remnant was suture ligated with absorbable suture, the umbilical vessels were ligated with silk ties, the fascia was closed, and the umbilicus was then inverted after fashioning a pursestring at the skin edge and then tacking the umbilicus to the underlying fascial closure. Final pathologic analysis revealed a patent urachal remnant. The child's post-operative course was unremarkable, and he was discharged to home on the first post-operative day.

Discussion

The differential diagnosis of an umbilical mass in the neonate includes persistent omphalomesenteric

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Fig. 1 Photograph of umbilical stump demonstrating the amniotic remnant with irreducible contents

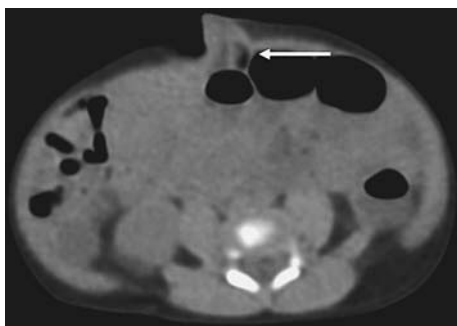


Fig. 2 Non-contrast CT-scan of the abdomen demonstrates possible herniation of bowel into the umbilical stump (arrow)

(vitelline) duct, urachal derivatives, as well as the rare findings of ectopic pancreatic [1, 2] and liver tissue [3]. Our patient was unique in that, unlike several case reports of a “giant umbilical cord” in the literature, he had a normal umbilical cord with a significant amniotic remnant with irreducible contents [4, 5]. The differential diagnosis upon examination of this patient in clinic included incarcerated hernia (small bowel or Meckel’s diverticulum) or a persistent urachal remnant. The CT-scan of the abdomen demonstrated what appeared to be intestinal bowel gas herniating into the large umbilical mass. This mandated emergent exploration of the umbilicus in an effort to reduce the hernia, inspect the umbilical cord contents, and repair the defect.

Umbilical anomalies may be broadly divided into two general classes: those arising from failed obliteration of the urachus or failed obliteration of the vitelline duct.



Fig. 3 Intra-operative image of two umbilical arteries, one umbilical vein, and a persistent urachal remnant within the umbilical stump

Urachal anomalies are rare in the pediatric population [7, 8]. Typical anomalies include urachal cyst, urachal sinus, patent urachus, or urachal diverticulum. Urachal cysts, the most common of the urachal anomalies, can present anytime from birth to adolescence with the average age of presentation being 4 years of age [7]. While typically asymptomatic, urachal cysts are often detected clinically following infection of the cyst. Patients may present with fever, periumbilical pain, or other symptoms of acute infection. *Staphylococcus* species are the most common infecting organism [7–9]. Spontaneous rupture of an infected cyst has been shown to lead to peritonitis [7]. A urachal sinus consists of a dilated, patent urachal end that opens into the umbilicus. Such sinuses may present clinically with either clear drainage or infection. Patients with urachal sinuses may demonstrate symptoms of a urinary tract infection if the sinus projects into the bladder [7]. The patent urachus, typically presents between 6 weeks and 6 years of age with urine leaking from the umbilicus [7]. An asymptomatic urachal diverticulum, another urachal anomaly, is often detected incidentally [10]. The presentation of other associated anomalies synchronously with urachal anomalies is quite rare, and subsequent work-up for associated anomalies is not typically warranted [9, 13].

Diagnosis of urachal anomalies begins with a thorough history and physical examination. Clinical suspicion is often further supported by ultrasound [9, 11]. In their analysis of 45 children with urachal anomalies, McCollum et al. [9] were able to correctly diagnose over 90% of the children using ultrasound. Contrast sinograms have also demonstrated diagnostic success

[12]. In contrast, voiding cystourethrograms have not been shown to be clinically effective in making the diagnosis of a urachal anomaly; urinalysis and urine culture are also non-diagnostic in this setting [8, 9].

Varied opinions exist in the literature with regard to the adequate treatment of a detected urachal anomaly. There is consensus that if the anomaly is symptomatic, it must be surgically excised, however, some argue that all anomalies should be excised due to possible malignant potential. Both adenocarcinoma and transitional cell carcinoma have been identified on pathological examination of urachal anomalies [13, 14]. Such findings support the prophylactic removal of both asymptomatic and symptomatic urachal anomalies.

When surgical management is indicated, exploration via a transverse infraumbilical incision is recommended. Extraperitoneal dissection of the urachal remnant is then initiated, as described in the case report, leading to subsequent excision of the remnant from the umbilicus to the bladder apex. However, in the setting of acute infection, primary incision and drainage followed by elective excision of the remnant may be necessary. Several groups have also investigated the possibility of laparoscopic repair of urachal anomalies, and determined that such an approach may lead to a shorter recovery time and decreased length of hospital stay [15, 16].

Like urachal anomalies, failed vitelline (omphalomesenteric) duct obliteration may also lead to umbilical anomalies resulting in the formation of umbilical cysts, umbilical sinuses, or umbilical-enteric fistulas [6]. Such fistulas most commonly involve the ileum but may occasionally include the appendix or the cecum [7, 18]. Clinically, umbilical-enteric fistulas can be detected by passage of gas or feculent, bilious, or mucous discharge through the umbilicus thereby implying connection to the bowel [7, 18]. Failed obliteration of the vitelline duct may also present as an umbilical polyp. Polyps may be recognized clinically by the presence of reddish umbilical tumors that demonstrate minimal bleeding or discharge [17]. An umbilical Meckel's diverticulum could also result due to vitelline duct persistence. Herniation of this lesion may lead to subsequent incarceration thus requiring prompt evaluation and treatment to prevent bowel necrosis.

Evaluation of a persistent vitelline duct is initiated with a thorough history and physical exam. If a umbilical polyp is suspected, ultrasound is the imaging modality of choice [17]. However, if an umbilico-enteric fistula is suspected, a fistulogram may be the best initial imaging approach [18]. CT-scan may also aid in diagnosis if the ultrasound or fistulogram are non-diagnostic. If a herniated Meckel's diverticulum is present, CT may

provide the most accurate diagnosis. An association between failed obliteration of the vitelline duct and malrotation of the intestines has been suggested in the literature [18]. This implies that in the child presenting with an anomaly caused by failed obliteration of the vitelline duct, an upper gastrointestinal series to rule out malrotation is indicated [18]. Treatment of anomalies resulting from failed obliteration of the vitelline duct includes surgical resection of the umbilical contents with possible bowel resection.

Consistent with the work-up that was conducted in our case, we propose that ultrasound is the best and most cost effective initial imaging modality to accurately diagnose umbilical anomalies. If ultrasound is non-diagnostic, CT-scan is an appropriate secondary study. If there is strong clinical suspicion for an umbilical-enteric fistula, a fistulogram may be the most helpful in identifying the fistula track. Following accurate diagnosis, surgical resection of the umbilical anomaly, as described in the case report, is the preferred method of treatment.

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