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CASE REPORT

Umbilical Enteric Fistula: a Rare Cause of Neonatal Umbilical Mass

JJK Ip[1], PKT Hui[1], L Lan[2], WWM Lam[1], MT Chau[1]

[1]Department of Radiology, and [2]Department of Surgery, Queen Mary Hospital, 102 Pokfulam Road, Hong Kong

ABSTRACT

Congenital umbilical anomalies are common presentations in paediatric patients, and are usually classified as persistent umbilical cord structures or failure of umbilical ring closures. Urachal anomalies, umbilical herniations, and vitelline duct anomalies are the main classifications for these disorders. Among these disorders, only congenital urachal cyst is common. All anomalies associated with the vitelline duct are rare, and umbilico-enteric fistula is the rarest of all. This report is of a rare umbilical fistula in a neonate with persistent vitelline duct, with a brief review of the embryology, anatomy, and classification of vitelline duct malformations. The differential diagnosis and management options are also discussed.

Key Words: *Congenital abnormalities; Umbilical cord; Umbilicus; Urachus; Vitelline duct*

中文摘要

臍腸瘻：新生兒臍部腫塊的罕見病因

葉精勤、許其達、藍傳亮、林慧文、周明德

小兒的先天性臍異常很常見，通常出現持續的臍膨出或臍環未能閉合。臍異常的主要病理可分為臍尿管異常、臍疝氣、及卵黃管發育異常，其中先天性臍尿管囊腫最普遍。而與卵黃管有關的異常很少，其中的臍腸瘻更為罕見。本文報告一名新生兒未閉的卵黃管出現罕見的臍瘺，從胚胎學、解剖學和分類學討論有關卵黃管異常的情況，並探討此症的鑒別診斷及醫治方法。

INTRODUCTION

Umbilical cord anomalies are the most frequent cause of newborn consultations for paediatric surgeons. Urachal anomalies, umbilical herniations, and vitelline duct anomalies are the main classifications for these disorders.[1] 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, umbilical enteric fistula, or umbilical polyp.[2] These disorders can be classified according to the embryonic remnants in the umbilicus, including the urachus, vitelline duct, round ligament of the liver, extraperitoneal paravesical spaces, umbilical ring and the umbilicus itself.[3] Of the five types of congenital urachal abnormalities, only urachal cyst is common.

Correspondence: *Dr Janice JK Ip, Department of Radiology, Queen Mary Hospital, 102 Pokfulam Road, Hong Kong, China. Tel : (852) 2255 3284 ; Email : jk.janice.ip@gmail.com*

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All anomalies associated with the vitelline duct are rare except for Meckel's diverticulum.[3] This report is of a neonate with umbilical enteric fistula presenting with an umbilical nodule. The differential diagnosis and management options are discussed.

CASE REPORT

A 15-day-old boy was admitted in July 2011 for evaluation of an abnormal umbilical mass. The patient's perinatal course was uncomplicated, and he was delivered vaginally at full term. The child was noted to have an umbilical cord of normal length and size at the time of delivery. He was discharged home three days after delivery. He had been tolerating feeds and had normal bowel and bladder functions.

A pink nodular growth was noted after sloughing of the umbilical stump, however. The growth was progressively enlarging and discharging serous material. At physical examination, the patient had a 1.5- to 2.0-cm soft pink umbilicated nodule protruding from the umbilicus (Figure 1). Abdominal radiographs, taken at admission, showed normal bowel pattern, with no abnormal calcification or displacement of bowel loops (Figure 2).

Initial ultrasonography of the umbilical zone excluded the presence of an urachal duct, since there was no tubular or cystic lesion along the midline below the umbilicus to the urinary bladder. Bowel loops were present beneath the lesion (Figure 3). The dome of the urinary bladder was smooth and normal looking.

Fluoroscopic examination was done. Contrast was instilled into the sinus opening at the stoma-like

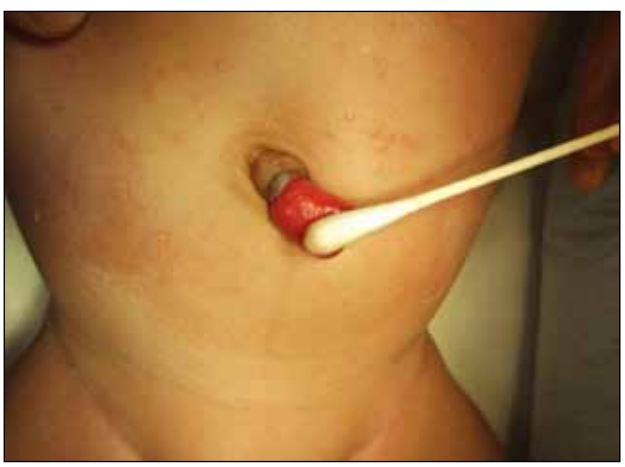

Figure 1. A 15-day-old baby boy with a small 1.5-cm soft pink umbilicated nodule protruding from the umbilicus.

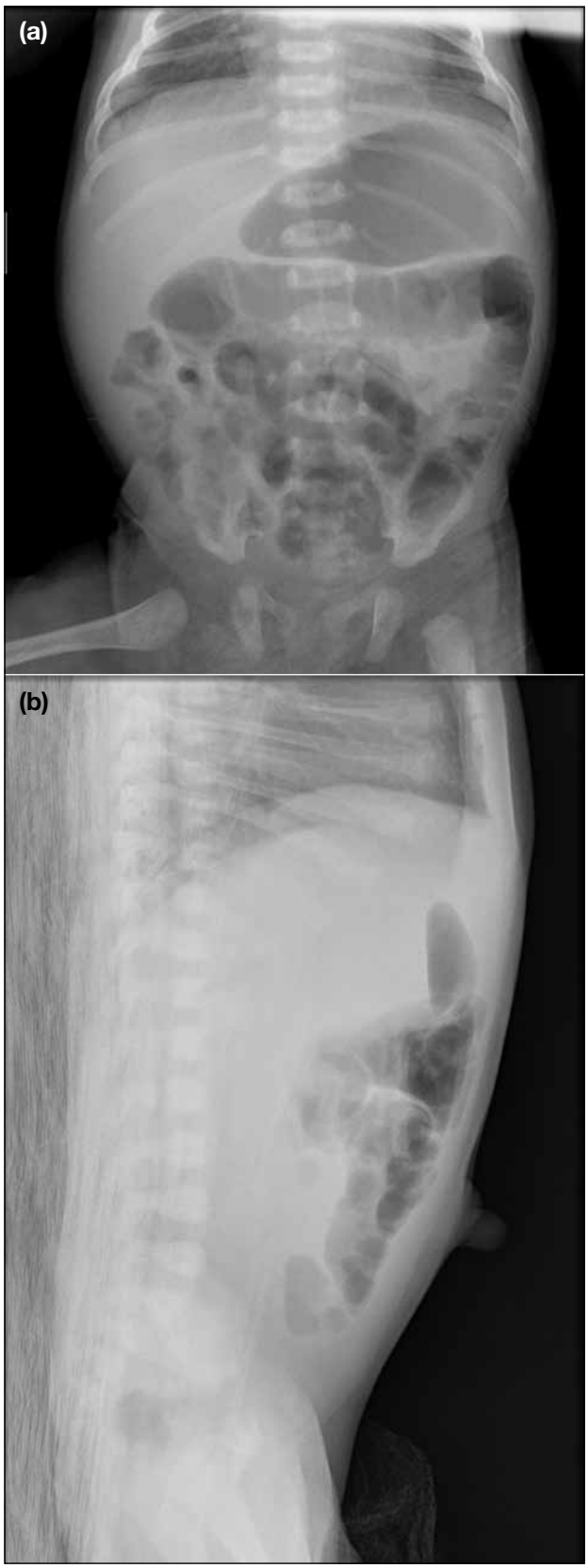

Figure 2. Abdominal radiographs showing (a) normal bowel pattern, with no abnormal calcification or displacement of bowel loops, and (b) the soft tissue umbilical nodule on lateral radiograph.

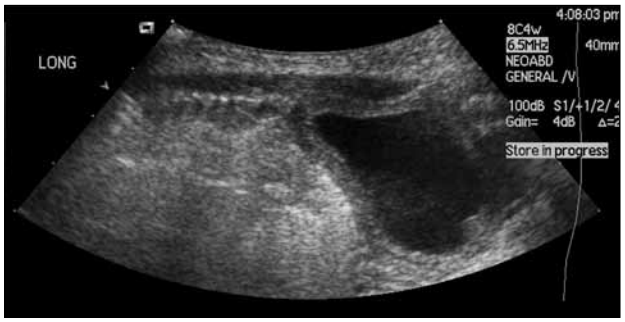

Figure 3. Initial ultrasonography of the umbilical zone shows only bowel loops beneath the lesion. No tubular or cystic lesion was seen along the midline below the umbilicus to the urinary bladder. The dome of the urinary bladder was smooth and normal looking.

umbilical polyp (Figure 4). Free passage of contrast through the umbilicus into the bowel loops was noted, without opacification of the urinary bladder or contrast spillage into the peritoneum. There was no abnormal bowel dilatation or herniation. The features were in keeping with an umbilical enteric fistula as a result of failed vitelline duct obliteration, which was confirmed intra-operatively (Figure 5).

DISCUSSION

Prenatally, the umbilicus is of paramount importance, providing the gateway between the mother and the foetus.[4,5] During the sixth week of embryogenesis, the mid-gut elongates and herniates into the umbilical cord. Within the umbilical cord, the mid-gut rotates 90° anticlockwise around the axis of the superior mesenteric artery. At the same time, the mid-gut elongates to form the jejunum and ileum, and the lumen of the vitelline duct closes. By the 10th week of embryogenesis, the mid-gut returns to the abdominal cavity and the vitelline duct becomes a thin fibrous band, which eventually disintegrates and is absorbed.[5,6] The vitelline duct will continue to grow if it fails to completely shrink and disintegrate.

Failure to obliterate the vitelline duct may produce different malformations, and involve a wide spectrum of anatomical structures and associated symptoms. These features may range from a completely patent vitelline duct at the umbilicus, as in this patient, to a variety of less common remnants such as cysts, fibrous cords connecting the umbilicus to the distal ileum, granulation tissue at the umbilicus, umbilical hernias, and the well-known Meckel's diverticulum (Figure 6).[6-8] The overall incidence of anomalies of the vitelline duct is 1 in 15,000 live births. Umbilical enteric fistula results

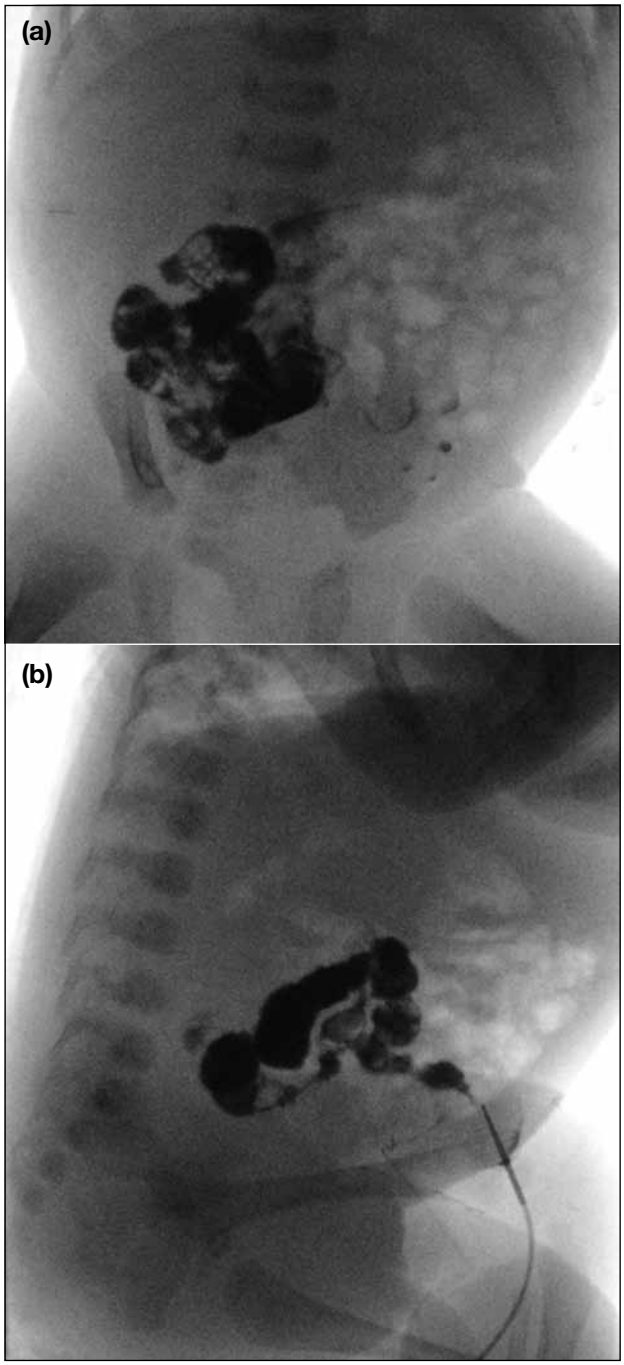

Figure 4. Fistulography demonstrates free passage of contrast through the umbilical nodule into the bowel loops. No contrast opacification of the urinary bladder or contrast spillage into the peritoneum was seen. The features are in keeping with failed vitelline duct obliteration, with umbilical enteric fistula.

from a completely patent vitelline duct, and is the least common of these anomalies. The duct remains open through its entire course. Patients with this anomaly may be brought to the attention of paediatricians or paediatric surgeons during the neonatal period due to mucous or faecal discharge from the umbilicus.

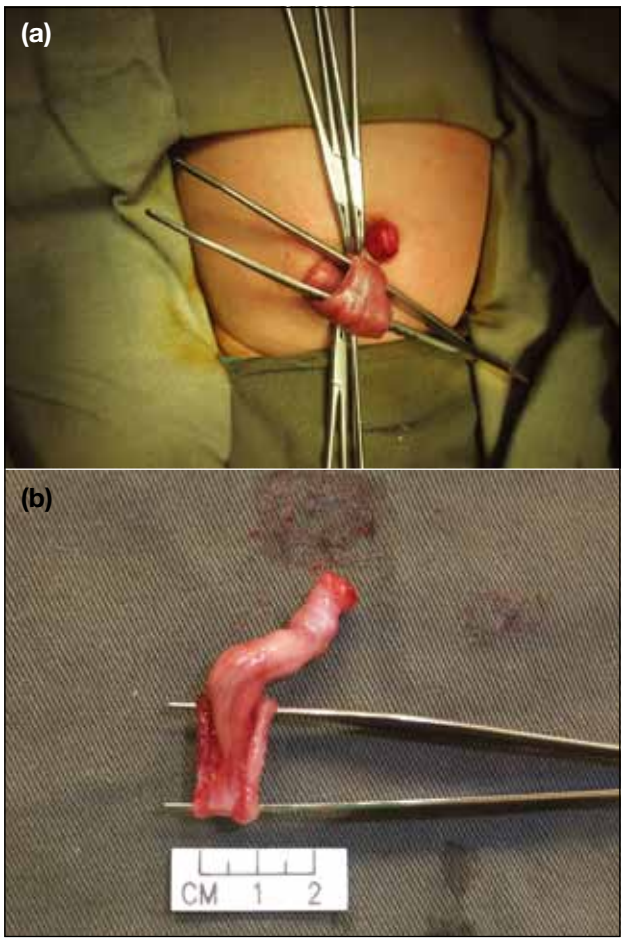

Figure 5. Laparotomy in this patient found an umbilical enteric fistula.

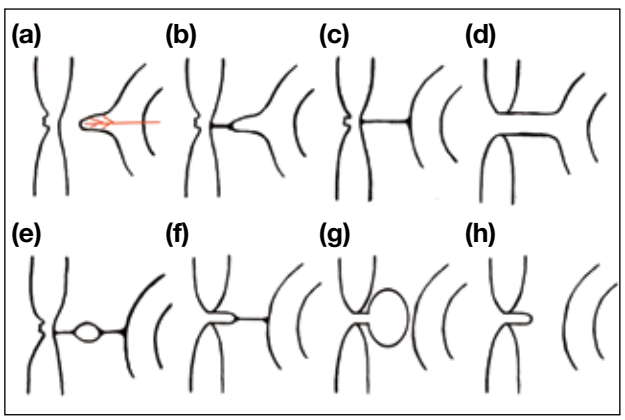

Figure 6. Diagrammatic illustrations of different malformations due to failure of obliteration of the vitelline duct. (a) Meckel's diverticulum with feeding vessel. (b) Meckel's diverticulum attached to the posterior surface of the anterior abdominal wall by a fibrous cord. (c) Fibrous cord attaching the ileum to the abdominal wall. (d) Umbilical enteric fistula: intestinal mucosa extends to the skin surface. (e) Vitelline cyst arising in a fibrous cord, the cyst may contain intestinal or gastric mucosa. (f) Umbilical sinus ending in a fibrous cord attaching to the ileum. (g and h) Vitelline cyst and sinus without intestinal attachments.

Ileoileal intussusception into the patent duct may occur, appearing clinically as ileal prolapse at the umbilicus. Partial obliteration of the vitelline duct may result in a vitelline duct sinus or cyst, a fibrous cord, or a Meckel's diverticulum. Symptoms may involve intussusception, intestinal obstruction from a variety of causes, melaena and anaemia, abdominal pain, and inflammation.[7]

Although symptoms occur most frequently during early childhood (especially in the first two years of life), they may also occur in adults.[7] Although these malformations are found with equal frequency among the sexes, a significantly higher incidence of symptoms is encountered in male patients. Yamada et al[9] reviewed 65 cases of patent vitelline duct in Japan and reported a male-to-female ratio of 2.8:1. The ducts averaged 3.8 cm in length and 1.1 cm in diameter.[9] Ectopic gastric mucosa was found in 10% of the patients.[9] A relatively high incidence (52.8%) of prolapse of the ileum was also noted.[9] In view of the high morbidity and mortality rates of patients with a prolapse of the ileum and the strong possibility of intestinal obstruction, surgical resection is the recommended treatment for patent vitelline ducts.

The differential diagnosis of an umbilical mass in neonates includes both developmental remnants and acquired entities (Table). Jauniaux et al[5] found embryonic remnants in 231 patients (23.1%), which included remnants of the allantoic duct (63%), vitelline duct (6.6%), and embryonic vessels (30.4%). Apart from developmental anomalies of the vitelline duct and urachus, umbilical masses may be related to lesions of the skin or an umbilical hernia. Masses associated with the skin include dermoid cysts, haemangiomas, and inclusion cysts. Among these, umbilical drainage is associated with granulomas, infection, and embryologic remnants. Granulation tissue may persist at the base of the umbilicus after cord separation. The tissue is composed of fibroblasts and capillaries, and can grow

Table. Differential diagnosis of umbilical mass according to the congenital structures involved.

Diagnosis	Congenital structures involved
Urachal derivatives	Urachal sinus
	Urachal cyst
	Urachal diverticulum
	Patent urachus
Vitelline duct derivatives	Herniated Meckel's diverticulum
	Umbilical enteric fistula
	Umbilical polyp

to more than 1 cm. Umbilical granulomas must be differentiated from other umbilical polyps, which do not respond to silver nitrate cauterization. Patients with omphalitis may present with purulent umbilical discharge or periumbilical cellulitis, and may have retained umbilical cord or ectopic tissue. Cellulitis may become severe within hours and progress to necrotising fasciitis and sepsis.

Abdominal radiography is not usually indicated for most children with umbilical lesions. However, radiography may be useful for children with omphalitis. Air in the subcutaneous tissue or muscle planes is an ominous sign. Ultrasonography is always helpful when a mass is present, and may be useful for identifying cysts of the umbilicus, as urachal cyst most commonly appears as a mass between the umbilicus and suprapubic area. Fistulography or sinography may be performed if a definitive opening is observed within the umbilicus by injecting water-soluble contrast medium into the opening. Cystography may also be indicated to identify bladder outlet obstruction in children draining frank urine from an urachal fistula. However, studies suggest that, in most patients, history and ultrasonography are sufficient for diagnosis.[10] The histopathology of umbilical remnants depends on the tissue of origin and may reveal intestinal or gastric mucosa in vitelline duct remnants or transitional or columnar epithelium in urachal remnants.

Surgery is the mainstay of treatment for vitelline remnants and other causes of paediatric umbilical masses, including urachal remnants, large umbilical granuloma, umbilical gangrene and necrotising fasciitis, and umbilical hernias that are symptomatic or do not spontaneously close. Surgical principles include identification of all structures of the umbilicus, excision of urachal or vitelline remnants, closure of the umbilical ring, and preservation of the natural appearance of the umbilicus. A patent vitelline duct must be traced to its origin and divided, as in this patient; the same applies to the urachus. When connected to the ileum, a patent vitelline duct may have a wide lumen or be a high-output fistula. This may lead to fluid and electrolyte deficits, especially in neonates, and treatment should be expeditious, with wedge or segmental resection. Morbidity and mortality can be avoided by prompt treatment. The outcome for infants and children with umbilical remnants is generally excellent, and no long-term problems occur in most patients.

Clinical manifestations of umbilical disorders are usually non-specific; use of ultrasonography and fluoroscopic examinations can help to identify most of these entities because of their typical locations and distributions in continuity with the urinary bladder or gastrointestinal tract. Knowledge of the embryology of the vitelline duct is necessary for good understanding of the various anomalies and their management. Early diagnosis and management are suggested in view of the possible complication of intestinal obstruction and the associated high morbidity and mortality rates.

REFERENCES

1. Ozel SK, Kazez A, Akpolat N. An unusual presentation of patent urachus: report of a case. Eur J Pediatr Surg. 2004;14:206-8.
2. Carlisle EM, Mezhir JJ, Glynn L, Liu DC, Statter MB. The umbilical mass: a rare neonatal anomaly. Pediatr Surg Int. 2007;23:821-4.
3. Khati NJ, Enquist EG, Javitt MC. Imaging of the umbilicus and periumbilical region. Radiographics. 1998;18:413-31.
4. Snyder CL. Current management of umbilical abnormalities and related anomalies. Semin Pediatr Surg. 2007;16:41-9.
5. Jauniaux E, De Munter C, Vanesse M, Wilkin P, Hustin J. Embryonic remnants of the umbilical cord: morphologic and clinical aspects. Hum Pathol. 1989;20:458-62.
6. Ballester I, Betlloch I, Pérez-Crespo M, Toledo F, Cuesta L. Atypical presentation of an omphalomesenteric duct cyst. Dermatol Online J. 2009;15:13.
7. Smith JR. Accessory enteric formations: a classification and nomenclature. Arch Dis Child. 1960;35:87-9.
8. Moore TC. Omphalomesenteric duct malformations. Semin Pediatr Surg. 1996;5:116-23.
9. Yamada T, Seiki Y, Ueda M, Yoshikawa T, Sempuku S, Kurokawa A, et al. Patent omphalomesenteric duct: a case report and review of Japanese literature. Asia Oceania J Obstet Gynaecol. 1989;15:229-36.
10. Little DC, Shah SR, St Peter SD, Calkins CM, Murphy JP, Gatti JM, et al. Urachal anomalies in children: the vanishing relevance of the preoperative voiding cystourethrogram. J Pediatr Surg. 2005;40:1874-6.