CASE REPORT

Umbilical Enteric Fistula: a Rare Cause of Neonatal Umbilical Mass

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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

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.

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All anomalies associated with the vitelline duct are rare except for Meckel’s diverticulum. 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](image1.png)

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

![Figure 2](image2.png)

*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.
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 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.
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.

Although symptoms occur most frequently during early childhood (especially in the first two years of life), they may also occur in adults. 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 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. Ectopic gastric mucosa was found in 10% of the patients. A relatively high incidence (52.8%) of prolapse of the ileum was also noted. 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 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>
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<th>Diagnosis</th>
<th>Congenital structures involved</th>
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| 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. 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.

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