

Ileal perforation and fistulated urachal remnant in Crohn's disease

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The urachus is an extra-peritoneal structure joining the bladder and the umbilicus; it lies between the transverse fascia and parietal peritoneum. Originating from the allantois and cloaca, the urachus provides a channel to allow drainage of the developing bladder in-utero. The lumen functionally closes before birth and the urachus atrophies in the post-natal period, leaving a persistent fibrous cord, known as the median umbilical ligament. If the lumen fails to fully close and atrophy in the early postnatal period, then it is known as a urachal remnant.

Case report

A 20-year-old male presented with an umbilical abscess and, under general anaesthesia, had an incision and drainage. He re-presented two months later with umbilical discharge and weight loss. Following re-admission, enteric contents was observed discharging from the umbilicus. Imaging identified an ileal perforation tracking extra-peritoneally and draining into the umbilicus via a fistula into a urachal sinus (Figure 1).

Figure 1: Sagittal slice of a CT scan of the abdomen and pelvis. The urachal remnant extends from the umbilicus to the bladder with inflamed small bowel lying immediately posteriorly.

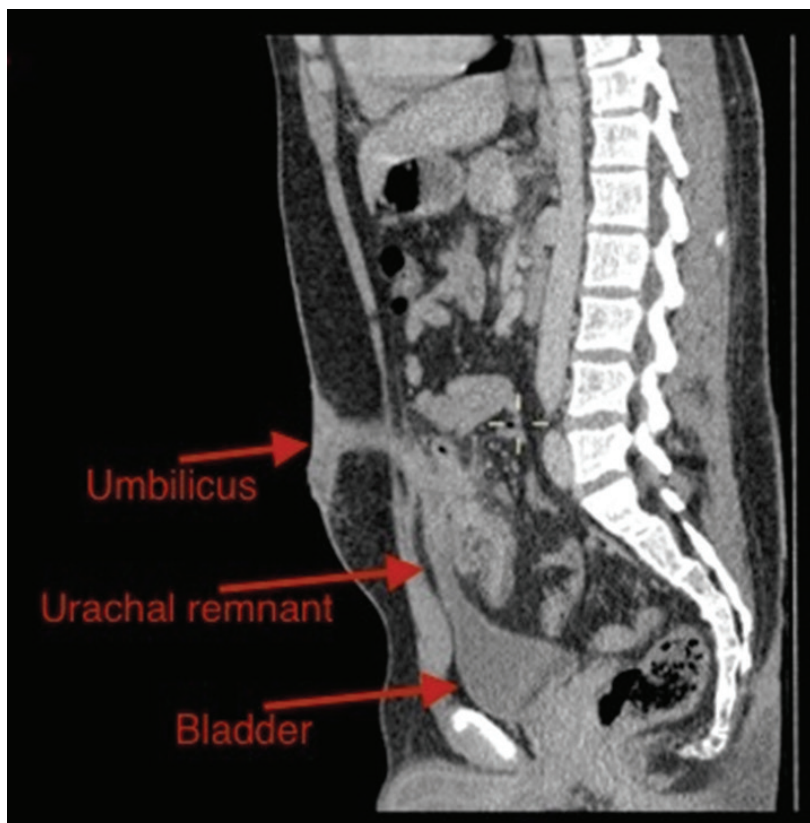
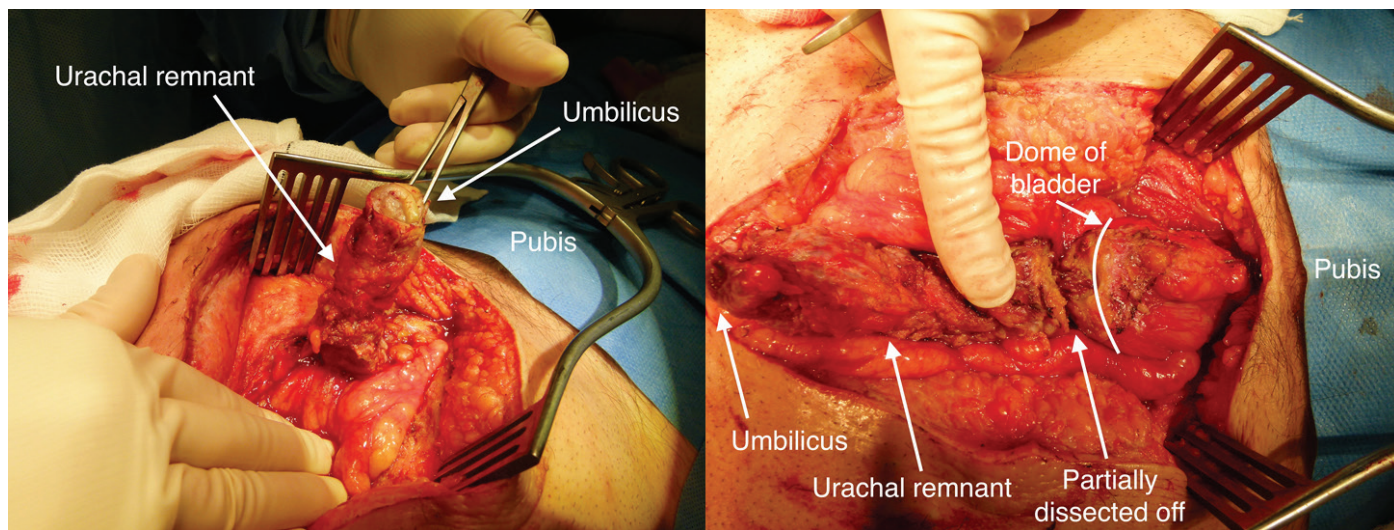


Figure 2: Midline laparotomy incision. Umbilicus dissected free and lifted with urachal remnant in continuum extending towards pubis in the pre-peritoneal plane. The urachal remnant joins the bladder at the apex, it was dissected off with a cuff of bladder tissue.



The patient underwent an open limited right hemicolectomy via a midline laparotomy, resection of the diseased segment of small bowel with excision of the umbilicus, urachal remnant and a cuff of bladder (Figure 2).

Intra-operative findings on laparotomy and histology were consistent with active Crohn’s disease. He made an uncomplicated recovery, progressing well at follow-up.

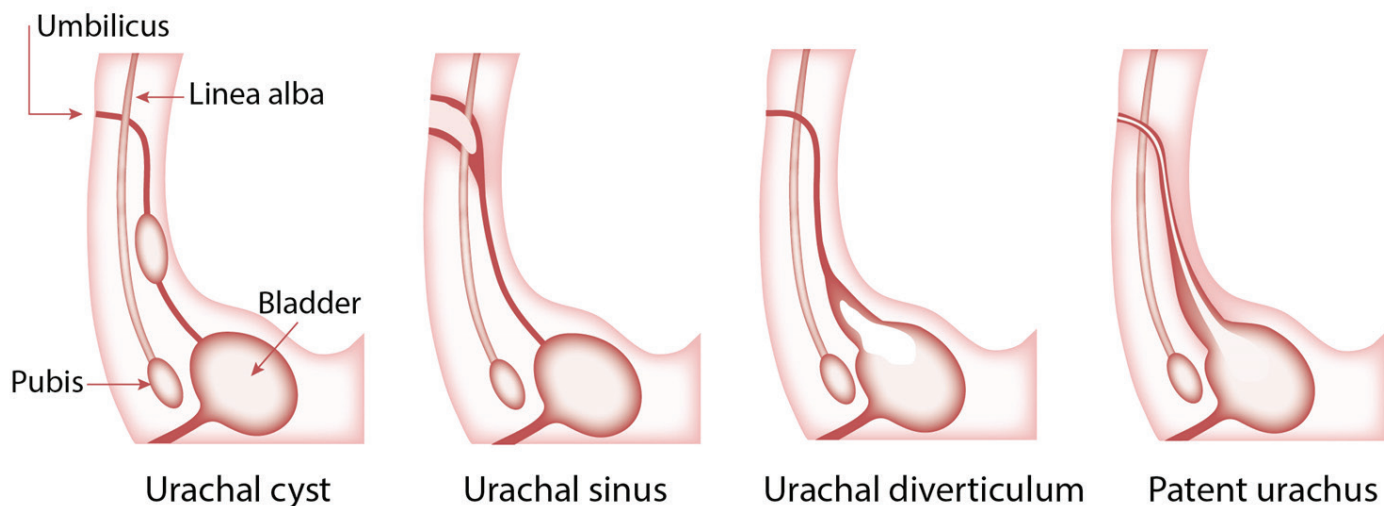
Discussion

Urachal remnants are rare, although the true prevalence of urachal remnants is unclear. A Japanese study included more

than 3,000 child and 40,000 adult abdominal ultrasounds performed in hospital. They found evidence of urachal remnants in 1.6% of children and 0.063% of adults.¹ In contrast, another small study identified urachal remnants in 32% of adults at post-mortem with a 2:1 male to female ratio.²

Urachal remnants may be categorised by the degree of patency. A urachal cyst is an open segment within the structure which is closed off at both ends and the most common presentation, a urachal sinus is a patent segment opening only into the umbilicus and a urachal diverticulum is a segment opening only into the bladder.

Figure 3: Classification of urachal remnants.



A patent urachus is a persisting canal throughout its entire length; it may also be a result of recanalisation due to urinary obstruction, in this case urine may leak from the umbilicus.

Most urachal remnants are asymptomatic, although recognised complications include urachal infections (most common), recurrent urinary infections, urinary calculi, fistulae and malignancy. Malignancy is typically adenocarcinoma despite the transitional cell urachal epithelium. Urachal cancer has a poor prognosis as presentation is often at an advanced stage, five-year survival is estimated to be around 50%.³

Case reports of fistulae between bowel and urachal remnants usually relate to Crohn's disease; other published causes include diverticulitis and appendicitis.⁴ Symptomatic umbilical remnants typically require surgical resection. Open,

laparoscopic and robotic approaches can be utilised.⁵ There is some evidence with infected urachal cysts and sinuses, performing a two-stage procedure may be advantageous to initially control the sepsis then separately resect the remnant.⁶

For asymptomatic structures, the risk of future malignant transformation is believed to be low. An estimation from local data in Toronto, Canada by Gleason et al found 5,721 excisions in asymptomatic children are needed to prevent one case of urachal adenocarcinoma.⁷ However, the value and optimal method of surveillance is also unclear; one study in adults recommended interval ultrasound with cystoscopy and cross-sectional imaging at the time of diagnosis.⁸ In the absence of formal guidelines and with limited evidence available, management plans need to be developed on a case-by-case basis.

Competing interests:

Nil.

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