Urachal remnant causing umbilical in-drawing during micturition

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ABSTRACT

The urachus is a vestigial remnant of the allantois, which is normally obliterated during fetal life to become the median umbilical ligament, which runs between the urinary bladder and umbilicus in adults. Failure of obliteration leaves a tubular urachal remnant, which may present with disease. We report a unique case of a urachal remnant causing umbilical pain and in-drawing on micturition in a nine-year-old boy. There was no urine discharge from the umbilicus and in-drawing did not occur on defecation. His urinary stream was normal. High frequency ultrasonography revealed a thick band with a narrow, anechoic, fluid filled central channel. Exploration via an infraumbilical curvilinear incision identified a thick urachal band that could be traced to the dome of the bladder. This was excised flush with the bladder. The patient remains well at nine months following surgery with complete cessation of symptoms.

KEYWORDS

Urological surgical procedures – Child health – Urogenital abnormality

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The urachus is a vestigial remnant of the allantois, a caudal structure that handles liquid waste during embryonic development. The allantois normally becomes a fibrous cord, the median umbilical ligament, which runs between the urinary bladder and the umbilicus.¹ Failure of obliteration leaves a tubular urachal remnant. This uncommonly presents as a patent urachus with umbilical urine discharge, an umbilical sinus or a urachal cyst,² which may become infected. Malignancy is rare and the most common histological type is adenocarcinoma.³ We present a unique case of a urachal remnant where detrusor contraction during micturition caused umbilical in-drawing and pain.

Case history

A previously healthy nine-year-old boy presented with an unusual one-year history of umbilical pain and umbilical in-drawing during micturition. There was no urine discharge from the umbilicus and in-drawing did not occur at any other time, including on defecation. There was no increased urinary frequency and his urinary stream was normal.

High frequency ultrasonography revealed a thick urachal band, which could be traced from the bladder apex to the umbilicus, with a narrow, anechoic, fluid filled central channel. Figure 1 demonstrates the relation of the urachus and the bladder. Figure 2 shows the thick urachal band in the midline.

Owing to the distress caused by the condition, surgery was planned. Exploration was undertaken via an infraumbilical curvilinear incision. This identified a thick urachal band, which was traced to the dome of the bladder (Fig 3), consistent with the ultrasonography findings. This was excised flush with the bladder (Fig 4). Microscopy identified a fibromuscular tube lined with glandular epithelium

Figure 1 Ultrasonography demonstrating a thick urachal band connecting the umbilicus to the bladder

Figure 2 Ultrasonography showing the urachal band deep to the rectus muscles

Figure 4 Urachus divided from umbilicus and traced down to bladder (view from head end of patient)

Figure 3 Urachus isolated at the umbilicus

consistent with a urachal remnant. The patient remains well at nine months following surgery with complete cessation of symptoms.

Discussion

During weeks 4–7 of development, the urogenital sinus forms from the cloaca. The proximal urogenital sinus forms the bladder and is continuous with the allantois, which runs to the umbilicus.¹ The allantois is obliterated, leaving a fibromuscular cord of tissue, the urachus, which becomes the median umbilical ligament in adults. The lining of the urachus may be transitional or columnar epithelium and its outer layer is muscular, continuous with the detrusor.²

Abnormalities associated with the urachus may be congenital or acquired. Congenital failure of obliteration leading to patency of the urachus is a relatively common

anatomical variation, which one series identified in a third of adult postmortem examinations.⁴ Urachal patencies may close after birth but reopen as a result of pathological conditions.² The degree and position of patency determines how a patent urachus may present. A completely patent urachus creates a fistula between the umbilicus and bladder, which may present with urine discharge at the umbilicus. Blind ended patency at one extreme of the urachus results in an umbilical-urachal sinus, which may also produce discharge, or a vesicourachal diverticulum in the bladder, which may be accompanied by abdominal muscle deficiency syndrome (prune belly syndrome).² An area of patency that is closed at both ends results in a urachal cyst, which may become infected and present as an acute abdomen.

Carcinoma of the urachus is rare, representing 0.35–0.7% of all bladder cancers.⁵ It typically presents with haematuria (in 73% of patients), which may be accompanied by abdominal pain (14%), dysuria (13%) and mucosuria (10%).⁵ The most common histological type is adenocarcinoma. Urachal carcinoma presents at a late stage and carries a poor prognosis, with a five-year survival rate of around 50%.⁵

Infected urachal cysts and urachal carcinomas may be difficult to differentiate on ultrasonography or computed tomography.² Owing to the high risk of reinfection of umbilical cysts, complete urachectomy is the mainstay of therapy. *En bloc* urachectomy and umbilectomy with clear surgical margins accompanied by partial or radical cystectomy is recommended in urachal carcinoma (along with chemotherapy) because of the typically late presentation of urachal carcinoma and frequent recurrence.⁵ It is therefore important to understand the structure and relations of the urachus.

The described case appears to be unique in the literature as a presentation of a congenital urachal abnormality. It seems likely that detrusor contraction during micturition acted via the muscular outer layer of the urachus and put tension on the umbilicus, causing in-drawing and associated pain. This striking presentation illustrates the course of this vestigial remnant.

Conclusions

This report describes a unique case of a urachal remnant where detrusor contraction during micturition caused umbilical in-drawing in a nine-year-old boy. Ultrasonography proved to be useful for diagnosis and preoperative planning, and excision *in toto* caused complete resolution of the symptoms. This is the first report of a urachal remnant presenting in this way, and it highlights the anatomical course and relations of this structure.

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