Intestinal obstruction caused by a Meckel's diverticulum adherent to an infected urachal cyst

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Urachal remnant diseases are rare and frequently misdiagnosed, as the clinical presentation is similar to more common causes of intra-abdominal and pelvic disease. We report the rare case of a 1½-year-old boy with small-bowel obstruction caused by an adhesion band between a Meckel's diverticulum and an infected urachal cyst. The condition was initially misdiagnosed and treated as a periappendiceal abscess on the basis of clinical examination and ultrasonography findings. In addition to the case report, we present an overview of urachal diseases. *Ann Pediatr Surg* 10:139–141 © 2014 Annals of Pediatric Surgery.

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Introduction

Embryologically, the urachus is an open connection between the fetal bladder and the allantois. Normally, the urachal lumen obliterates from approximately gestational week 6, but in some individuals, this obliteration is incomplete or entirely absent [1,2]. A patency of the embryologic urachal remnant may give rise to several clinical problems including cyst, sinus tract, diverticulum, infection, and malignancy [3]. The rarity of urachal abnormalities and diverse clinical presentations lead to frequent misdiagnosis. In the present case, a Meckel's diverticulum (MD) was adherent to an infected urachal cyst, causing small-bowel obstruction.

Case

A 1½-year-old boy with no medical history presented at the Department of Paediatric Surgery with a 1-day history of abdominal pain, vomiting, reduced appetite, and fever. He did not have complaints of constipation, diarrhea, or dysuria. On initial examination, he had tenderness in the lower abdomen, without guarding or rebound. Bowel sounds were normal. The white blood cell count was 30.3×10^9/l and C-reactive protein was 70 mg/l. Urinalysis was normal.

An ultrasonography scan (US) demonstrated a 2.8×2.9 cm cavity located in the right fossa behind the bladder (Fig. 1). The appendix was not visualized. The radiologist interpreted the cavity as consistent with a periappendiceal abscess. An abdominal radiograph demonstrated only meteorism. The patient was treated with an US-guided drainage supplemented by 1 week of antibiotics and was discharged in his habitual condition.

Two days after discharge, the boy returned to the department with continuous complaints of vomiting, abdominal pain, and reduced defecation and flatus. At this time, fever was absent and laboratory results were normal. An acute abdominal radiograph demonstrated mechanical bowel obstruction. A new US showed no intraperitoneal free fluid or residual abscess. On laparotomy, a normal appendix was identified. Further exploration demonstrated distended small intestines extending to an MD, which was connected to an infected urachal cyst by an adhesion band. The small intestines were strangulated by the adhesion band. The urachal cyst and the MD were excised *en bloc* (Fig. 2), and the ileum was closed. Even though we found no lumen between the urachal cyst and the bladder, an open catheter was placed in the bladder for 3 days. The postoperative course was uneventful, and the boy was discharged after 5 days. Subsequent histopathological analysis revealed an MD with lymphoid hyperplasia. The urachal cyst was lined with hyperplastic gastric mucosa and partial small-intestine mucosa with peptic ulcerations and underlying necrosis and inflammation. The urachal cyst was connected to the MD by a dense, fibrotic, edematous, and well-vascularized adhesion band. There was no sign of malignancy. Furthermore, the pathologist found no urothelium or squamous epithelium in the specimen.

Discussion

The urachus is an embryological remnant of the fetal bladder, a canal that connects the fetal bladder with the allantois. The length varies from 3 to 10 cm and diameter from 8 to 10 mm. The urachus has no known function. The descent of the bladder between the fourth and the fifth months of development probably stretches the urachus, causing its lumen to obliterate and become the median umbilical ligament that connects the vesical apex to the umbilicus. Some authors argue that the obliteration of the lumen is due to segmental obstruction by the desquamated epithelium [1,2]. Histologically, the urachus is composed of three layers: in 70% of the cases, the lumen is lined with a transitional cell epithelium, and in the rest of the cases, a columnar epithelium lines the lumen; this forms the innermost layer, which is followed by a middle connective tissue and the outermost smooth muscle layer [1,4]. Involution of the tubular urachus normally occurs before birth, but occasionally, the obliteration is incomplete and results in urachal

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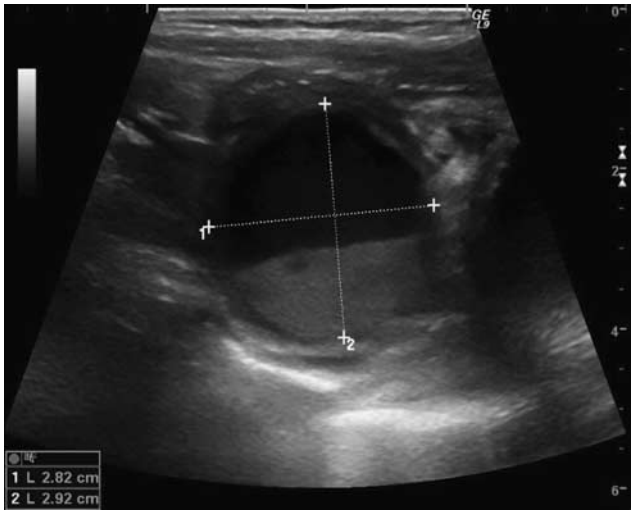
Fig. 1



Ultrasonography scan of the 2.82 × 2.92 cm urachus located just in the right posterolateral region of the bladder.

abnormalities. The overall prevalence of persistent urachal remnants varies within the literature. Schubert *et al.* [4] found urachal remnants in 32% of adult autopsy specimens. However, the prevalence of clinically relevant urachal abnormalities seems to be much rarer. These urachal abnormalities can be divided into four main types according to the patency of the duct: the patent urachus also called the urachal fistula, the urachal sinus, the vesicourachal diverticulum, and the urachal cyst [1,5]. A patent urachus accounts for 10–15% and usually presents in the neonatal period with urine discharge from the umbilicus. The incidence of patent urachus is one in 761 in autopsy studies [1]. The latter three types of urachal remnants may be closed at birth, but then reopen under pathological conditions, and therefore are also categorized as acquired disease [6]. Urachal cysts account for 29–43% and are found in 1 : 5000 in autopsy series [1]. Most commonly, urachal cysts present as a secondary infection. The route of infection may be lymphatic, hematogenous, or vesical [7]. The clinical presentation of urachal

Fig. 2

Urachus cyst adherent to the Meckel's diverticulum.

abnormalities varies, but symptoms of infected cysts typically include abdominal pain, tenderness, fever, nausea, vomiting, and leukocytosis. Noninfectious presentations of urachal cysts include progressive enlargement, calculus formation, intraperitoneal/extraperitoneal rupture, malignant transformation, fistula formation, acute hemorrhage, mechanical urinary obstruction, intestinal obstruction, and the development of Reiter syndrome [8]. Typically, symptoms related to all of these complications are misdiagnosed as appendicitis, intra-abdominal or pelvic abscess, strangulated umbilical hernia, acute prostatitis, MD, urinary tract infection, pelvic inflammatory disease, and bladder carcinoma [9–12]. Because of the location of the urachal remnant posterior to the anterior abdominal wall with no interfering structures, US is regarded as a suitable imaging modality for the diagnosis. The success rate of US has been reported to range from 75 to 100% [8]. When in doubt, a computed tomography or MRI should be performed [13].

Surgical excision is the treatment of choice due to the high rate of recurrence with other modalities [8]. Complete excision is essential due to a 30% reinfection rate if the urachal remnant is not completely resected. Furthermore, urachal remnants may transform into aggressive urachal malignancy in adulthood [14]. In view of the poor prognosis of urachal cancer patients, of whom up to 20% have metastases at debut of symptoms, surgical intervention should be performed at the first diagnosis. Copp and colleagues, in contrast, proposed that only incidentally detected urachal remnants lined with an epithelium should be excised, as remnants with no epithelium carry little risk of malignant transformation. However, asymptomatic urachal remnants are as likely as symptomatic ones to have epithelial elements. Therefore, it is not possible to predict the risk of malignant transformation on the basis of the presentation [15].

Although the urachal remnant and MD have a common embryological origin from the endoderm in the yolk sac, a synchronous presentation of the two congenital abnormalities is extremely rare. A few cases of intestinal obstruction caused by a urachal remnant have been published [6]. In these cases, the obstructions were caused by entrapment of the small bowel in the opening between an intraperitoneally located infected urachal cyst and the abdominal wall [6].

Conclusion

Urachal abnormalities are rare and may cause considerable morbidity. Detailed history, careful examination, knowledge of the embryological anatomy, and ultrasonography expertise are important to ensure an early and accurate diagnosis. The clinical presentation is often diverse and nonspecific. US is sometimes inconclusive, and in such cases, computed tomography or MRI should be considered. When a symptomatic urachal remnant is diagnosed, complete excision is recommended due to the risk of recurrence and of later urachal malignancy. In our opinion, complete excision is also the treatment of choice regarding incidentally found urachal abnormalities.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

References

1 Nguyen HT, Cilento BG Jr. Bladder diverticula, urachal anomalies, and other uncommon anomalies of the bladder. In: Gearhart JP, Rink RC, Mouriquand PDE, editors. *Pediatric urology*. Philadelphia: Saunders; 2010. pp. 417–422.
2 Sadler T. Urogenital system. In: Sadler T, editor. *Langman's medical embryology*. Philadelphia: Lippincott Williams & Wilkins; 2010. pp. 243–245.
3 McCollum MO, Macneily AE, Blair GK. Surgical implications of urachal remnants: presentation and management. *J Pediatr Surg* 2003; **38**: 798–803.
4 Schubert GE, Pavkovic MB, Bethke-Bedürftig BA. Tubular urachal remnants in adult bladders. *J Urol* 1982; **127**:40–42.
5 Blichert-Toft M, Nielsen OV. Congenital patient urachus and acquired variants. Diagnosis and treatment. Review of the literature and report of five cases. *Acta Chir Scand* 1971; **137**:807–814.
6 Blichert-Toft M, Nielsen OV. Diseases of the urachus simulating intra-abdominal disorders. *Am J Surg* 1971; **122**:123–128.
7 MacNeily AE, Koleilat N, Kiruluta HG, Homsy YL. Urachal abscesses: protean manifestations, their recognition, and management. *Urology* 1992; **40**:530–535.
8 Allen JW, Song J, Velcek FT. Acute presentation of infected urachal cysts: case report and review of diagnosis and therapeutic interventions. *Pediatr Emerg Care* 2004; **20**:108–111.
9 Ash A, Gujral R, Raio C. Infected urachal cyst initially misdiagnosed as an incarcerated umbilical hernia. *J Emerg Med* 2012; **42**:171–173.
10 Goldberg R, Pritchard B, Gelbard M. Umbilical inflammatory conditions: case report and differential diagnosis. *J Emerg Med* 1992; **10**:151–156.
11 Goldman IL, Caldamone AA, Gauderer M, Hampel N, Wesselhoeft CW, Elder JS. Infected urachal cysts: a review of 10 cases. *J Urol* 1988; **140**:375–378.
12 Nimmonrat A, Na-ChiangMai W, Muttarak M. Urachal abnormalities: clinical and imaging features. *Singapore Med J* 2008; **49**:930–935.
13 Yu JS, Kim KW, Lee HJ, Lee YJ, Yoon CS, Kim MJ. Urachal remnant diseases: spectrum of CT and US findings. *Radiographics* 2001; **21**:451–461.
14 Ashley RA, Inman BA, Routh JC, Rohlinger AL, Husmann DA, Kramer SA. Urachal anomalies: a longitudinal study of urachal remnants in children and adults. *J Urol* 2007; **178 (Pt 2)**:1615–1618.
15 Copp HL, Wong IY, Krishnan C, Malhotra S, Kennedy WA. Clinical presentation and urachal remnant pathology: implications for treatment. *J Urol* 2009; **182 (Suppl)**:1921–1924.