Radiology Case Reports 14 (2019) 1181–1184

Available online at www.sciencedirect.com

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journal homepage: www.elsevier.com/locate/radcr

Case Report

The primary considerations and image guided diagnosis of an infected urachal cyst in a pediatric patient

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ARTICLE INFO

Article history:
Received 6 June 2019
Revised 24 June 2019
Accepted 26 June 2019
Available online 23 July 2019

Keywords:
Pediatric radiology
Diagnostic radiology
Ultrasound
Urology

ABSTRACT

Urachal cyst is a rare condition that is typically asymptomatic and will often have symptomatology that is misdiagnosed or missed. A urachal cyst occurs in 1 out of 5000 live-births, but is only clinically relevant in 1 out of 150,000 of the population often as an incidental finding. The urachus is the embryological remnant of the allantois, which connects the apex of the bladder to the umbilicus, and usually fully obliterates to become the median umbilical ligament.

Urachal defects are uncommon and cysts are usually asymptomatic until infection results. An infected cyst may present mimicking a wide range of intra-abdominal and pelvic disorders, and accurate diagnosis is often delayed. Children may present with umbilical discharge; adults often have hematuria. Computed tomography (CT) and ultrasound are ideally suited for demonstrating urachal remnant diseases; however, infected urachal cysts commonly display increased echogenicity with ultrasoundsography and thick-walled cystic or mixed attenuation with CT. Drainage and excision of the urachal remnant is the definitive treatment.

Case report

A 6-year-old obese Hispanic boy with no significant past medical history presented to the emergency room with a 2-month history of intermittent abdominal pain associated with diarrhea. His visit to the emergency department was prompted by a reported a 2-day history of loose stools as well as dysuria. His mother described it as ongoing intermittent periumbilical pain. The abdominal pain was nonradiating without alleviating or aggravating factors and was associated with a tactile fever. His physical exam was significant for abdominal tenderness and guarding upon palpation of both right and left lower quadrant. The child was admitted with partial small bowel obstruction due to intussusception as the primary consideration.

His laboratory work demonstrated nonspecific leukocytosis and elevated inflammatory markers with an unremarkable basic metabolic panel.

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https://doi.org/10.1016/j.radcr.2019.06.012

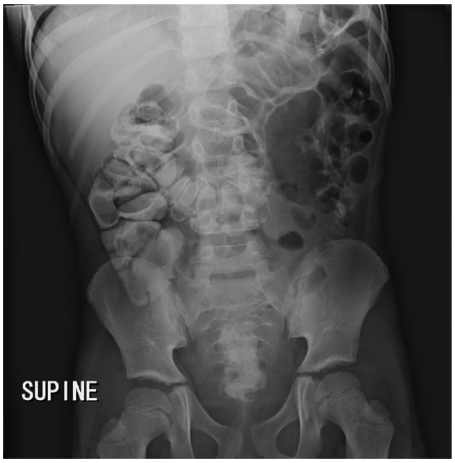

Fig. 1 – Supine abdominal radiographs of the 6-year-old male diagnosed with an infected urachal cleft cyst. A nonspecific bowel gas pattern with no evidence for free air demonstrated. Small volume residual enteric contrast opacifies several loops of bowel in the right lower quadrant.

As the clinical suspicion favored small bowel obstruction secondary to intussusception though the differential remained somewhat extensive, abdominal computed tomography (CT) with contrast was obtained following a negative abdominal radiograph (Fig. 1). The exam delineated nonspecific increased soft tissue superior to the urinary bladder (Fig. 2), for which neoplasm, perforated appendicitis, or nonspecific inflammatory process were the primary considerations. Ultrasound was recommended to further characterize the lesion. The ultrasound of the abdomen (Fig. 3) revealed a midline soft tissue mass superior to the dome of the bladder consistent with an inflammatory process measuring 3.86 cm × 2.1 cm (Fig. 2). The patient began a regimen of antibiotics after pediatric infectious disease was consulted, which failed to resolve the symptomatology, necessitating a pediatric urology consultation. The diarrhea, unusual given the differential, desisted shortly after admission, and was believed to reflect a self-limiting viral illness.

Cystoscopy demonstrated an indentation along the anterior bladder dome, typical of a urachal cyst, which precipitated an anterior abdominal wall incision and direct visualization of a cystic lesion arising from the bladder dome, consistent with a urachal cleft cyst. A single stage surgical excision of the lesion was performed. The procedure was tolerated with no complications. The specimen expressed purulent material with microabscesses, which when cultured was found to be negative for any micro-organisms. Postoperatively the patient received antibiotics for 7 days as an inpatient. The patient was discharged after his antibiotic therapy regimen with scheduled follow-up with pediatric urology and infectious disease clinics.

Imaging

Ultrasonography is the preferred modality for diagnosing a urachal cyst given the extra-peritoneal location of the abnormality, the ability to determine the presence of any potential communication with the bladder, and absence of harmful radiation exposure. However, CT is often obtained given the nonspecific presentation to exclude alternative diagnoses. The typical CT appearance is a simple fluid attenuating lesion immediately posterior to the abdominal wall, often contiguous with the bladder dome. When infected, it may mimic a soft tissue mass, for which bladder adenocarcinoma is a necessary consideration. This is particularly important given the predisposition of adenocarcinoma due to prolonged urinary stasis and infection of urachal cysts. The sensitivity and specificity of ultrasound are not well established in the presence of concomitant inflammation or complicated disease. A bladder diverticulum, mesenteric cyst, or less

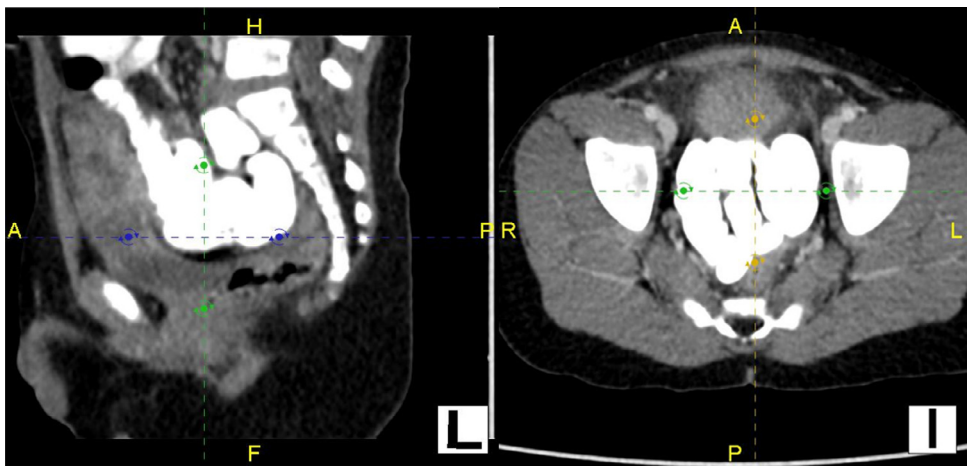

Fig. 2 – Sagittal and axial reconstructed images at the level of the ischial tuberosities of the CT of the abdomen and pelvis enhanced with IV and enteric contrast. A soft tissue density is demonstrated superior to the bladder dome with infiltration of the adjacent fat planes. It appears to extend superiorly to and is questionably contiguous with the umbilicus. The bladder, while underdistended, displays a grossly smooth wall contour.

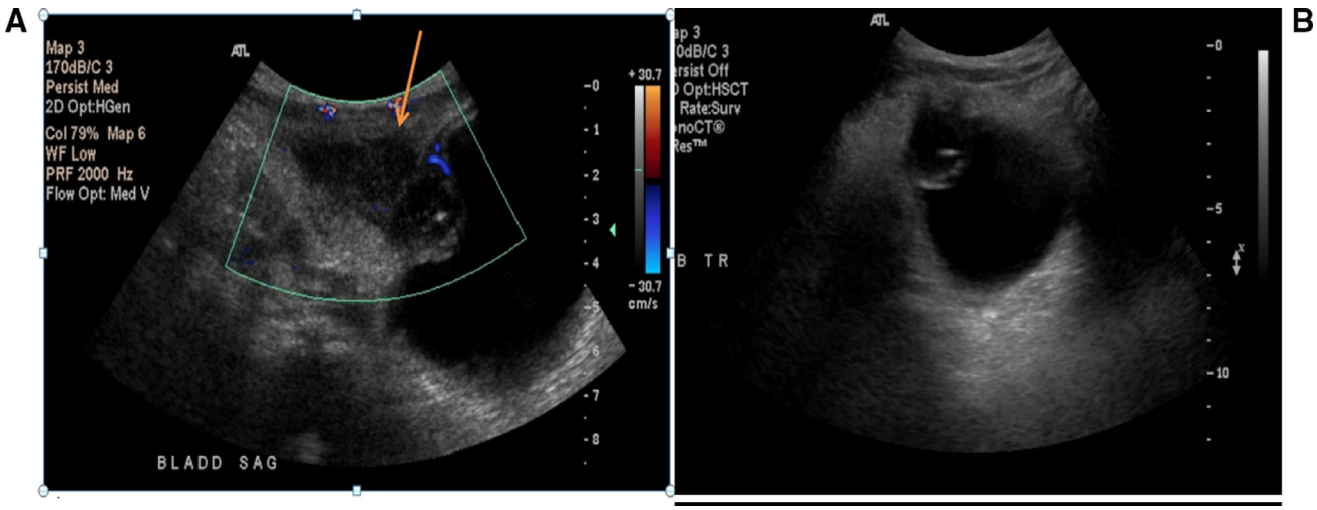

Fig. 3 – Midline sagittally oriented images at the level of the bladder dome obtained during ultrasound of the abdomen and pelvis utilizing color Doppler and grayscale images. (A) Heterogeneous lesion extends superiorly from the bladder dome lacking internal vascularity. This was a key finding in distinguishing neoplasm from an inflammatory reaction. (B) Intravesicular echogenic lesion abutting the bladder dome; this was felt to represent the extension of the inflammatory reaction into the bladder lumen rather than an intrinsic bladder wall lesion.

likely Meckel's diverticulum may mimic its appearance. In the case of a nondiagnostic ultrasound, MRI is an acceptable alternative. [1] Fistulography, though not performed in our case, may be useful in ruling out the presence or determining the extent of any communication with the gastrointestinal tract, urinary tract, or umbilicus preoperatively. [2]

Management

Drainage of the infected abscess and excision of the urachal remnant is the definitive treatment. [1,4,8,9] The preferred surgical treatment method remains a subject of debate. The single-stage excision involves a primary excision of the infected urachal cyst and bladder cuff, whereas the two-stage procedure involves a primary incision and drainage, followed by a later excision of the urachal remnant and bladder cuff after ensuring that any potential infection was treated. The two-stage procedure produces a shorter postoperative hospital stay with no complications. However, in the case of small and localized infections, a single-stage excision can be considered in young adults without comorbidity. A single-stage excision procedure was opted over a two-stage procedure in our case. After undergoing the single stage procedure, this patient was followed in outpatient pediatric urology and infectious disease clinics and no complications were reported. [3]

Discussion

A urachal cyst is one of several potential bladder defects which form when the lumen of the intraembryonic portion of the allantois fails to obliterate after fetal development [1,4]. The persistence of the central portion of the urachus defines a urachal cyst, vs a sinus or fistula [5]. This lesion may be found incidentally on imaging in asymptomatic older children and, more frequently, in adults with infection. Normally the allantois, the precursor tissue to the urachus, obliterates during embryogenesis becoming a fibrous vestigial cord called the median umbilical ligament, which rests between the umbilicus and the dome of the bladder, by the fourth or fifth month of gestation [6]. When the regression is incomplete, urachal abnormalities can result. Possible types of remnants include patent urachus, urachal cyst, urachal sinus, and urachal diverticulum [7]. Although rare, these anomalies serve as a nidus for abscess formation, cyst rupture leading to peritonitis, uracho-colonic fistula, stone formation, or neoplastic transformation.

A child with a urachal cyst can present asymptomatically as an incidental finding or with nonspecific symptomatology including abdominal pain, palpable mass, fever, and urinary dysfunction [3–7]. The reported incidence of urachal cysts varies among studies. The clinical differential diagnosis of urachal cyst is similarly broad and includes partial bowel obstruction, celiac disease, inflammatory bowel disease, urinary tract infection, abdominal tuberculosis, acute appendicitis, and infectious gastroenteritis. Urachal abnormalities are often misdiagnosed as appendicitis, problematically leading to readmission [8]. A urachal remnant should be suspected in any patient with calcifications or inflammation involving the bladder dome on imaging.

A well-documented complication involves postoperative enterocutaneous fistula which may necessitate an additional operative procedure. Cases of an extensive and severely infected urachal cyst typically involve excision using the single-stage procedure. In our case example, given that the urachal cyst infection was extensive and severe, the 2-stage procedure may have been considered [6]. Urachal cysts are a rare phenomenon that may present with misleading

symptoms suggestive of an acute abdomen. Because the clinical presentation is nonspecific, it is essential to have a high index of suspicion and be familiar with the underlying anatomy to reach the correct diagnosis. A urachal cyst should be considered in patients with periumbilical and suprapubic abdominal pain even in the absence of a discernable mass.

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