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Urachal cyst infection as a cause of acute abdomen: A case report and discussion about controversial issues of its management in children

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ABSTRACT

Defects of the urachus obliteration may lead to urachal abnormalities such an urachal cyst, which tend to rest asymptomatic. We report a case of a four-old-year boy who referred a three-week history of abdominal pain, dysuria and fever. A hypogastric mass was observed at the exploration. The ultrasonography and the computed tomography were compatible with an infected urachal cyst. Intravenous antibiotic therapy was started, with an optimal response. Four months later, a chirurgical complete excision of the urachal cyst was performed. Among different congenital disorders of the urachus, urachal cysts and patent urachi are the most frequent. Urachal cysts may become infected, causing suprapubic pain, dysuria, fever and abdominal mass, as well as signs of peritonism. The main tool for diagnose these lesions is ultrasonography. They could potentially degenerate into adenocarcinoma in adults, which is rare but associated to bad prognosis. Urachal remnants have been classically managed surgically. However, an active surveillance of these lesions with a conservative treatment could be an alternative, since spontaneous resolution is more common than previously thought.

Key Words: Urachal cyst, urachal remnant, children, non-operative management.

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Introduction

The urachus is an embryonal tubular structure which extends from the bladder dome to the posterior umbilicus. It is composed of three layers; an innermost layer of modified transitional epithelium similar to the urothelium, a middle fibroconnective tissue layer, and an outer layer of smooth muscle continuous with the detrusor [1]. The downgrade of the bladder into the fetal pelvis from the fifth month of development pulls the urachus with it, causing its progressive obliteration, remaining as a fibrous cord called median umbilical ligament. Urachal remnants (UR) are secondary to an incomplete urachal obliteration. They tend to remain asymptomatic, except when a complication occurs [2]. Here we report a case of an infected

urachal cyst (UC) in a pediatric patient and we present a bibliographical review focused on its management, which still remains controversial.

Case report

A four-old-year Caucasian child with no relevant medical history applied to our Emergency Department. He had a three-week evolution colic abdominal pain, localized at the hypogastrium and exacerbated with the urination. He presented fever in the last 48 hours, without vomits or diarrhea.

The abdominal examination showed a painful hypogastric mass, without hepatomegaly or splenomegaly. The rest of the physical examination was normal. The urinalysis was normal, while the blood test showed a hemoglobin of 11.8g/dl, leucocytes of 12,500/ml (mature neutrophils 9,400/ml, immature neutrophils 300/ml and lymphocytes 2,100/ml) and a C-reactive protein (CRP) of 110.8mg/L. No germens grew at the blood or the urine cultures. An abdominal ultrasound (US) showed a flux-less, heterogenic, bad-defined mass, which extended from the front of the bladder to the umbilicus (Fig. 1).

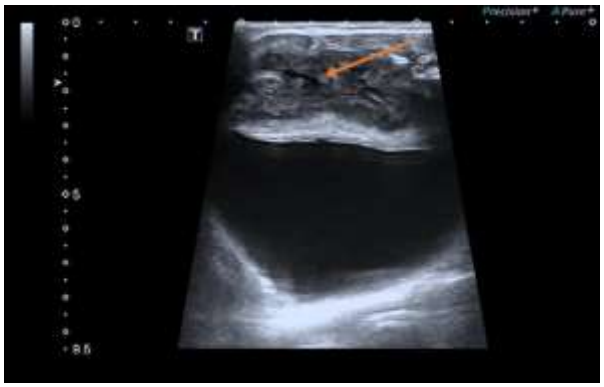

Fig. 1. Ultrasonography. Orange arrow points the infected urachal cyst.

An abdominal Computed Tomography (CT) showed a 50x34x47mm hypodense collection at the anterosuperior bladder angle, with thickened walls. The retropubic space's fat tissue showed signs of inflammation, while the adjacent bladder wall was remarkably thickened. No communication between the collection and the umbilicus was seen (Fig. 2).

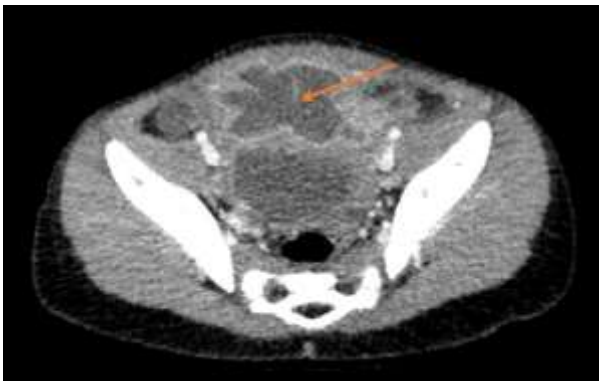

Fig. 2. Computed tomography scan. Orange arrow points the infected urachal cyst.

Suspecting an infected UC, the patient was admitted, receiving intravenous amoxicillin/clavulanic acid (100mg/Kg/day) for five days. At the third day of treatment, the fever had receded, the abdominal pain had sharply improved, and the CRP had decreased to 83mg/L. A new US showed a reduction in collection size (38x20mm) at the fifth day of admission, so he was discharged and continued with prophylactic oral amoxicillin/clavulanic acid (60mg/Kg/day). A magnetic resonance imaging performed after one month showed a persistent mass of 18x9x5mm without signs of inflammation. The patient remained asymptomatic, so the antibiotic prophylaxis was stopped. Four months after the initial symptoms, a chirurgical complete excision of the UC was performed. Through an infraumbilical laparotomy, a longitudinal incision of the aponeurosis was made. Then, the umbilical vein remnant was bilaterally identified, while the urachus was correctly obliterated from the umbilicus through the

cyst. The urachal cyst was correctly identified and completely resected. The intervention was performed without any complications. No residual mass was observed at the ultrasonographic surveillance. Twelve months after the surgery, he remains asymptomatic.

Discussion

The urachus is supposed to progressively obliterate during the fetal life, remaining as a fibrous cord between the transversalis fascia anteriorly and the peritoneum posteriorly, attaching the umbilicus to the bladder dome [3]. With the improvement of US techniques, it has been observed that its obliteration delay is more common than previously thought [4]. In fact, up to 50% of newborns may have a permeable urachus, comparing to 2% of adults. Nevertheless, it is clinically relevant in only 2 every 300,000 newborns [2,5]. Meanwhile, from autopsy studies, it was estimated that 1 every 5,000 newborns may have a UC [5].

UR are generally divided into four main categories: patent urachi (communication between the bladder and the umbilicus), umbilical sinuses (Urachus' opening into the umbilicus), vesico-urachal diverticula (urachus' wide patent opening into the bladder) and urachal cysts (presence of a cyst-like structure within urachus' length) [2,6]. In pediatric patients, the most frequent disorders are UC (30-54% of cases) and patent urachi (13-50%) [6–8].

Symptomatic patent urachi debut at the newborn stage manifesting as umbilical drainage [9,10]. However, UC tend to be asymptomatic unless a complication occurs. Infections are the most common complications, caused mainly by direct migration from the umbilicus or the bladder [11]. The most frequently cultured bacteria are *Staphylococcus aureus* [2,5,7,11–13].

Clinical manifestations of infected UC include acute suprapubic abdominal pain, dysuria, fever [1,2,6,12], vomiting and voiding difficulty [14]. A suprapubic mass might also be observed at the exploration [1,2,4,12], as in our case. Abscesses may drain into the umbilicus, presenting with umbilical drainage [12,15,16], which is the most reported symptom in some series [8,13]. Although it is rare, UC may also drain into the abdominal cavity, resulting in acute peritonitis [4,15,17]. Their differential diagnosis includes other causes of acute abdominal pain like acute appendicitis and Meckel's diverticulum [16], as well as cellulitis, umbilical hernia, and tumors [6].

UC diagnosis is usually based on US. It shows an elliptical, hypoechoic structure in the middle of the anterosuperior surface of the urinary bladder [17]. Only if ultrasonography is inconclusive, CT is indicated [6]. On urinalysis, bacteriuria and pyuria are absent in more than 80% of the cases, and urine culture is generally negative [6].

Seventy-two per cent of UR have epithelial components, which could theoretically degenerate into malignant tissue [15]. Although cylindrical metaplasia of the transitional epithelium may be seen up to 31.6% of individuals with UR [18], malignant degeneration into an adenocarcinoma is rare and occurs almost exclusively in adults older than 55 years old [7]. In fact, no association between UR in childhood and urachal carcinoma later in life has been established [10,13]. However, it is something to bear in mind, especially because these tumors are difficult to diagnose and limited responders to chemo and radiotherapy, so their prognosis is usually bad [15].

If surgical excision of an infected UC is decided, a two-stage procedure should be the

Table 1: Sum up of the main publications that analyze the non-surgical management of urachal remnants.

Main author	Patients and methods	Results
Ueno [17]	Prospective surveillance of 44 children with non-operated UR.	Clinical relapse in one case (2.3%).
Gleason [18]	In 13 years, 64,803 pediatric patients underwent some imaging study. The data was cross-referenced with the incidence of urachal carcinoma.	UR were seen in 667 patients. NNT to prevent one case of adenocarcinoma: 5,721.
Nogueras-Ocaña [12]	Ultrasonographic surveillance of 6 children with infected urachal cysts treated only with antibiotics and 4 with asymptomatic cysts.	Three patients of each group showed a spontaneous resolution (60%). Two (20%) needed a surgical intervention.
Naiditch [8]	Retrospective review of 103 patients with UR. 19 patients (11 symptomatic and 8 asymptomatic) were followed-up with ultrasonography.	Spontaneous resolution in 8/11 (72.7%) symptomatic and in 7/8 (87.5%) asymptomatic remnants.
Dethlefs [10]	Retrospective review of 153 patients diagnosed with a UR. 39 primarily received non-operative management. 118 were finally operated.	12% patients with conservative management failed (the rest of case presented resolution in a median of 35.5 months), whereas 19,5% of operated patients had complications.

NNT: number to treat

election. It is that parenteral antibiotic therapy is performed weeks or months before the surgery [1,2]. This approach is related to less complications and a shorter hospital stage compared to one-stage procedures [14]. However, non-infected UC could be removed in a single-stage excision [2].

Since preventing malignancy is one of the theoretical reasons of surgical treatment, excisions must be imperatively radical. However, umbilectomies could be avoided [16], since malignant changes tend to take place at the junction of the urachal ligament and the bladder dome [19]. Moreover, incomplete resection is associated to 30% risk of reinfection [20]. It is important to remind that some series describe complications in up to 14.7%-20% [8,10] of the patients (mainly infections), some of whom requiring reinterventions. Surgical excision involves a transverse or midline infra-umbilical incision. Recently, some authors defended that a laparoscopic approach is related to a minor risk of wound infection [6], although it has not been related to a reduction of the operation time or the inpatient stay [19].

Whereas in adults surgery is the most defended treatment for UC [6], there is not a global consensus about its management in children [12,18]. Previously, general recommendation was a surgical excision for children older than 6 months, considering the high probability of spontaneous resolution in younger infants [16]. However, since the US technique has strongly improved, more incidental diagnosis could be made, whereas performing imaging surveillance is expected to become easier [8,17].

Lately, more and more authors defend an active surveillance as a strong alternative to universal intervention. According to some of them, asymptomatic non-infected UC should be surgically removed, while infected UC could be managed with only antibiotics and periodically controlled by ultrasound [4]. Conversely, a Japanese group showed a relapse of only 2.3% of non-operated patients [17] (see Table 1).

In an Ontarian tertiary pediatric center, UR were incidentally seen in 1.03% of patients who underwent an abdominal image study for any reason. They estimated a number needed to treat to prevent a single case of adenocarcinoma of 5,721 (see Table 1) [18]. This shows that treating all incidentally found UC could imply an important overtreatment. According to this, some authors argue that continuous observation with periodic ultrasound examinations could not necessary for asymptomatic cases [17].

Other studies have performed ultrasonographic surveillance of both infected and non-infected UC. A Spanish group followed a cohort of children with UC. After 5 years of follow-up, 60% of them showed spontaneous resolution and just 20% needed a surgical intervention because of a reinfection [12]. An American group observed spontaneous resolution in 78.9% of the cases [8]. Both groups described spontaneous resolution both in infected and asymptomatic UC (see Table 1).

Thus, surgical resection could be restricted to patients with multiple symptomatic episodes who are older than one year, since spontaneous resolution is so likely in younger patients [17]; both in infected and non-infected remnants and regardless of the UR type [4,8,14]. Initial drainage of infected UC may help their non-surgical resolution [4].

Conclusion

We would like to highlight that UC infections may be confused with other causes of acute abdomen, since signs of peritonism may be

present. If the triad of tender midline infraumbilical mass, umbilical discharge and sepsis were present, an infected UC should be suspected. Its surgical excision has traditionally been the treatment of choice. However, surveilling these patients is an alternative, since the risk to benefit ratio for resection is not clear. Nevertheless, if a urachal anomaly is not resected, patients and their families should be aware of a potential future risk of infection and the need of screening.

Compliance with ethical statements

Conflicts of Interest: None.

Financial disclosure: None.

Consent: We obtained the informed consent from the patient's parents to report this case.

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