CASE REPORT

Large serous urachal cyst in an adult

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Lesions of the urachus are rarely manifested clinically in adulthood. They more commonly cause clinical problems in children. The urachus is obliterated in early infancy and its remains persist as the medial umbilical ligament. The lumen of the lower part of the urachus may persist throughout life and communicate with the cavity of the bladder[1]. Persistent urachal remnants are most likely a normal phenomenon and clinically important only when complicated by infection, neoplasia or cystic dilatation[2].

We present an adult patient with an unusually large urachal cyst causing hydronephrosis of the right kidney.

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A 33-year-old male patient presented with a 24-month history of slowly progressive abdominal distention and mild discomfort. He had no fever and admitted with no clinical symptoms related to the urinary or gastrointestinal system. On examination he was hemodynamically stable, well hydrated, with distended abdomen (Fig 1). A large, tense, painless mass was palpated, occupying the whole abdomen. Hematological and biochemical tests were normal and urine analysis was unremarkable.

Computed tomography examination of the abdomen (Fig 2) showed a large well circumscribed fluid filled cyst with thin wall, occupying the whole abdomen (dimensions: rostrocaudal 25 cm, anteroposterior 15 cm, lateral 23 cm). The origin of the cyst could not be resolved by the radiologic examination.

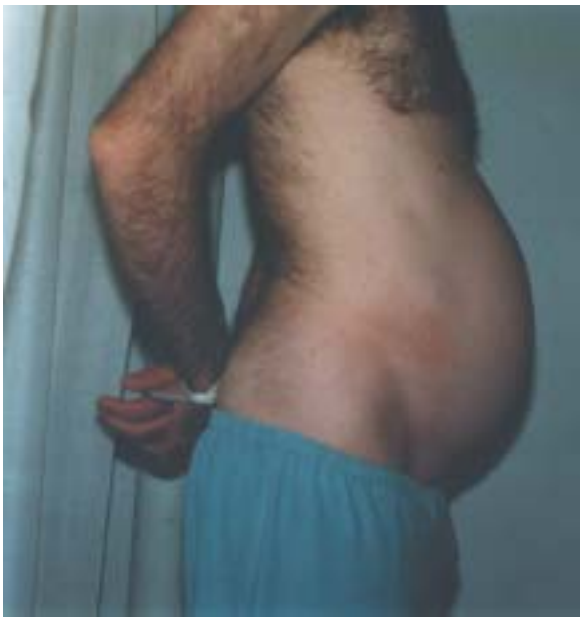

Fig. 1. Lateral view of the patient.

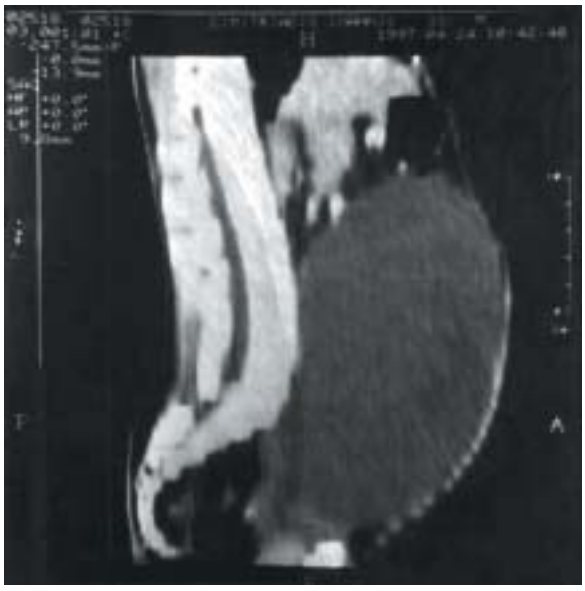

Fig. 2. Abdominal CT scan, median sagittal view. The cyst extents from the anterior abdominal wall to the spine posteriorly causing hydronephrosis of the right kidney and displacement of the bowel posteriorly and laterally.

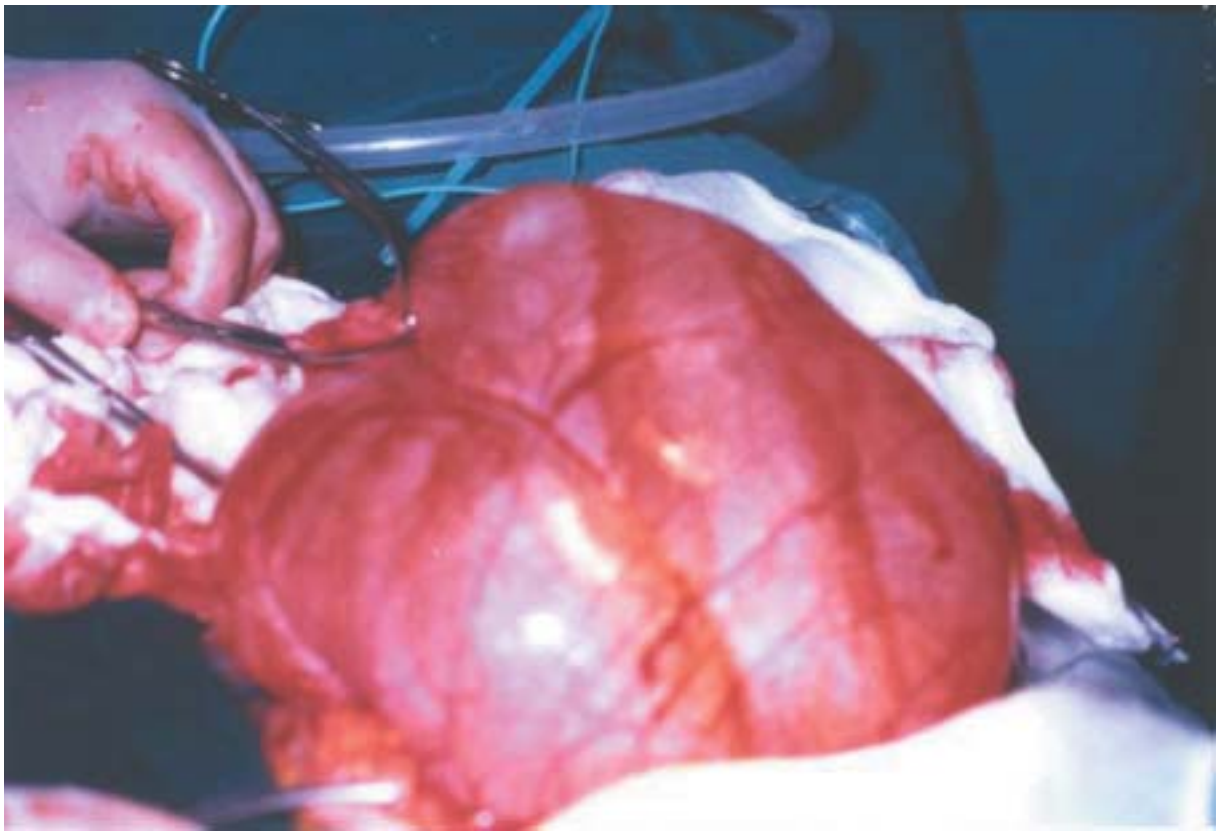

Fig. 3. Itraoperative transilluminated view of the cyst.

Fig. 4. Hematoxylin-Eosin X100. The cyst is lined by simple cuboidal epithelium and has a thin submucosal layer of connective tissue and a fibromuscular wall. This type of layering is reminiscent of the normal urachus.

An IV urography showed delayed urinary excretion and hydronephrosis of the right kidney due to compression of the right ureter by the cyst.

At laparotomy a large, well circumscribed, transluscent cyst filled with clear serous fluid was seen attached to the umbilicus, the anterior abdominal wall below the umbilicus, and the dome of the urinary bladder (Fig 3). It did not have any attachement to any other intraabdominal organ. A total excision of the cyst with part of the attached dome of the urinary bladder was performed. The patient had a uneventfull postoperative course and was discharged on the 8th postoperative day.

On pathologic examination a portion of the muscular bladder wall and the attachment to the umbilicus were identified on the outer surface of the cyst. The two medial umbilical ligaments were identified along the anterior wall of the cyst. The cyst had a smooth inner lining. It had a thin fibromuscular wall and was lined by a simple cuboidal epithelium (Fig 4). The epithelium was intact. The epithelial cells had, small round hyperchromatic nuclei, with no atypia, little cytoplasm and showed small paranuclear vacuoles, negative for PAS and alcian blue. Mitoses were not seen. They expressed cytokeratins of low and high molecular weight and were negative for epithelial membrane antigen, vimentin, S100 protein, and carcinoembryonic antigen. Sections of the excised urinary bladder wall did not show any urachal remnants.

Comment

The urachus is the embryologic remnant of the allantois and the adjoining ventral cloaca. It has a tubular structure and its lumen becomes obliterated with advancing age. However, in a small proportion of the adults (2%), patency with the urinary bladder persists [3]. Urachal lesions can be classified into the patent urachus, urachal sinus, vesicourachal diverticulum, urachal cyst and alternating sinus [4]. In patent urachus its entire tubular structure is intact. In urachal sinus there is drainage to the inferior umbilicus. In vesicourachal diverticulum the lower end of the urachus remains connected to the lumen of the bladder. In alternating sinus, a cyst-like structure retains patency either to the bladder or the umbilicus. Whereas in children the most common type of urachal lesion is patent urachus [5,6] in adults the urachal cysts prevail [4,7]. These are usually small remnants in the lower third of the urachus or the bladder wall and are usually found incidentally in autopsy or cystectomy specimens [2]. They present clinically whenever they enlarge significantly, they become infected [4,8,9,11,12] or a neoplasm develops in them [13-15]. They usually become infected due to persistent communication with the bladder, so the bacteria can gain access to the lumen of the cyst.

If they are not infected, they can attain a very large size like the present case. A case of huge urachal cyst, which contained 55 liters of clear fluid, has been described [16]. The phenomenon of progressive enlargement of these cysts could be explained either by continuous secretory activity of the lining epithelium without drainage or by neoplastic potential of the epithelial cells. In our case, the preservation of the normal architecture of the wall of the urachus and the presence of cytoplasmic vacuoles the epithelial cells are findings suggestive of the enlargement of the cyst being probably due to secretory activity of the lining epithelium. On the other hand, in case of a neoplastic lesion, e.g. a cystadenoma, we would expect to see evidence of stromal reaction, similar to the one seen in serous cystadenomas in other locations, such as ovary, and other morphological signs of proliferative epithelium.

Summarizing, cysts are rare intraabdominal lower midline lesions, and they should be considered in the differential diagnosis of cystic lesions of that region. In most cases, diagnosis is established easily by clinical and radiographic examination. However, in case of very large cysts, such as our case, the origin of the lesion is more difficult to establish preoperatively. They should be excised totally, with a cuff of bladder in case of vesiculourachal abnormality, in order to rule out carcinoma and to avoid the possibility of neoplastic recurrence. In case of acute infection, they should be treated by incision and drainage, followed by total excision after remission of the infection. Laparoscopic excision has been performed successfully in a case [17].

Περίληψη

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Η κύστη του ουραχού είναι σπάνια ενδοκοιλιακή βλάβη με συχνότερη εντόπιση το κάτω τριτημόριο της κοιλίας. Στις περισσότερες περιπτώσεις η διάγνωση τίθεται προεγχειρητικά από τα κλινικά και ακτινολογικά ευρήματα. Εντούτοις σε περίπτωση ευ-

μεγέθους κύστης όπως στην περίπτωση μας η προέλευση της βλάβης δύσκολα καθορίζεται προεγχειρητικά. Η χειρουργική αντιμετώπιση περιλαμβάνει την πλήρη αφαίρεση της κύστης με τμήμα του θόλου της ουροδόχου κύστης, για να αποκλειστεί η πιθανότητα υποτροπής ή κακοήθους εξαλλαγής.

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