Allantoic remnants presenting as a giant retroperitoneal cyst

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Urachal anomalies are usually found in early childhood or just after birth. These usually involve patent ductus urachus, urachal cyst, umbilical-urachal sinus or vesicourachal diverticulum. Very rarely are urachal anomalies found in adults, usually as an infected urachal cyst.

We are presenting a case of surgically removed giant urachal retroperitoneal cyst that was found by chance during the abdominal ultrasound examination of a 22 year old man who was initially treated for idiopathic hypertension.

Key words: allantoic remnants, retroperitoneal cyst, urachus, urachal cyst

INTRODUCTION

Urachus is a normal embryonic remnant of the primitive bladder dome. It is derived from the allantois and the ventral cloaca, but exact contribution of each is contentious. It occupies the potential midline space between the peritoneum and the transversus fascia and has obliterated umbilical arteries on each side.

In fetus it extends with its growth to form an epithelialized tube at four months.

This tube has transitional (70%) or columnar epithelium (30%) surrounded by connective tissue and the outermost muscular layer is in continuity with the detrusor muscle.

Its lumen obliterates in utero in 98%. It rarely remains patent in part or as whole, resulting in urachal anomalies, further classified as congenital or acquired.

Congenital urachal anomalies present as patent urachus, umbilical-urachal sinus, vesico-urachal diverticulum and urachal cyst.

By this case report we wanted to show another rarity of the presentation of urachal remnants in an adult.

CASE REPORT

A young male, 22 years old, had one year history of hypertension. Additional ultrasonographic examination showed a large cystic formation in the right side of the abdomen, extending from the pelvis to the liver.

On the admittance to the hospital all blood, biochemical and urine analysis were within normal ranges (RBC 5.72, WBC 5.8, Hgb 169, Hct 48.2, PLT 237, Glucose 5.2, Urea 6.2, Creatinin 80, K 4.4, Na 141). Physical examination revealed a large, non tender mass in the right half of the abdomen, extending up to about 5 cm to the left of the umbilicus, giving dull sound on percussion.

Repeated ultrasound of the abdomen revealed dislocation of the right kidney towards the lateral abdominal wall and the diaphragm by a large cystic formation, as well as few other, smaller cysts in the region of the left kidney and the right side of the prostate, appearing to be communicating with the main one.

CT scan of the abdomen confirmed the position of the right kidney and showed the gigantic septated (Fig. 1) cystic formation extending from the right kidney to the periprostatic area, compressing pancreas to the left and givi-
ng the impression in the bladder wall by the cyst from the direction of the prostate. (Fig. 2). Radiologist was under the impression that there were few satellite cysts in the left kidney area.

Intravenous urography (IVU) showed slight distension of the right pyelocalyceal system, with the widening of the right ureter and the compression by a low density formation in the right half of the abdomen. There were no extralumination of the contrast in the bladder, which was slightly impressed from the direction of the prostate. (Fig. 3)

Intraoperatively a large irregularly shaped (tortuous) retroperitoneal cyst was revealed, extending from the diaphragm to the ileocecal region. There were neither satellite cysts nor other abnormalities in the abdomen. It was carefully separated from the surrounding structures and a lower, tubular extension of this formation was found, extending into the pelvis. At this point the cyst was pierced and about 4000 ml of the clear, serous liquid was evacuated. This tubular part of the cyst, which had several projections, extended under the prostate and anteriorly towards the inferior wall of the bladder, was ligated and carefully extirpated. (Fig. 4)

There were no postoperative complications and the patient was released from the hospital seven days later.

Pathohistological examination revealed transitional type of epithelium, what brought us to the conclusion that this formation was the cyst of the urachus. Thick muscular layer with the desquamated mucosa was showed on some slides, but in the wall of the wider part of the cyst even thicker muscular layer with the normal or slightly decreased stratified epithelium with recognizable Dogiel’s umbrella cells on the surface was revealed. (Fig. 5)

**DISCUSSION**

During gestation allantois appears on abut day 16 as a small fingerlike projection from the caudal wall of the yolk sac. The bladder develops from the ventral portion of the expanded terminal part of the hindgut, the cloaca, which is continuous with the allantois ventrally, so the
bladder initially extends all the way to the umbilicus. By the 4th or 5th month of gestation the bladder descends into the pelvis and its apical portion progressively narrows to a small epithelialised fibromuscular strand-urachus which varies from 3-10 cm in length and from 8-10 mm in diameter. Congenital urachal remnant diseases are due to urachal remnants that stay patent and are often subject to infections. An umbilical-uralchal sinus (representing about 15% of cases), vesicourachal diverticulum, (3-5%) or urachal cyst may all close normally after birth, but than reopen in association with pathologic conditions. This categorizes them as acquired diseases. Signs and symptoms in this case can often be confused with a wide spectrum of midline intraperitoneal or pelvic inflammatory disorders at examination, or with malignant tumors at imaging.

CT and ultrasound are diagnostic means that are suited for demonstrating urachal remnant diseases. Patent urachus presents at longitudinal ultrasound and occasionally at CT as a tubular communication of the umbilicus and bladder. A vesicourachal diverticulum is usually discovered incidentally at axial CT or at US of the bladder as extraluminally protruding fluid filled sack that does not communicate with the umbilicus. In this case, the vesicourachal diverticulum extended anteriorly, inferiorly into the pelvis and superiorly filling the retroperitoneal space all the way to the lower edge of the liver. The communication with the bladder had previously spontaneously closed, leaving the irregularly shaped cystic formation. Such a large cyst is an extremely rare anomaly. All the diagnostic procedures could only show the presence of the cyst, without its origin. Since there was no communication with the bladder, only pathohistology could offer the final diagnosis.

CONCLUSION
Urachal remnants are very rare entities, but can be revealed with commonly available diagnostic procedures. In this case the initial diagnosis could only be given intraoperatively and stated with certainty after the pathohistological examination. By this report we wanted to draw the attention of doctors to the possibility of having allantoic remnants presenting as a retroperitoneal cyst.

SUMMARY
Anomalije urahusa se obično otkrivaju u ranom detinjstvu ili odmah po rodjenju. U njih obično spadaju perzistentni ductus urachus, umbiliko-uralchal sinus ili veziko-uralchal divertikulum. Jako retko se anomalije urahusa nalaze kod odraslih ljudi i to obično kao inficirana urahalna cista. Ovom prilikom prikazujemo slučaj 22 godine starog muškarca, inicijalno lеченog od idiopatske hipertenzije, kod koga je slučajno otkrivena gigantska retroperitonealna cista urahusa ultrazvučnim ispitivanjem trbuha.

REFERENCES


