AFIP ARCHIVES

Best Cases from the AFIP

Urachal Carcinoma[1]

Ingrid M. Koster, MD • Pieter Cleyndert, MD • Raimond W. M. Giard, MD, PhD

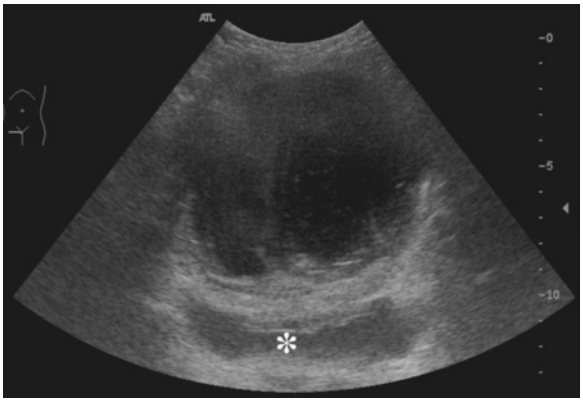

Figure 1. Transverse US image (obtained with a C5–2 MHz transducer with a convex array) of the lower abdomen shows a poorly filled bladder and focal thickening of the bladder wall (*). On top of the bladder, a large complex hypoechoic cystic lesion is seen.

History

A 58-year-old man presented with a 2-month history of painless gross hematuria. Physical examination revealed no abnormalities. The patient had a history of hepatitis A, sterilization, and a jaw operation. Urinalysis showed signs of hemorrhage but no malignant cells. Other laboratory test results were normal.

Imaging Findings

Ultrasonography (US) showed irregular focal thickening of the bladder wall and intravesical blood clots. At the dome of the bladder, a cystic mass with mixed echogenicity was seen (Fig 1). Contrast material–enhanced computed tomography (CT) revealed a supravesical midline mass of approximately 11.5 cm that extended toward the anterior abdominal wall. The mass had a mixed cystic and solid aspect. Peripheral punctate and curvilinear calcifications were seen within the solid part of the tumor, near the dome of the bladder (Figs 2, 3). Because there was no fat interface with the bladder, invasion of the bladder wall was likely. There were no signs of local or distant metastases.

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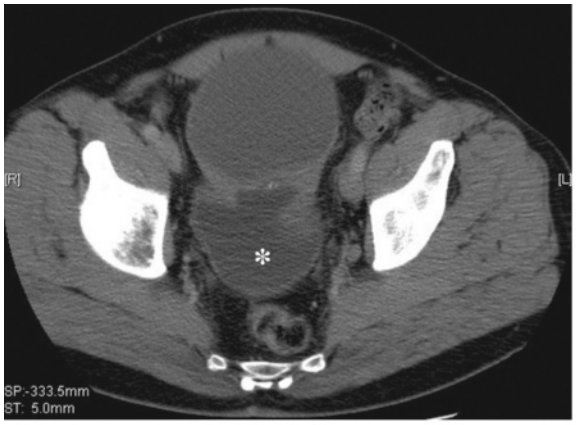
[1]From the Departments of Radiology (I.M.K., P.C.) and Pathology (R.W.M.G.), Maasstad Ziekenhuis, Olympiaweg 350, 3078 HT Rotterdam, the Netherlands. Received June 2, 2008; revision requested June 24 and received July 29; accepted July 29. All authors have no financial relationships to disclose. **Address correspondence to** I.M.K. (e-mail: *Ingrid.M.Koster@gmail.com*).



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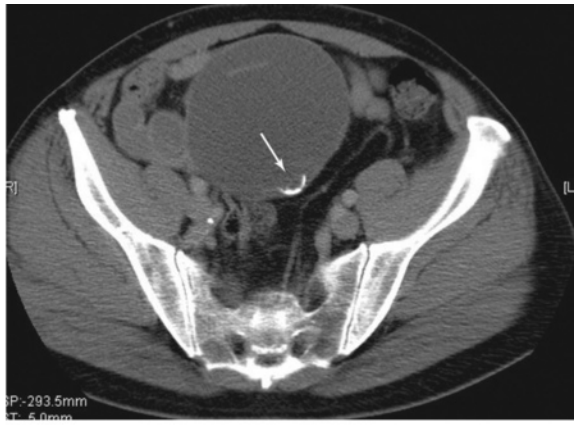

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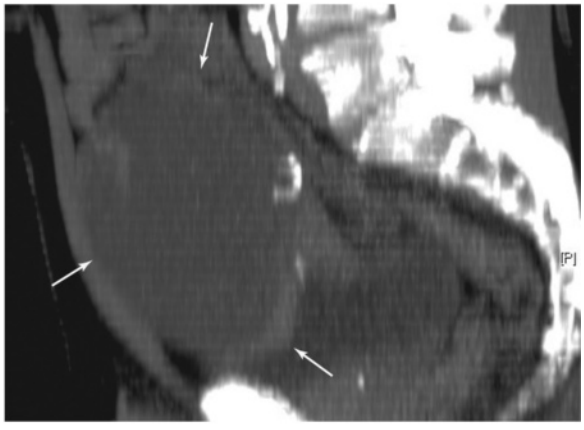


3.

Figures 2, 3. **(2a)** Contrast-enhanced CT scan shows a mixed cystic and solid midline mass with small punctate calcifications anterocranial to the bladder (*). The mass is slightly less attenuating than adjacent soft tissue, a finding suggestive of a mucus-filled structure. **(2b)** Contrast-enhanced CT scan shows peripheral curvilinear calcifications (arrow) in the wall of the cystic mass. Note the thick enhancing septum in the anterior portion of the mass. **(3)** Sagittal reformatted CT image shows the supravesical position of the extraperitoneal cystic mass (arrows). Focal thickening of the bladder wall is seen near the zone of contact with the bladder (lower right arrow).

Pathologic Evaluation

At cystoscopy, polypoid tissue was seen at the dome of the bladder. Biopsy specimens of the bladder wall showed malignant epithelial cells with a signet-ring cell type.

Radical cystectomy with en bloc resection of the mass was performed (Fig 4). Analysis of the gross specimen revealed a cystic 13 × 10-cm lesion filled with mucus and located at the dome of the bladder. Near the contact zone with the bladder, polypoid formations were seen and calcifications were palpable (Fig 5). Histologic analysis showed a poorly differentiated mucinous adenocarcinoma of the signet-ring cell type that extended into the serosa and submucosa of the bladder wall (Fig 6). These findings are consistent with malignant transformation of a urachal cyst.

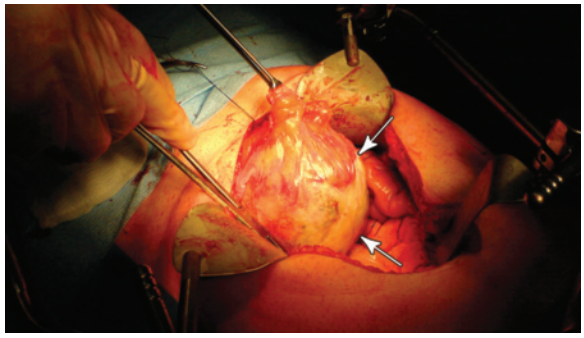
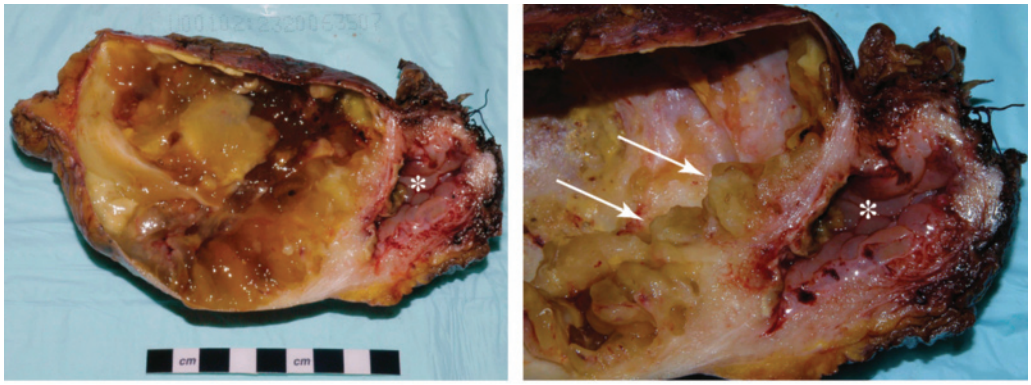


Figure 4. Intraoperative photograph shows a well-circumscribed oval mass in the lower anterior abdomen (arrows).

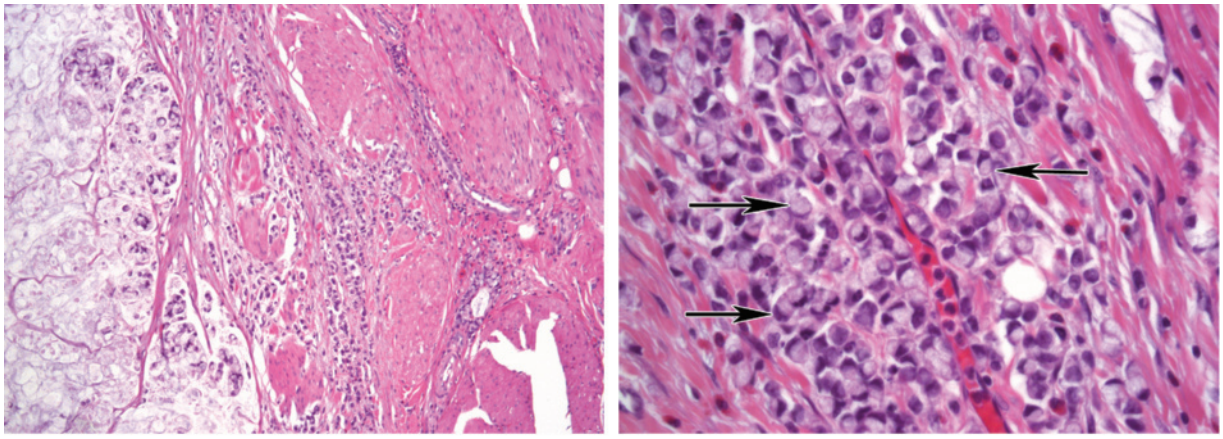
Discussion

The urachus is a midline remnant of at least two embryonic structures: the cloaca and allantois. It is located extraperitoneally and is bounded by the transverse fascia ventrally and the parietal peritoneum dorsally. This area is called the Retzius space (1). In late fetal life, the urachus usually deteriorates to a fibrous band, also known as the median umbilical ligament, which extends from the anterior dome of the bladder toward the umbilicus. Incomplete regression of the urachus results in four types of congenital urachal anoma-



a. b.

Figure 5. **(a)** Photograph of the gross specimen shows a large, cystic, partially septated lesion that is filled with mucus and located at the dome of the bladder (*), findings indicative of a urachal cyst. **(b)** Photograph of the gross specimen, obtained after removal of the mucus, shows polypoid formations within the cyst (arrows) near the zone of contact with the bladder (*). Calcifications were palpable at the dome of the bladder.



a. b.

Figure 6. **(a)** Photomicrograph (original magnification, ×40; hematoxylin-eosin stain) shows diffuse infiltration of poorly differentiated mucinous adenocarcinoma in the muscularis. **(b)** High-power photomicrograph (original magnification, ×250; hematoxylin-eosin stain) shows classic morphologic features of a signet-ring cell type adenocarcinoma (arrows).

lies: patent urachus, umbilical-urachal fistula, vesicourachal diverticulum, and urachal cyst. In our case, malignant transformation of a urachal cyst occurred. Urachal cysts account for 30% of all urachal anomalies and primarily occur in the lower one-third of the urachus. They do not communicate with the bladder or umbilicus (2).

Although the urachus is normally lined by transitional epithelium, most urachal tumors are adenocarcinomas (90%). These are thought to result from metaplasia of the urachal mucosa into columnar epithelium, followed by malignant transformation (1). Adenocarcinoma of the bladder is an uncommon neoplasm, accounting for only 0.5%–2% of all bladder carcinomas, and is classified as primary vesical, urachal, or metastatic. Thirty-four percent of bladder adenocarcinomas are urachal in origin (1–4). Urachal carcinomas tend to have a male predilection and are found in adults who are between 40 and 70 years old (1–3). The most common clinical feature is hematuria. Other signs and symptoms are dysuria, abdominal pain, a suprapubic mass, and discharge of blood, pus, or mucus from the umbilicus (5,6).

US, CT, and magnetic resonance (MR) imaging have the ability to display cross-sectional images and therefore are ideally suited for demonstrating urachal anomalies. As in our case, most urachal carcinomas arise in the juxtavesicular portion of the urachus. US may demonstrate a midline fluid-filled cavity with mixed echogenicity and calcifications adjacent to the anterior abdominal wall. A characteristic CT feature of urachal carcinoma is a midline mass anterosuperior to the dome of the bladder with low-attenuation components, which represent pools of mucin at pathologic examination (1). Peripheral calcifications in the soft-tissue–attenuation mass occur in 50%–70% of cases. They may be punctate, stippled, or curvilinear and are considered pathognomonic for urachal adenocarcinoma (1,2,7). MR imaging is an excellent staging tool. Because of the presence of mucin within the tumor, increased signal intensity is seen on T2-weighted spin-echo MR images (5). Both CT and MR imaging are useful for demonstrating intra- and extravesical extension of the tumor.

Benign urachal neoplasms, such as adenomas, fibromas, fibroadenomas, fibromyomas, and hamartomas, are rare but may mimic urachal malignancy (2,6). The radiologic differential diagnosis also includes adenocarcinomas of nonurachal origin, transitional cell carcinomas, infected urachal remnants, and metastases originating from primary lesions of the colon, prostate, or female genital tract (2,8). The typical extension of urachal carcinomas along the Retzius space helps differentiate them from vesical carcinomas (7).

Because of their extraperitoneal location, urachal carcinomas typically are silent and often show local invasion or metastases to the pelvic lymph nodes, lung, brain, liver, or bone at presentation (9). The prognosis is slightly better than that of nonurachal adenocarcinomas (4). Depending on the histologic subtype, stage, and resectability of the tumor, the 5-year survival rate for patients with urachal carcinoma ranges from 6.5%–61% (2,4,6). As in our case, signet-ring tumors are aggressive and tend to have the worst prognosis.

Surgery is the primary treatment of choice. In a retrospective study by Ashley et al (3), there was no difference in survival between patients who underwent partial cystectomy and those who underwent radical cystectomy. However, performance of complete urachectomy and umbilectomy was a significant predictor of survival. Urachal carcinoma can have metachronous or synchronous tumors along the urachal tract. Therefore, partial cystectomy and en bloc resection of the urachal mass, urachal tract, umbilicus, and pelvic lymph nodes are preferred. No definitive improvement in survival is seen after treatment with chemotherapy and radiation therapy (3,6,9). Local recurrence often is seen within 2 years of surgery (6).

Our patient underwent a cystectomy with en bloc resection of the urachal mass and Bricker deviation. Six months after surgery, peritoneal metastases were found. Despite undergoing a chemotherapy regimen of oxaliplatin and capecitabine, our patient died of metastatic disease within a year of the initial treatment.

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