U R O G E N I T A L

Cornelia M. Thali-Schwab
Paula J. Woodward
Brent J. Wagner

Computed tomographic appearance of urachal adenocarcinomas: review of 25 cases

C. M. Thali-Schwab · P. J. Woodward
Department of Radiologic Pathology,
Armed Forces Institute of Pathology,
Washington, DC, USA

B. J. Wagner
Reading Hospital,
Reading, PA, USA

C. M. Thali-Schwab (✉)
Insel Hospital,
University of Berne,
3010 Berne, Switzerland
e-mail: cornelia.thali@bluewin.ch
Tel.: +41-31-6322111
Fax: +41-31-6324874

Abstract Twenty-five cases of surgically proven urachal carcinomas were retrospectively reviewed. The radiological archives at the Armed Forces Institute of Pathology were searched for cases of surgically proven urachal carcinomas that had a computed tomographic (CT) scan as a part of their radiologic evaluation. CT images from all cases were evaluated to determine tumor morphology, presence and localization of calcification, extent of bladder invasion and metastases. Tumor size at presentation averaged 6 cm. Twenty-one of 25 (84%) were mixed cystic solid lesions and 4/25 (16%) were completely solid. Calcifications were present in 18/25 (72%), with 11 peripherally located, 3 central only and 4 both. Bladder wall invasion was present in 23/25 (92%), but was seen as an intraluminal mass in only 13/25 (52%). The bulk of the mass was extravesicular in 22/25 (88%). Metastases were present in 12/25 (48%). Our series supports observations from other smaller series that a midline, calcified, supravesicular mass is highly suspicious, if not pathognomonic, for urachal carcinoma.

Keywords Urachus · Urachal adenocarcinoma · Allantois · Computed tomography · Bladder development · Bladder neoplasm

Introduction

The urachus is a vestigial remnant of the intraembryonic portion of the allantois, which extends from the bladder dome to the umbilicus. Urachal carcinoma is a rare malignant neoplasm comprising only 0.01% of all adult cancers [1]. They are even rare among bladder tumors, accounting for only 0.34% of all bladder neoplasms; however, up to 39% of bladder adenocarcinomas are of urachal origin [2, 3]. Most adenocarcinomas of the urachus are mucinous. Less common histologic types include transitional cell and squamous cell carcinoma.

Several reviews have discussed the clinical aspects of urachal carcinoma [3–5]. Because of the rarity of this tumor, the radiological literature consists primarily of case reports and small series [6–10]. In this paper, we describe the computed tomographic (CT) findings in 25 pathologically proven cases, which, to our knowledge, is the largest series in the radiological literature.

Materials and methods

The radiological archives at the Armed Forces Institute of Pathology were searched for cases of surgically proven urachal carcino-

mas that had a CT scan as a part of their radiologic evaluation. These 25 cases were received between 1981 and 2001. The CT images were evaluated to determine] tumor morphology (cyst vs. solid) [1], presence [2] and localization [3] of calcification, extent of bladder invasion [4] and metastases [5]. The pathology and surgical reports were reviewed for all patients. The films were compared to the gross specimens and, where possible, clinical information regarding symptoms was obtained.

Results

There were 13 males and 12 females with an average age of 48 (range 21–69 years). Tumor size averaged 6 cm in the largest cross-sectional diameter (range 2–12 cm). Twenty-one cases (84%) appeared as mixed cystic-solid lesions (16 predominantely cystic and 5 predominantely solid); 4 (16%) were completely solid (Figs. 1, 2, 3). All were adenocarcinomas. The CT appearance correlated well with gross pathologic findings. Calcifications were

radiologically visible in 18/25 (72%) and varied from fine punctate deposits to extensive areas of calcification. Eleven cases had purely peripheral calcifications, four had both peripheral and central calcification, and three showed purely central calcification (Figs. 4, 5, 6).

Invasion of the bladder wall was very common and occurred in 23/25 (92%). Invasion, however, was not always obvious by CT. In only 13/25 (52%) was there obvious invasion into the bladder lumen correlating with mucosal invasion by histology (Fig. 7). In 10/25 (40%), there was histologic invasion into the muscularis layer of the bladder, but the mucosa was preserved. The bladder wall did not appear abnormal in any of these cases. In 22 cases (88%), the extraluminal portion of the mass was larger than the intraluminal portion. In the remaining two cases, the mass was totally extrinsic to the bladder without invasion.

Sixteen of 25 (64%) had pathologically proven local perivesical fat invasion; in addition, two had lymphatic and two small bowel invasion (Fig. 8). Local invasion was
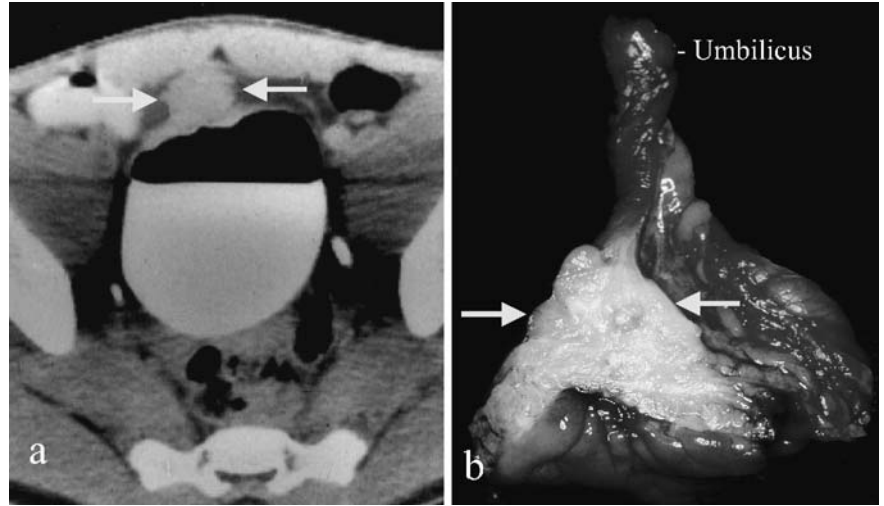

Fig. 1 a Axial and **b** sagittal reformatted CT images show a mixed cystic-solid lesion (*arrows*); *B* bladder


Fig. 2 a Axial CT image demonstrates a predominantly cystic lesion with a small solid component (*arrow*). **b** Photograph of the resected tumor (*arrow*) and bladder (*B*)
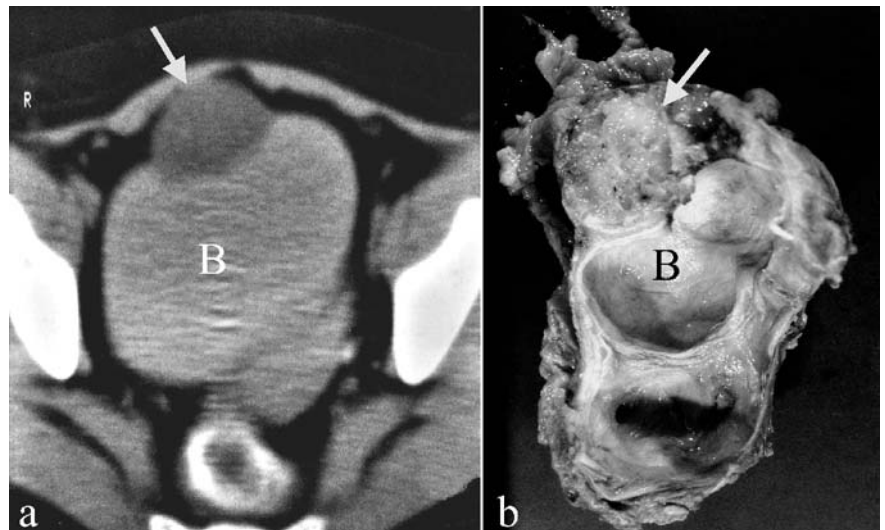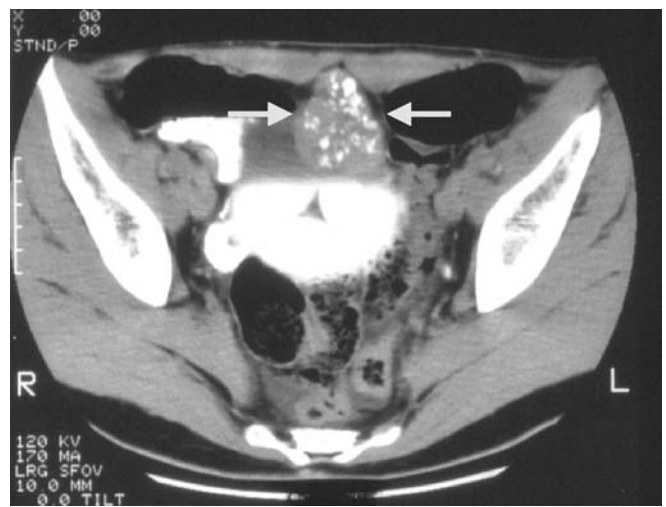
Fig. 3 a Axial CT image of a totally solid lesion (*arrows*) with the **b** corresponding pathological specimen

Fig. 4 a Abdominal radiograph and **b** corresponding axial CT image shows a mass with peripheral calcifications (*arrows*)
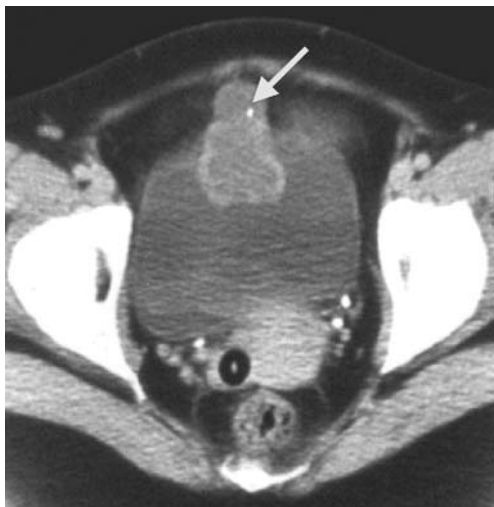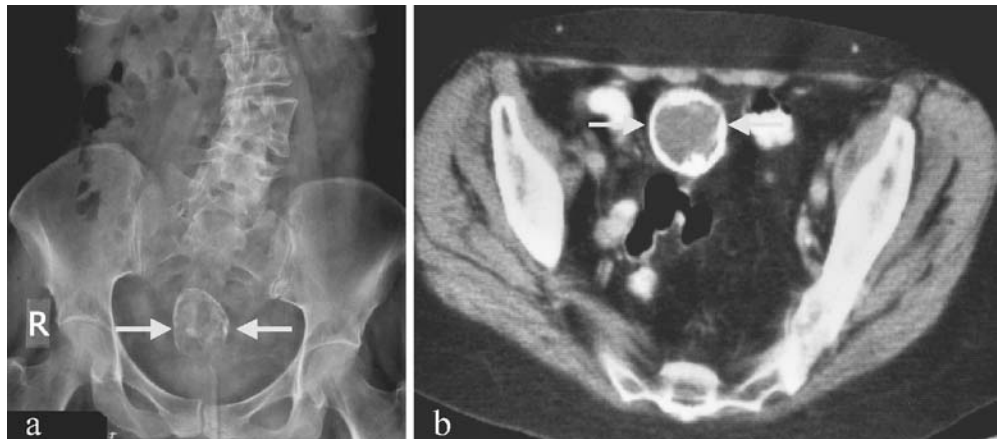

Fig. 5 Axial CT image showing a single punctate central calcification within the mass (*arrow*)


Fig. 6 Axial CT image with extensive peripheral and central tumor calcifications (*arrows*)

Fig. 7 Bladder invasion by urachal carcinoma. **a** Sagittal ultrasound image shows a solid mass fungating into the bladder lumen (*arrow*). **b** Axial CT image shows this mass has both cystic and solid components (*arrows*). **c** Photograph of the resected specimen shows a predominately extraluminal mass (*curved arrow*) with involvement of the bladder mucosal (*straight arrows*). *B* bladder
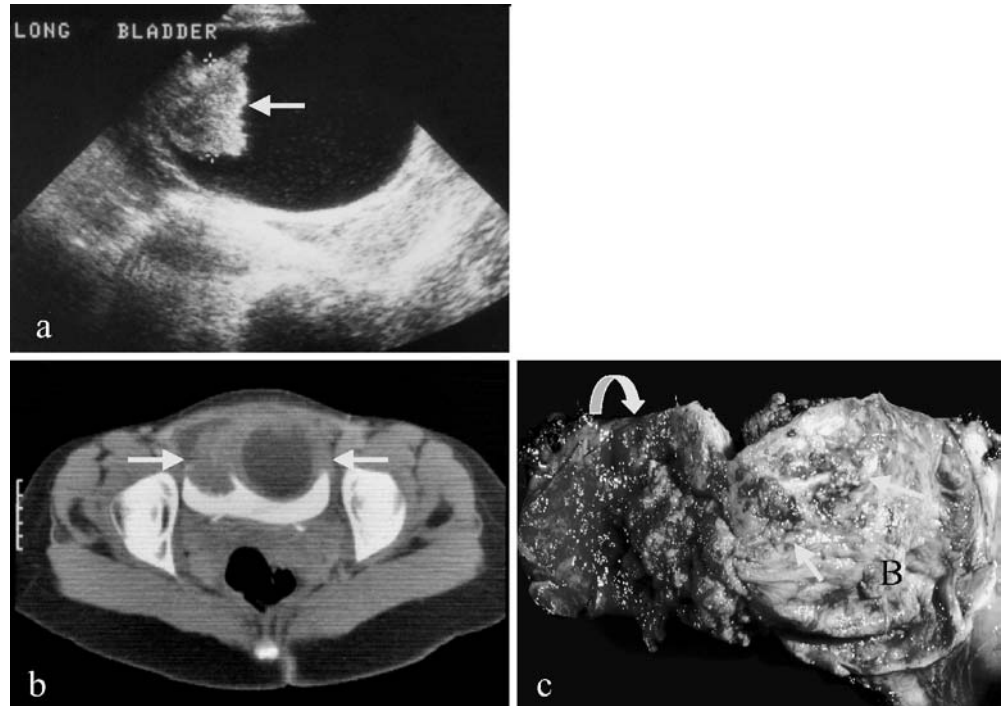
Fig. 8 Urachal carcinoma with invasion of the small bowel. **a** Abdominal radiograph after instillation of contrast into the bladder shows a vesicoenteric fistula (*arrow*) with filling of multiple small bowel loops. **b** Axial CT image shows both enteric contrast and gas within the tumor
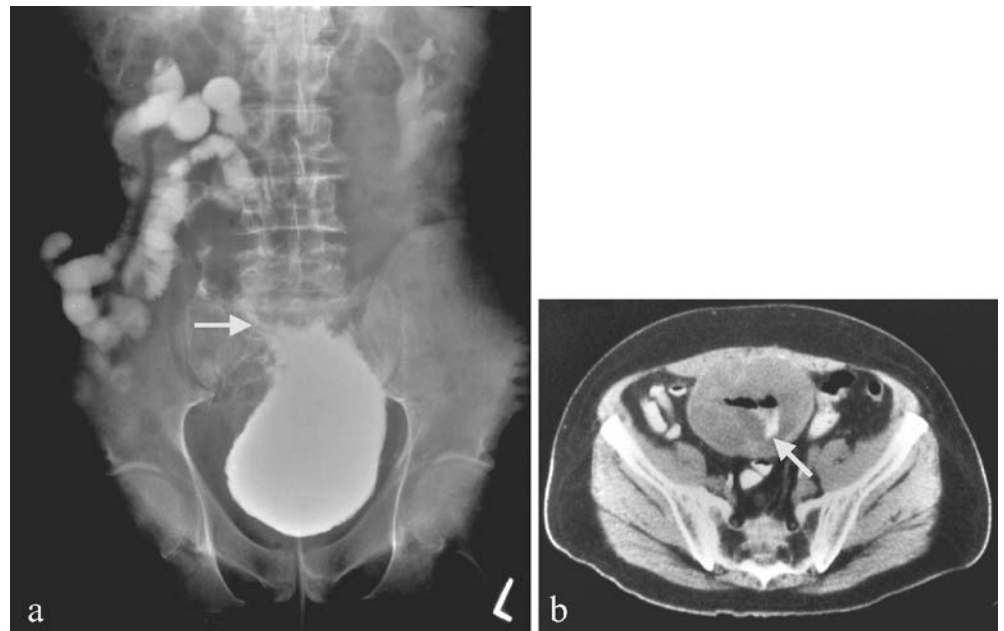
suggested by CT when the borders of the mass were irregular. Two cases of local invasion were not radiologically evident. At the time of initial diagnosis of the primary lesion, distant metastases were present in 8/25 (32%) and were noted in the following locations: bone (2), cerebellum (1), peritoneal cavity (1), bilateral ovary (1), umbilical skin (1), perirectal soft tissues (1) and omentum (1).

Clinical information was available in 22/25 patients. In our series 13/22 patients (59%) presented with hematuria as the first clinical sign, 5/22 (23%) had pain (diffuse pain and dysuria 1, suprapubic pain on voiding 1, painful pelvic mass 1, dyspareunia 1, low back pain 1). Only 2/22 (9%) presented with mucinuria. One patient (5%) had no symptoms.

Discussion

The intraembryonic portion of the allantois forms a connection from the apex of the bladder to the umbilicus. As the bladder enlarges, the allantois involutes and forms a fibrous cord called the urachus or median umbilical ligament [10]. It is an extraperitoneal midline structure running within the space of Retzius, between the transversalis fascia and the parietal peritoneum [1]. It can be quite variable in length (2–15 cm) and appearance (a well-defined ligament vs. a fine, fibrous plexus) [2]. Up to 30% of adults have been shown to have a microscopic lumen at autopsy; however, functionally, the urachus is closed by the third trimester of fetal life [10]. Failure of complete urachal closure gives rise to a variety of anomalies, including a patent urachus, urachal cyst, urachal sinus or a vesicourachal diverticulum [10]. In addition to closure anomalies, benign and malignant neoplasms can arise in the urachus.

Although the normal urachus is most commonly lined by transitional epithelium, the majority of urachal carcinomas are adenocarcinomas [6]. Two theories have been suggested as possible explanations. The first, and more commonly accepted theory, proposes there is columnar metaplasia of the transitional epithelium, which subsequently undergoes malignant transformation [1, 3]. An alternative, although less likely, explanation is that adenocarcinomas could arise from embryonic rests within islands of mucus-containing hindgut epithelium persisting within the urachus [7, 8].

In our series, most tumors were complex, mixed, cystic and solid masses. The cystic portion was often filled with mucinous material. A high mucin content contributes to the low-density appearance often seen on CT [2]. The minority (16%) were purely solid. In none of the cases was a purely cystic mass seen (a potential confusion for a urachal closure abnormality). As with mucinous tumors elsewhere, urachal carcinoma has a propensity for psammomatous calcification. Calcification is generally extensive enough that it is easily seen by CT. In our series these calcifications occurred more often in the periphery, but were also central or a combination of both.

Histologically, urachal carcinoma is often indistinguishable from a primary bladder adenocarcinoma. In all but three of our cases, the bulk of the tumor was extraluminal. This finding can be very helpful in differentiating it from a bladder adenocarcinoma, which would be expected to be primarily intraluminal [9]. Bladder wall invasion is common in urachal carcinoma and was present in 92% of cases in our series. This is not always an obvious CT finding, however. In 40% of the cases, invasion was confined to the muscularis and was not apparent by CT. The area of the dome of the bladder is often difficult to evaluate secondary to volume averaging effects. With newer multidetector CT scanners, thinner sections and better multiplanar reformatting are possible and may improve the evaluation for invasion. It is important to note that with conventional axial imaging, bladder wall invasion may be missed and a normal appearing bladder does not exclude invasion.

The most common clinical symptoms described for urachal adenocarcinomas include hematuria, irritative bladder symptoms (frequency, urgency and dysuria) and pain. This was also true in our series. In a review series by Sheldon et al. [3], they reported mucinuria (either microscopic or gross) in 25% of the patients. It was lower in our series, occurring only in 2 of 22 patients (9%) for whom history was available. This discrepancy may be due to incomplete data. Three of our cases had no history listed, and the other histories were written by the case contributor, which may or may not have been complete. Our data varied further in that this series did not show a significant male predilection, and the average age was younger than reported by others [2, 3].

Because the tumor growth occurs in an area that is relatively clinically silent, patients with urachal carcinoma generally present late in their course, making the prognosis poor. They do far worse than patients with a primary bladder adenocarcinoma, which is histologically similar, but typically presents much earlier. In our series, 28% had metastases at the time of presentation. Metastases have been reported most commonly to involve the regional lymph nodes, omentum, liver, lung and bone [2–5]. We found additional sites, including the cerebellum, peritoneal cavity, bilateral ovaries, umbilical skin and perirectal soft tissues.

In conclusion, our series supports observations from other smaller series that a midline, calcified, supravesicular mass is highly suspicious, if not pathognomonic, for urachal carcinoma. Bladder wall involvement is common and cannot be excluded, even in the absence of radiological findings. In view of the high frequency of bladder invasion at the time of presentation, the pathologist is often unable to establish if the lesion is a primary tumor of the urachus or bladder by histologic examination alone. Therefore, based on the location of an extravesical component of the lesion, the radiologist may often be of value in suggesting that the neoplasm is urachal in origin.

References

1. Hori S (1983) Staging of urinary bladder cancer—evaluation by CT. Nippon Igaku Hoshasen Gakkai Zasshi 43:1024–1035

2. Brick SH, Friedman AC, Pollack HM, Fishman EK, Radecki PD, Siegelbaum MH, Mitchell DG, Lev-Toaff AS, Caroline DF (1988) Urachal carcinoma: CT findings. Radiology 169:377–381

3. Sheldon CA, Clayman RV, Gonzalez R, Williams RD, Fraley EE (1984) Malignant urachal lesions. J Urol 131:1–8

4. Kakizoe T, Matsumoto K, Andoh M, Nishio Y, Kishi K (1983) Adenocarcinoma of urachus. Report of 7 cases and review of literature. Urology 21:360–366

5. Mattelaer P, Wolff JM, Jung P, IJzerman W, Jakse G (1997) Adenocarcinoma of the urachus: 3 case reports and a review of the literature. Acta Urol Belg 65:63–67

6. Mangiacapra FJ, Scheraga JL, Jones LA (2001) Mucinous colloid adenocarcinoma of the urachus. Radiographics 21:965–969

7. Mekras GD, Block NL, Carrion HM, Ishikoff M (1980) Urachal carcinoma: diagnosis by computerized axial tomography. J Urol 123:275–277

8. Mostofi FK, Thomson RV, Dean AL (1954) Mucous adenocarcinomas of the urinary bladder. Cancer 8:741

9. Narumi Y, Sato T, Kuriyama K, Fujita M, Saiki S, Kuroda M, Miki T, Kotake T (1988) Vesical dome tumors: significance of extravesical extension on CT. Radiology 169:383–385

10. Yu JS, Kim KW, Lee HJ, Lee YJ, Yoon CS, MJ Kim (2001) Urachal remnant diseases: spectrum of CT and US findings. Radiographics 21:451–461