Image Quiz for surgeons

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A Rare Case of Metastatic Squamous Urachal Carcinoma

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Rezumat

Caz rar de carcinom scuamos de uracă metastatic

Carcinomul scuamos este un tip foarte rar de tumoră malignă de uracă, doar câteva cazuri fiind raportate în literatura medicală. Prezentăm cazul unui pacient în vârstă de 49 de ani diagnosticat cu carcinom scuamos de uracă suprainfectat cu multiple metastaze pulmonare, după ce acuză dureri de etaj abdominal inferior, tumoră abdominală și febră, fără simptome respiratorii. Ecografia abdominală și tomografia computerizată au relevat o masă tumorală la nivelul abdomenului inferior, în contact cu peretele abdominal și domul vezicii urinare, deplasând intestinului subțire. Leziuni pulmonare nodulare au fost descrise în piramida lobului stâng. Diagnosticul intraoperator a fost de tumoră necrozată de uracă cu invazie în domul vezicii urinare și suspiciune de metastaze pulmonare pentru care s-a practicat ablația tumorii în bloc cu rezecția domului vezical și cistorafie. Rezultatul histopatologic a fost carcinom scuamos cheratinizat slab diferențiat (G3), cu margini de rezecție negative. Pacientul s-a recuperat după intervenția chirurgicală, dar prognosticul este nefavorabil datorită stadiului metastatic în care tumora a fost diagnosticată, deoarece până în prezent nu există nici un regim chimioterapic eficient pentru carcinomul metastatic de uracă.

Cuvinte cheie: carcinom scuamos de uracă, metastaze pulmonare, ablația tumorii

Abstract

Squamous cell carcinoma is a very rare type of urachal malignancy, only a few cases being reported in the medical literature. We present the case of a 49-year-old male patient diagnosed with infected squamous cell urachal carcinoma with multiple pulmonary metastases, after complaints of lower abdominal pain, abdominal mass and fever, without respiratory symptoms. The abdominal ultrasonography and the CT scan revealed a tumoral mass in the lower abdomen in contact with the abdominal wall and the urinary bladder dome, displacing the small bowel. Pulmonary nodular lesions were described in the left lobe pyramid. The intraoperative diagnosis was necrotic urachal tumor with urinary bladder dome invasion and suspected pulmonary metastases, and tumor ablation with bladder dome resection and suture of the bladder were performed. The histopathological result was poorly differentiated squamous cell carcinoma (G3), with negative resection margins. The patient recovered well after surgery, but the prognosis is very poor due to the metastatic stage in which the tumor was diagnosed, no standard chemotherapy regimen for the treatment of metastatic urachal carcinoma being known as effective until now.

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Key words: squamous urachal carcinoma, pulmonary metastases, tumor ablation

Introduction

Squamous cell carcinoma of the urachus is a very rare finding; a review of the literature found only eight cases reported from 1870 to 2006 from the total experience of little less than 300 cases of urachal malignancies, and all eight cases were diagnosed as localised malignancy. The first case of squamous urachal carcinoma with metastatic disease at the time of diagnosis was reported by Kodali et al. in 2006, and since then we did not find another case of squamous type carcinoma of the urachus to be published in the last 7 years. This is a case of squamous cell carcinoma of the urachus with pulmonary metastases which became symptomatic and was revealed because of the infection of the primary tumor.

Case presentation

We present a case of a 49-year-old male patient admitted with complaints of progressive lower abdominal pain, abdominal mass and fever. Physical examination revealed a painful palpable hard mass situated on the midline in the lower abdomen, of approximately 20 cm in diameter, adherent to the abdominal wall, without other abnormalities. Biochemical findings: systemic inflammation with elevated neutrophil count and fibrinogen level. Imagistic investigations (abdominal ultrasonography and abdominal CT scan) revealed an intra-peritoneal mass of 11/8/8.5 cm in the lower abdomen, with thick and partially calcified walls and a few septa separating the fluid content (maximum density 23 UH at CT scan). This mass was in contact with the abdominal wall without a demarcation limit, and its inferior part was tight to the urinary bladder dome, compressing the small bowel which was displaced. The right iliac vascular bundle was tangent to the described tumor, but without evidence of tumoral invasion (Fig. 1). No metastases were found in the abdomen, but in the inferior thoracic images intercepted nodular lesions were described in the left lobe pyramid, the largest one being of 47/27 mm, with central excavation and adjacent small alveolar infiltrations presenting air bronchogram (Fig. 2). The radiologist concluded that the abdominal images were suggestive for an infected hydatid cyst, and the pulmonary lesions could represent secondary pulmonary abscesses.

The chest X-ray revealed a well-shaped homogenous nodular opacity of 20/26 mm in the left lobe pyramid, of moderate intensity and a few emphysematous bullae in the posterior segment of the superior right lobe.

The treatment choice for this patient was surgical resection of the infected abdominal mass. The intraoperative finding was: necrotic urachal tumor with urinary bladder dome invasion and suspected pulmonary metastases for which tumor ablation with bladder dome resection and suture of the bladder were performed. (Fig. 3, 4)

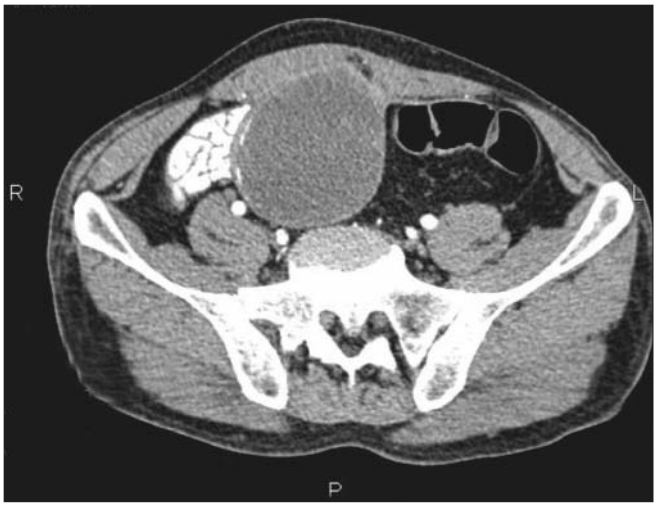

Figure 1. *Abdominal CT scan - calcified tumor in the lower abdomen*

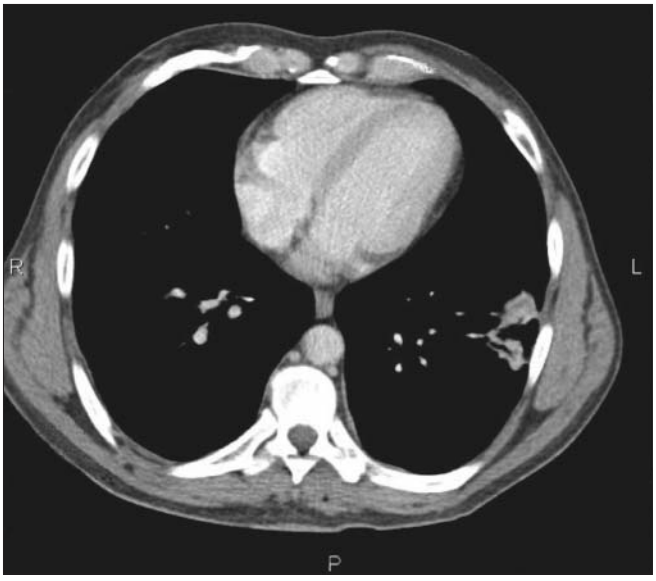

Figure 2. *Pulmonary CT scan – nodular lesion of the left lobe pyramid*

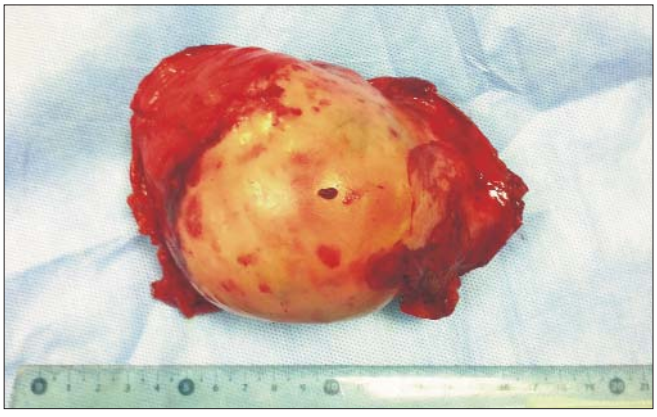

Figure 3. *Resection piece - necrotic urachal tumor*

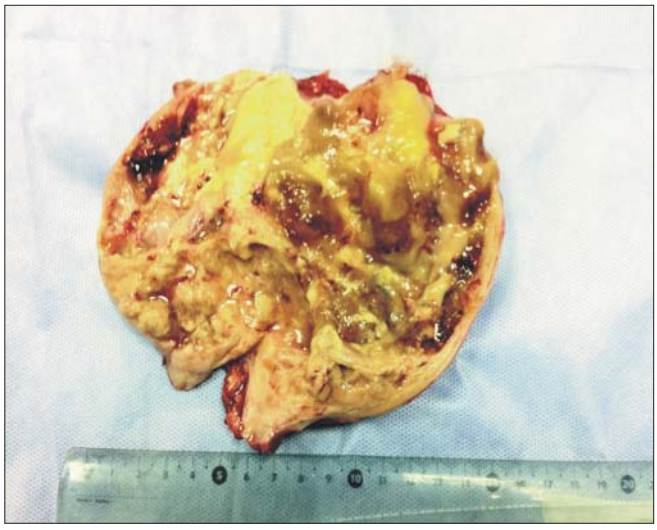

Figure 4. *Resection piece - necrotic urachal tumor*

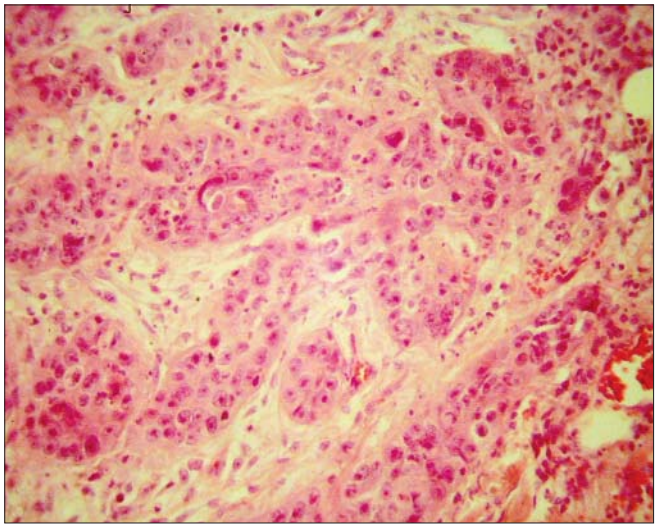

Figure 5. *Squamous carcinoma, poor differentiated, HE stain, X200*

Histopathology study of the resected piece showed an encapsulated tumor of 14/7/6 cm, with nodularities on its surface, with focal calcifications, which microscopically represents a poorly differentiated squamous cell carcinoma (G3), with negative resection margins (Fig. 5, 6).

The patient recovered well after the surgical intervention and was referred to the local oncologist for specific treatment and medical survey.

Discussions

The urachus is a vestigial remnant of two embryological structures: the cloaca and the allantois. It is an extraperitoneal midline tubular structure, which extends from the anterior dome of the bladder to the umbilicus (Schubert GE cited by 1). After birth, this structure remains as a fibrous band, known as the median umbilical ligament. The persistence of the urachal remnant can lead to various benignant or malignant diseases, both in childhood and in adult age. Congenital urachal anomalies are twice more frequent in men, and are usually asymptomatic due to their extraperitoneal location, unless they complicate with infection or malignancy. Malignant urachal neoplasms account for 0.01% of all malignancies (2) and represent less than 0.5% of bladder cancers (3), the majority of cases being adenocarcinomas (90%) (4). The first urachal cancer was described in 1863 by Hue and Jacquin in a report translated later by Sheldon (3). Mostofi and associates developed the totipotential theory in 1955, according to which squamous cell carcinoma develops secondary to the squamous metaplasia in the urachal epithelium (5).

Patients have a poor prognosis, because they usually have no early symptoms and therefor are diagnosed late (6-8), frequently in the metastatic stage, when no effective treatment is available (9,10). The most common symptoms of squamous cell carcinoma are abdominal pain (75%), pollakiuria with dysuria (75%) and haematuria (50%) (3). Our patient had no urinary symptoms, but abdominal pain and abdominal mass were present.

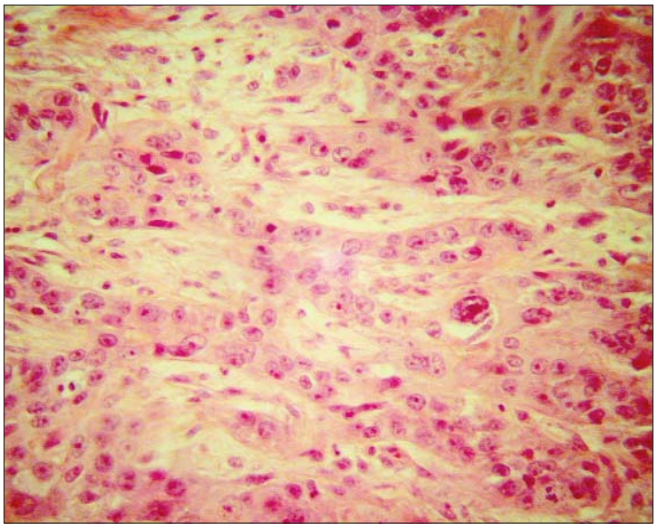

Figure 6. *Squamous carcinoma, poor differentiated, HE stain, X400 (detail)*

Abdominal US and CT scan can identify most urachal remnant diseases (1) and solid masses or those with calcification are more likely to be malignant (11), some authors considering that calcifications in a midline supravesical mass (also present in our case) are nearly diagnostic for urachal carcinoma (12,13). In our case, the abdominal CT scan images were considered by the radiologist to be suggestive for an infected hydatid cyst due to the liquid content and the calcifications, which are often present in echinococcus granulosus infections (14-16).

The diagnosis can be confirmed in some cases by cystoscopy with endoscopic biopsy (17). This patient having no dysuria or haematuria, but an infected abdominal mass was referred for surgery without a cystoscopy.

The MD Anderson Cancer Center developed a set of criteria for the diagnosis of urachal cancer (18): a. location in the bladder dome or elsewhere in the midline of the bladder, b. sharp demarcation between tumor and normal surface

epithelium and c. supportive criteria: enteric-type history, absence of urothelial dysplasia, absence of cystitis cystica or cystitis glandularis transitioning to the tumor and absence of primary adenocarcinoma of another organ.

It is known that the traditional staging system of urothelial tumors does not apply in urachal tumors, which grow outside the lumen of the bladder (2,19). In 1984 Sheldon et al. proposed a first staging system for urachal cancer (3):

- Stage I Urachal cancer confined to urachal mucosa;
- Stage II Urachal cancer with invasion confined to urachus itself;
- Stage IIIA Local urachal cancer extension to bladder;
- Stage IIIB Local urachal cancer extension to abdominal wall;
- Stage IIIC Local urachal cancer extension to peritoneum;
- Srage IIID Local urachal cancer extension to viscera other than bladder;
- Stage IVA Metastatic urachal cancer to lymph nodes;
- Stage IVB Metastatic urachal cancer to distant sites.

In 2006 a simplified Mayo staging system was proposed (17) as a modification of the Henly staging system from 1993 (20):

- Stage I Urachal cancer confined to the urachus and/or bladder;
- Stage II Urachal cancer extending beyound the muscular layer of the urachus and/or bladder;
- Stage III Urachal cancer infiltrating the regional lymph nodes;
- Stage IV Urachal cancer infiltrating the non-regional lymph nodes or other distant sites.

The authors from the Mayo Clinic found a high correlation of the two staging systems (Mayo and Sheldon) in terms of predicting cancer-specific survival and recommend the use of their simplified staging system, considering that the Sheldon system is over-specified and has an unnecessary complexity, but no staging system has been validated yet, the cause being the limited experience with urachal cancer. Our patient is a stage IV urachal carcinoma, the pulmonary metastases leading to a very poor survival expectation.

Ashley et all. reported a 50 year experience and studied a big cohort of urachal tumors (a total of 130 patients, 66 of them having malignant tumors), finding two important predictors of malignancy in the multivariate regression model: the presence of haematuria (17-fold risk) and the age over 55 years (3-fold risk) (17). Also mucosuria is an very rare urological symptom and should raise the possibility of urachal pathology when it is present (but unfortunately it is found in under 10% of the urachal diseases cases) (3). None of these factors were present in the urachal carcinoma case we exposed above.

The most common sites for metastasis of urachal cancer are bone, lung and liver, but peritoneal carcinomatosis and other organs are possible secondary tumor sites too (2,21).

The multivariate analysis of predictors of cancer-specific survival found that tumor grade and surgical margins were the most important prognostic factors (17), different from other studies that found omission of umbilectomy and increased cancer stage as important indicators of poor outcome (21,22). Tumor stage was found as an important prognostic factor also in a recent review of the Memorial Sloan-Kettering Cancer Center experience, published by Gopalan and all (23).

Umbilectomy still remains an important aspect of surgical treatment in urachal cancer, the treatment of choice in this kind of tumors being demonstrated to be total urachectomy, which entails excision of the urachal remnant along with its peritoneal, fascial and umbilical segments and excision of the bladder dome, with negative margins. (3,9,20-22) Lymphadenectomy is not recommended because it does not improve the outcome of these patients. Also, the multimodal approach in the treatment of urachal cancer is not convincing yet, the benefit of radiation or chemotherapy being unclear at this point (3,21). There is no standard chemotherapy regimen for the treatment of metastatic urachal carcinoma (24,25). Recent case reports show the benefit of combined chemotherapy in isolated cases of urachal cancers, most of them adenocarcinomas: the association of 5-FU, cisplatin or oxaliplatin, irinotecan and bevacizumab in different combinations demonstrated usually a partial and limited response (26-30).

Patients should be monitored carefully after surgery to identify early local tumor recurrences, which usually occur within 2 years from surgery (3,18), because the prompt surgical resection of these recurrences can be curative (17).

Overall survival for all stages of urachal carcinomas is 62 months, with 34% survival at 5 years (31). For stage IV urachal malignancies (as is our case) the survival does not usually exceed 12 months.

Conclusion

Squamous cell carcinoma of the urachus with additional infection of this cystic tumor and pulmonary metastatic disease is an extremely rare association, this case being the first reported in the literature. This patient had no urinary symptoms and was diagnosed because of the associated infection and the rapid growth of the tumor. Despite the complete resection of the primary tumor, the prognostic is poor due to the multiple pulmonary metastases.

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