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Primary Signet Ring Cell Adenocarcinoma of the Urinary Bladder: A Report of 2 Cases☆

Wiem Boukettaya [a,*], Jihene Feki [a], Slim Charfi [b], Nabil Toumi [a], Afef Khanfir [a], Tahia Boudawara [b], Jamel Daoud [c], Mounir Frikha [a]

[a] Department of Medical Oncology, CHU Habib Bourguiba, Sfax, Tunisia
[b] Anatomopathology Laboratory, CHU Habib Bourguiba, Sfax, Tunisia
[c] Department of Radiotherapy, CHU Habib Bourguiba, Sfax, Tunisia

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ABSTRACT

Primary signet ring cell carcinoma of the urinary bladder is a rare and aggressive histologic subtype of adenocarcinoma. In general, this tumor occurs in the middle age, and clinical presentation does not differ from transitional cell carcinomas. The prognosis is often poor, given the advanced stage at diagnosis. To our knowledge, <100 cases of signet ring cell adenocarcinoma of the urinary bladder have been reported. We report 2 cases with bladder linitis plastica primitive, and we draw attention to its pathologic, anatomoclinical, and evolution specificity to optimize its therapeutic management.

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Introduction

Primary signet ring cell adenocarcinoma of the urinary bladder, also called linitis plastica urinary bladder, is rare, accounting for only 0.24% of all malignant tumors of the urinary bladder.[1]

Case presentation

Case 1

A 72-year-old patient consulted for intermittent painless total gross hematuria, urgency, and pollakiuria. The medical and familial histories were unremarkable. Physical examination was normal. The abdominal and pelvic ultrasound showed a bilateral hydroureteronephrosis with thickening of the urinary bladder wall. Cystoscopy visualized a solid mass in the left-side wall of the urinary bladder. Histologic examination of cystoscopic biopsy showed a proliferation of round-cell aspect of signet ring. An immunohistochemical study demonstrated positivity for cytokeratin 7 and negativity for cytokeratin 20. The diagnosis of signet ring cell adenocarcinoma of the bladder was established. Abdominal computed tomography (CT) showed no locoregional lymph nodes, metastases, or a primary tumor in other abdominal or pelvic organs.

We performed a complete gastrointestinal endoscopic evaluation to exclude an extravesical primary tumor site, but no other primary site was found. The tumor was therefore treated as a primary signet ring cell carcinoma (SRCC) of the urinary bladder. The patient underwent a radical cystoprostatectomy. The intraoperative examination found a budding tumor inserted to the left-side wall. Histologic examination concluded to a signet ring cell adenocarcinoma with a colloid component estimated about 40%. The tumor was invasive; it extended into the perivesical fat with carcinomatous lymphangitis and nerve sheathing perished (Fig. 1).

Surgical resection margins were free of tumor cells. The tumor was classified pT3N0M0. The patient had no adjuvant treatment. The patient consulted again after 16 months for hematuria and perineal pain. Endoscopy showed stenosis of the anterior urethra and the biopsy confirmed tumor relapse in the urethra. Radiotherapy at a dose of 64 Gy was delivered: the first dose of 44 Gy at 5 fractions of 2 Gy/wk in the pelvis and then an additional 20 Gy in a limited volume in the urinary bladder. The patient was followed up every 6 months, and a thoracoabdominal CT scan was done every 6 months. The patient has radiological stability and kept a preserved quality of life after 3 years of follow-up.

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* Corresponding author. Tel.: +2-169-764-1475.
E-mail address: wiembouk@yahoo.fr (W. Boukettaya).

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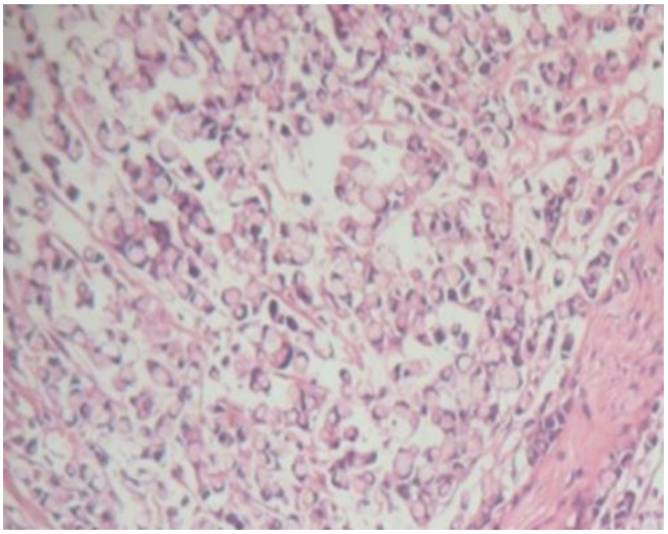

Figure 1. Histopathology of the bladder lesion showing multiple signet ring cells (hematoxylin-eosin ×200).

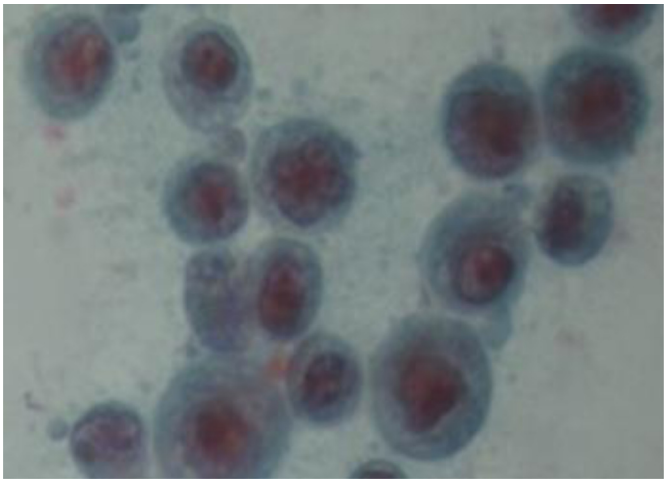

Figure 2. Neoplastic cells in the ascitic fluid.

Case 2

A 64-year-old patient without medical history consulted with a history of 2 months of total hematuria. Pelvic ultrasound showed an infiltrating mass in the posterolateral wall of the urinary bladder associated with a left hydroureteronephrosis. Cystoscopy showed a pseudopolypoid mass on the left posterolateral urinary bladder. Endoscopic resection of the tumor was performed. Pathologic examination found a poorly differentiated invasive signet ring cell adenocarcinoma. An abdominal CT scan showed a large effusion occupying the entire abdomen and peritoneal cavity without evidence of peritoneal carcinomatosis.

The digestive exploration (gastroduodenoscopy and colonoscopy) showed no suspicious location. The evolution was marked by the appearance of ascites. Cytologic analysis of the peritoneal fluid revealed the presence of neoplastic cells (Fig. 2).

Palliative chemotherapy has been proposed but not performed because of the deterioration in the general condition of the patient. He was followed in the palliative care consultation. The patient died 5 months after diagnosis.

Discussion

Primitive bladder adenocarcinoma accounts for only 0.5%-2% of all primary malignant tumors of the bladder.[1] Most adenocarcinomas of the urinary bladder result from direct extension from adjacent organs (eg, colon, prostate). Rarely, there can be metastatic spread to the bladder of SRCC originating in another organ.[2] The variant signet ring cell is a poorly differentiated form, is exceptionally described, and its incidence is about 0.24% of bladder cancers.[2]

Hematuria, which was the reason for consultation in all our patients, is the most common clinical presentation. Other symptoms that have been reported are dysuria, pollakiuria, and urinary incontinence or retention.[3]

It is essential to distinguish this carcinoma from metastases as different therapeutic strategies are often necessary. Primary SRCC of the urinary bladder has the same histology as that of the gastrointestinal tract, breast, lung, and prostate; therefore, further evaluations for other primary sites are mandatory to exclude metastasis.[4] In our case, the gastrointestinal evaluation included esophagogastroduodenoscopy and colonoscopy, but we found no other tumor lesions. The histologic diagnosis was based on the presence of signet ring cells filled with cytoplasmic mucus-containing vacuoles compressing and displacing the nucleus into a peripheral crescent alongside the cell wall. The component signet ring cells are variable; it is >75% in almost half the cases.[5] Our first case was an invasive tumor, which extended to the perivesical fat. Indeed, the insidious progression of this entity explains the local character already advanced at diagnosis. At the time of diagnosis, about 25% of patients have distant metastases and approximately 50% have stage IV disease.[6]

Primary signet ring cell carcinoma of the urinary bladder has an ominous prognosis as it is diagnosed at an advanced stage. The treatment is surgical and consists of an early radical cystectomy. Resection is often incomplete with no clear margins on the specimen.[7] Considering the rarity of this histologic type of tumor, there is no consensus regarding the management after surgical care. Chemotherapy and radiation therapy are discussed. Adjuvant chemotherapy with 5-fluorouracil associated with adriablastin or bleomycin seems to give favorable responses, by analogy with stomach plastic linitis.[8] Our second patient had no palliative chemotherapy because of altered general condition.

Conclusion

The primary SRCC of the urinary bladder is a rare and aggressive tumor; the histologic type justifies a surgical strategy associated with a multidisciplinary approach. Prognosis is poor although some patients may benefit from surgical resection. Adjuvant chemotherapy should be discussed even if consensual attitude has not been defined.

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