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

This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/3.0/).
* Corresponding author. Tel.: +2-169-764-1475. E-mail address: wiembouk@yahoo.fr (W. Boukettaya).

A R T I C L E   I N F O
Article history:
Received 27 February 2014
Accepted 5 March 2014
Available online 8 April 2014

Keywords:
Signet ring cell carcinoma
Urinary bladder
Linitis plastica
Adenocarcinoma

A B S T R A C T
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, anatomic clinical, and evolution specificity to optimize its therapeutic management.

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 hydronephrosis 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. 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.
Cancers.2

Typically described, and its incidence is about 0.24% of bladder

The component signet ring cells are variable; it is >75% in almost half the cases.2 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.3

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.2 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 limitis.3 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.

References

1. Cruz-Gonzáles GH, Sánchez-Salas RE, Palmer-Román KJ, et al. Primary signet ring cell bladder adenocarcinoma. Case report with a multidisciplinary therapeutic approach. Actas Urol Esp Sep. 2007;31:919–922.
2. Singh J, Zherebitskiy V, Grynspan D, et al. Metastatic signet ring cell adenocarcinoma of the bladder: responsive to treatment? Can Urol Assoc J. 2012;6:e15–e19.
3. Ozeki Z, Kobayashi S, Machida T, et al. Transitional cell carcinoma of the urinary bladder accompanied by signet-ring cell carcinoma: a case report. Hinyokika Kiyo. 2003;49:411–413.
4. Krichen Makni S, Ellouz S, Khabir A, et al. Primary signet ring cell carcinoma of urinary bladder. A case report. Cancer Radiothéér. 2005;9:332–334.
5. Del Sordo R, Bellezza G, Colella R, et al. Primary signet-ring cell carcinoma of the urinary bladder: a clinicopathologic and immunohistochemical study of 5 cases. *Appl Immunohistochem Mol Morphol.* 2009;17:18–22.

6. Akamatsu S, Takahashi A, Ito M, Ogura K. Primary signet-ring cell carcinoma of the urinary bladder. *Urology.* 2010;75:615–618.

7. Spinoit AF, Petit T, Elalouf V, et al. Signet-ring cell primitive bladder carcinoma: a rare and aggressive tumor. *Progrès en urologie.* 2011;21:651–653.

8. Calabro F, Sternberg CN. Neoadjuvant and adjuvant chemotherapy in muscle-invasive bladder cancer. *Eur Urol.* 2009;55:348–358.