Adénocarcinome à cellules indépendantes de la vessie : A propos de deux cas

Primary signet-ring cell carcinoma of the urinary bladder: A report of two cases

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RÉSUMÉ

Introduction : Les adénocarcinomes vésicaux primitifs représentent environ 2% des tumeurs de vessie. L'adénocarcinome à cellules indépendantes est une variante rare des adénocarcinomes vésicaux. Il est caractérisé par le retard au diagnostic et par le mauvais pronostic. L'objectif du travail est d'étudier les caractéristiques cliniques, anatomopathologiques et évolutives de cette entité rare.

Observations cliniques : Il s'agit de deux patients un homme et une femme âgés respectivement de 53 ans et 70 ans. Le motif de consultation principal était des lombalgies avec pollakiurie, brûlures mictionnelles et hématurie totale. L'AUSP a montré une opacité de 8cm sur le trajet de l'uretère lombaire. L'échographie a révélé une lésion bourgeonnante intra vésicale avec un calcul du bas uretère gauche de 5mm. Une cystoscopie a été réalisée montrant une lésion tissulaire d'allure atypique blanchâtre effilochée, solide à la coupe. Une IRM pelvienne réalisée chez la femme, a montré une masse tissulaire mal limitée au col utérin envahissant la paroi antérieure du vagin. Une résection endoscopique de la tumeur vésicale a été réalisée dans un cas et une biopsie de la lésion vésicale pour l'autre cas.L'étude immunohistochimique a montré une positivité des cellules tumorales à la cytokératine 7. Le diagnostic d'adénocarcinome à cellules indépendantes a été retenu.

Conclusion: Le carcinome à cellules indépendantes de la vessie est un cancer agressif d'évolution rapide et de mauvais pronostic. Sa prise en charge est multidisciplinaire et fait appel à la chirurgie. Une chimiothérapie adjuvante doit être discutée même si une attitude consensuelle n'a pas été définie.

Mots-clés

Adénocarcinome à cellules indépendantes, vessie, linite plastique

SUMMARY

Introduction: Primitive bladder adenocarcinomas account for about 2% of urinary bladder tumors. Primary signet-ring cell carcinoma is a rare variant of bladder adenocarcinoma. It is characterized by a late diagnosis and a poor prognosis. The aim of the study was to discuss the clinical, pathological and evolutive features of this rare entity.

Clinical Observations: We report two patients a 53-year-old man and a 70-year-old woman. The main reason for consultation was right back pain, mictional burn and total hematuria. Abdominal ultrasound showed a thickened bladder wall measuring 8 mm in thickness. Ultrasonography revealed a budding intravesical lesion with a calculation of the lower left ureter measuring 5mm of great diameter. Cystoscopy showed an atypical tissular lesion frayed whitish and solid in the cut. Pelvic MRI performed in women showed a cervix ill-limited tissue mass invading the anterior vaginal wall. A transurethral resection of the bladder tumor was performed in one case and a biopsy of the bladder lesion for the other case. The immunohistochemical study showed positive tumoral cells to CK7. The diagnosis of signet ring cell adenocarcinoma of the bladder was established.

Conclusion: Signet ring cell adenocarcinoma of the bladder is an aggressive tumor of rapid development and poor prognosis. Its management is multidisciplinary and involves surgery. Adjuvant chemotherapy should be discussed even if consensual attitude has not been set.

Key-words

Signet ring cell carcinoma, Urinary bladder, Linitis plastica.

INTRODUCTION

Bladder adenocarcinoma is a rare histological subtype of bladder cancers, with variant subtypes including primary signet-ring cell carcinoma (PSRCC) which accounts for only 0.5- 2% of all primary neoplasms of the bladder (1). To our knowledge, Less than 100 cases of signet ring cell adenocarcinoma of the urinary bladder have been reported (2). The origin of signet-ring cell in the gallbladder is unknown. We presented two rare cases of signet-ring cell carcinoma of the gallbladder.

CASES REPORTS

Case report 1:

A 70-year-old woman with a history of ischemic stroke 5 years ago, presented with right back pain, mictional burn and total hematuria. Ultrasonography revealed a budding intravesical lesion with a calculation of the lower left ureter measuring 5mm of great diameter. Cystoscopy showed an atypical tissular lesion frayed whitich and solid in the cut. A biopsy of the bladder lesion was performed. Several chips were sent for histological examination. It showed a malignant epithelial proliferation forming clusters of cords and dissociating the muscular wall. The immunohistochemical study showed positive tumor cells to cytokeratin 7. The diagnosis of signet ring cell carcinoma of the bladder was made. Three years after the diagnosis, the patient is still alive.

Case report 2:

A 53-year-old man with no pathological history consulted for urinary disorders with a gross hematuria. Abdominal ultrasound showed a thickened bladder wall measuring 8 mm in thickness. Cystoscopy showed an inflammatory bullous lesion on the right side face and bottom that suggest carcinoma in situ or chronic cystitis. The gastrointestinal fibroscopy showed a nodular antral gastropathy. Colonoscopy revealed a sessile polyp of the left colon which was resected. The PSA was normal 1,2ng/ml. A thoraco-abdominal-pelvic CT scan objectified circumferential tissue thickening of the bladder wall above the level of the anterior right side wall estimated to be more than 15mm with irregular surface with infiltration of perivesical fat compared peri-bladder without satellite node. The patient had an endoscopic resection of the lesion. Histological examination concluded signet ring cell carcinoma of the bladder, invading the muscular wall (Fig.1). The immunohistochemical study showed positive tumor cells to cytokeratin 7 (Fig.2), negative to cytokeratin 20 and prostatic specific antigen. The patient didn't have a cystectomy. Two years after the diagnosis, the patient is still alive.

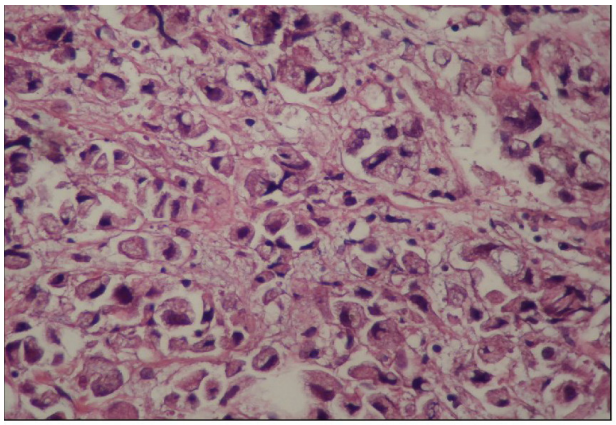

Figure 1 : HEx400: Primary signet-ring cell carcinoma of the urinary bladder- Tumoral cells with a large vacuole pushing the nucleus to one side called signet ring cells.

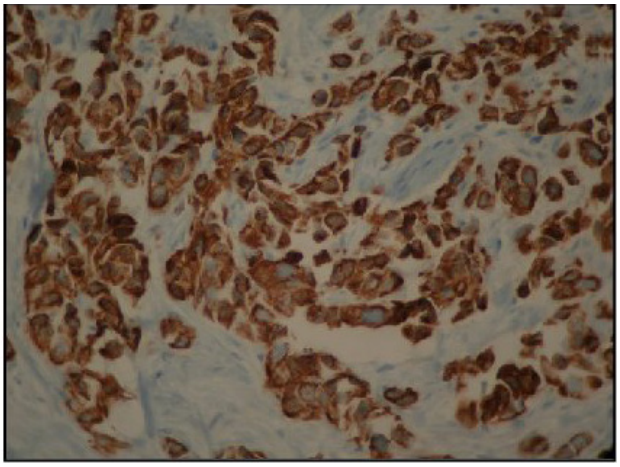

Figure 2 : IHCx400: Primary signet-ring cell carcinoma of the urinary bladder- strong immunostaining to cytokeratin 7.

DISCUSSION

Primary signet-ring cell carcinoma of the urinary bladder is a relatively rare subtype of adenocarcinoma and comprises only 0.24% to 2% of all primary epithelial urinary bladder tumors (3, 4). The first two cases were described by Saphir et al in 1955. Generally, this neoplasm occurs

in middle age with a male preponderance and is usually diagnosed at an advanced stage, usually demonstrating a subsequently poor prognosis (5). Clinically, the common presenting symptoms were irritative voiding symptoms and hematuria. Urinary retention and flank pain due to ureteral obstruction were less common (6). The lesion is described in cystoscopy as pedunculated, polypoid, sessile, and ulcero-infiltrative (7). Primary signet-ring cell carcinoma of the urinary bladder has the same histology as that of the gastrointestinal tract, breast, lung, gallbladder, and prostate; therefore, further evaluations for other primary sites are mandatory to exclude metastasis (7, 8). The histogenesis of primary signet cell cancer in the bladder is not well-understood, and includes metaplasia of transitional cell carcinoma (9, 10). Treatment modalities for signet ring cell carcinomas include surgery, radiotherapy, and chemotherapy. Radical cystectomy is the only therapy that offers the possibility of a cure when the tumor is localized.

This disease usually presents at advanced stages, and patient survival is therefore usually poor with a reported mean 5-year survival rate of 27%-30% (11, 12). One quarter of the patients were found to have distant metastases at the time of diagnosis and 60% of patients died within 1 year (13).

CONCLUSION

Primary signet-ring cell carcinoma is a rare and aggressive tumor; the histological type justifies a surgical strategy associated with a multidisciplinary approach. Recently, successful

Treatments with chemotherapy alone have been reported.

No conflicts of interest.

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