A case of signet ring cell adenocarcinoma of the bladder with spontaneous urinary extravasation

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ABSTRACT

Primary signet ring cell adenocarcinoma (PSRCC) of the bladder is a relatively rare variant of adenocarcinoma of the bladder with poor prognosis. Also PSRCC of the bladder presenting with spontaneous urinary extravasation is very rare. We present the case of a 48-year male who presented with spontaneous urinary extravasation and was diagnosed to have PSRCC of the urinary bladder on evaluation. He was treated with radical cystectomy and adjuvant chemotherapy. This report emphasizes the need to rule out other primary sites of adenocarcinoma in the body, which may metastasize to the urinary bladder.

Key words: Primary signet ring cell adenocarcinoma of the urinary bladder, spontaneous urinary extravasation, CK 7, CK 20

INTRODUCTION

Primary signet ring cell adenocarcinoma (PSRCC) of the urinary bladder presenting with spontaneous urinary extravasation is a rare variant. We present the case of a 48-year male patient who presented with spontaneous urinary extravasation and was diagnosed to have signet ring cell adenocarcinoma of the urinary bladder. Further workup to rule out other primary sites in the body showed it to be primary in origin. He underwent a radical cystectomy with adjuvant chemotherapy. He is alive and free of recurrences till 1 year of follow up. This case is discussed for its rare presentation and also with the diagnostic dilemma it represents as other primary adenocarcinomas as from GIT, lungs, breast, prostate have to be ruled out before the diagnosis.

CASE REPORT

A 48-year male presented with dull aching right loin pain since 3 months. There was no history of hematuria. Blood and urine examination were normal. Ultrasound examination of abdomen showed right hydroureteronephrosis with right periureteric collection. Intravenous pyelography showed urinary extravasation from the mid-ureter with right hydroureteronephrosis. CT scan of abdomen showed right hydroureteronephrosis with urinary extravasation from the right mid-ureter with thickening of the right lateral wall of the bladder [Figure 1]. Cystoscopy showed a sessile tumor in the right lateral wall. Transurethral resection

Figure 1: CT scan abdomen reconstruction showing right sided hydroureteronephrosis with right urinary extravasation from the ureter and thickening of the right lateral wall of the bladder
biopsies were taken. Histopathology showed signet ring cell feature with abundant mucin and confluent necrosis with muscle infiltration [Figure 2]. Immunohistochemistry of the specimen was positive for CK 7, CK 20, and HMW [Figure 3]. An extensive search for other sites of primary tumor was done. Upper and lower gastrointestinal endoscopy were negative for any tumor. Serum carcino-embryogenic antigen (CEA) was undetectable. Diagnostic laparoscopy of the abdomen also failed to pick up any primary elsewhere. 18FDG-PET (fluoro deoxy glucose-positron emission tomography) scan of the abdomen showed avid FDG uptake in the bladder [Figure 4]. This proved it to be PSRCC of the bladder.

The patient underwent radical cystectomy with ileal conduit done. The resected specimen showed cords and strands of cells filled with mucin infiltrating up to the perivesical fat (pT3NoMo). He was given four cycles of cisplatin and gemcitabine combination chemotherapy. He is alive and free of any recurrences for 1 year of follow-up.

DISCUSSION

PSRCC of the bladder is a relatively rare variant of adenocarcinoma of the bladder comprising only 0.5-2% of all primary cancers of the urinary bladder. Most of the patients are in their seventh to eight decade and there is male predominance. In our case, the patient developed signet ring cell adenocarcinoma in the fifth decade.

The usual clinical presentation of PSRCC of the bladder does not vary from other bladder malignancies. Hematuria is the most common presenting symptom. Mucinuria has been reported in 3-12% of the patients. Iqbal et al. reported a case of infiltrating PSRCC of the bladder which presented with acute renal failure and acute duodenal obstruction mimicking retroperitoneal fibrosis on imaging. Our patient had spontaneous urinary extravasation as the presenting symptom caused by the backpressure effect which is the only one reported till date in the world literature.

One of the main problem in case of PSRCC of the bladder is the exclusion of metastasizing primary tumor at any other site in the body and therefore the presence of a predominantly signet ring cell component should lead to a thorough search to exclude primary at any other site in the body. Patients with PSRCC of the bladder present with symptoms related to the urinary bladder, while other primaries present with secondaries to the bladder present with extravesical symptoms. Also the presence of other elements as small cell carcinoma, transitional cell carcinoma, and carcinoma in situ would help to rule out primary bladder tumor from metastases to the bladder. The presence of glandular metaplasia has long been associated with a risk of malignancy, particularly adenocarcinoma of the bladder. However, cystitis glandularis is a well-known response of the urothelium to chronic irritation and the relationship between cystitis glandularis and adenocarcinoma of the bladder is no longer accepted.
Adenocarcinoma arising in the urachus must be distinguished from adenocarcinoma of the bladder as it can be managed with partial cystectomy while adenocarcinoma of the bladder mandates radical cystectomy. Chuang-Gang et al. reported a case of signet ring cell adenocarcinoma in the urachus.[3] Johnson et al. proposed criteria to classify tumor as urachal in origin: (1) tumor in the bladder dome, (2) sharp demarcation between the tumor and the surface epithelium, and (3) exclusion of primary adenocarcinoma located elsewhere with spread to the bladder.[4]

The histogenesis of nonurachal signet ring cell adenocarcinoma of the urinary bladder is unclear as mucin-producing glands and columnar epithelium are not present in the normal bladder. Such adenocarcinomas are thought to arise from totipotent cells of the transitional epithelium or from the remnants of the cloaca.[5]

PSRCC of the urinary bladder has an ominous prognosis as it is diagnosed at an advanced stage. Cystoscopy is usually normal or may show edematous, bullous, or erythematous mucosa. If a mass lesion is present, it is pedunculated or sessile.

PSRCC of the urinary bladder has the same histology as adenocarcinoma arising from the gastrointestinal tract, prostate, female reproductive tract; therefore an extensive search should be done to rule out any other primary site of adenocarcinoma metastasizing to the urinary bladder.

Although there is no specific marker for the diagnosis or prognosis of PSRCC of the urinary bladder, elevated CEA has been reported to be elevated. Yamamoto et al. reported that the serum CEA levels normalized after surgery and increased as the disease progressed. In our case, the serum CEA level was 0 ng/ml.

Treatment modalities of PSRCC of the bladder are surgery, radiotherapy, and chemotherapy. Surgical options are transurethral resection, partial cystectomy (for urachal lesion or tumor in diverticulum), and radical cystectomy with urinary diversion. Unfortunately, no standard chemotherapy exists for PSRCCs of the bladder because of their rarity. The effects of chemotherapy on signet ring cell carcinoma arising from other organs provide limited information, because PSRCC of the urinary bladder may have characteristics differing from those of such tumors. Ota et al.[7] reported effective treatment with intra-arterial chemotherapy with cisplatin and metotrexate and radiation therapy. Cobo-Dols et al. reported a case of PSRCC treated successfully with total cystectomy followed by systemic chemotherapy with cisplatin and gemcitabine, a standard combination for transitional carcinoma of the urinary bladder.[8] Hirano et al. reported success with primary treatment of signet ring cell adenocarcinoma of the bladder with intra-arterial chemotherapy.[9]

So, in the case of diagnosis of adenocarcinoma of the urinary bladder, all the primary sites of adenocarcinoma as gastrointestinal, lung, breast should be ruled out before labeling it as a PSRCC of the urinary bladder.

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