Primary signet-ring cell linitis plastica type adenocarcinoma of the urinary bladder

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KEY WORDS

linitis plastica ◗ adenocarcinoma ◗ urinary bladder ◗ lower urinary tract symptoms

ABSTRACT

Adenocarcinoma of the urinary bladder is unusual, constituting about 2% of all primary carcinomas of the bladder. Bladder adenocarcinoma consisting chiefly of signet ring cells and those that have a *linitis plastica* pattern of infiltration is extremely rare. Not many cases have been reported in literature. To the best of our knowledge less than 100 cases of signet-ring cell adenocarcinoma of the urinary bladder have been reported and that of *linitis plastica* are even less. We hereby report an unusual case and provide a relevant review of literature.

CASE REPORT

A 53 year old female presented with complaints of irritative lower urinary tract symptoms (LUTS), mainly frequency and urgency, for duration of 4 weeks. She also had complaints of urge incontinence for 10 days. Routine investigations including renal functions were normal. General physical examination was unremarkable.

Ultrasonography (USG) of the abdomen showed a thickened bladder wall with bilateral hydroureteronephrosis. Contrast enhanced computer tomography (CECT) of the abdomen showed diffuse thickening of bladder wall with perivesical fat stranding, bilateral hydroureteronephrosis (left >right), multiple enlarged pelvic lymph nodes (Fig. 1). She was further evaluated with cystopanendoscopy (CPE); findings were: small contracted bladder, bilateral ureteric orifices were not seen, and bladder mucosa was thickened. Bladder biopsy was taken from multiple sites. The histopathology (HPE) report showed poorly differentiated adenocarcinoma of the urinary bladder with signet-ring cell features.

Her course of illness was complicated by issues that she developed contrast induced nephropathy (s-creatinine – 4.5 mg%), right hemi-paresis, and aphasia. Neurological involvement created a suspicion that she had brain metastasis. Whole body PET scan showed an FDG (fluorodeoxyglucose)-avid thick bladder wall (Fig. 2) and there was no FDG-avid lesion in any other part of body including the brain. MRI of the brain showed an area of hypoperfusion therefore, a diagnosis of right middle cerebral artery stroke was made. She was managed with indwelling catheterization, hydration, aspirin, and antihypertensives. We waited until the time when her kidney

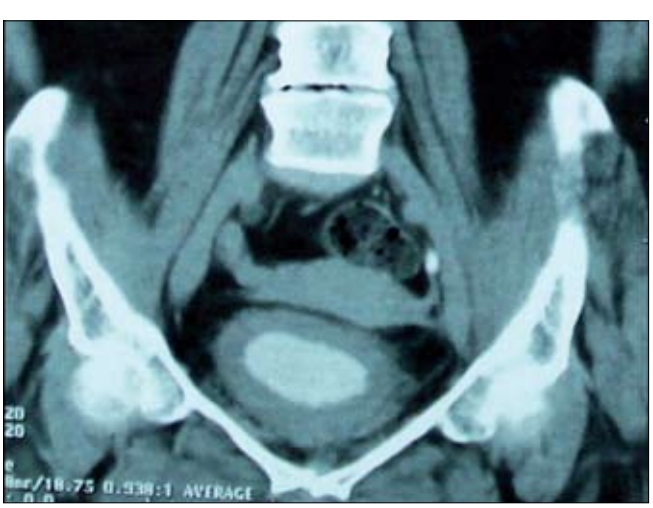

Fig. 1. Thickened bladder wall with perivesical fat streaking.

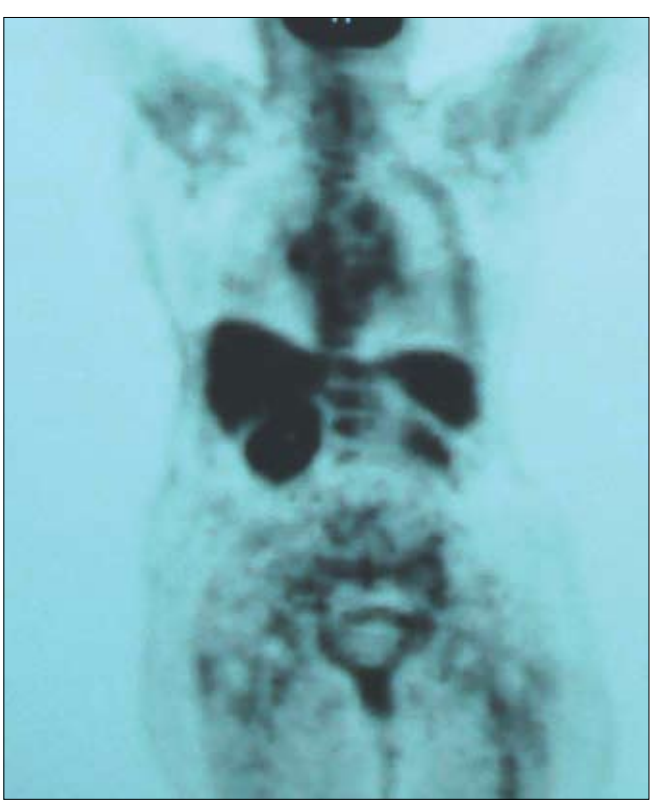

Fig. 2. FDG-avid bladder wall.

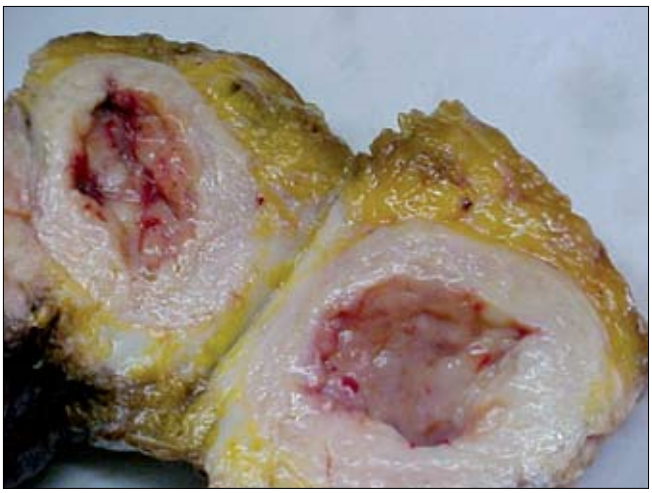

Fig. 3. *Linitis plastica* of urinary bladder, severely compromised lumen.

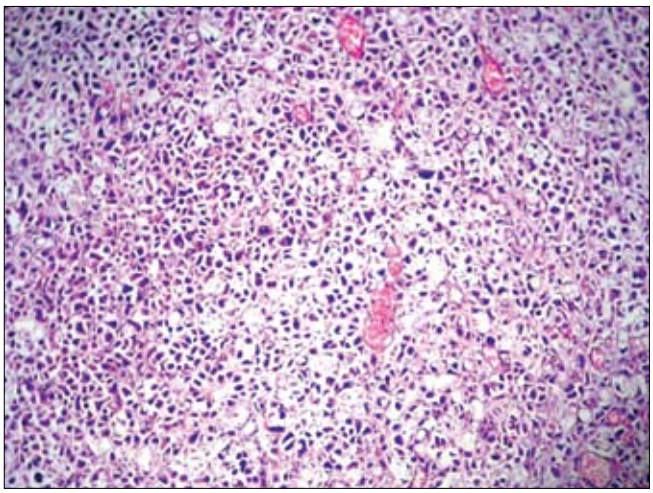

Fig. 4. Poorly differentiated adenocarcinoma of the urinary bladder with signet-ring cell features.

functions normalized, but there was no marked improvement in her neurological status. She was not in a state to give informed consent, but the fact that she has a highly aggressive bladder tumor where metastasis was a concern, radical surgery was the only option possible. We discussed these issues with family members. In consultation with the neurologist, the decision to perform surgery with the consent of next of kin was made. Anterior pelvic exenteration with pelvic lymphadenectomy and ileal conduit urinary diversion was done. A cut section of bladder showed a small contracted bladder and thickened bladder wall with typical features of *linitis plastica* (Fig. 3). Post operative recovery was uneventful.

The histopathology revealed poorly differentiated muscle invasive adenocarcinoma of the urinary bladder with signet-ring cell features (more than 70% signet-ring cells seen); pathological stage T4bN2M0 (Fig. 4). Immunohistochemistry was positive for cytokeratin and epithelial membrane antigen. She had received Gemcitabine and cisplatin based combination chemotherapy. At 7 months of follow-up she is doing well.

DISSCUSSION

Primary pure adenocarcinomas of the urinary bladder are infrequent. Based on the histological appearance; primary adenocarcinoma is classified as glandular carcinoma, colloid carcinoma, papillary carcinoma, signet-ring cell adenocarcinoma, and clear cell carcinoma. Primary signet-ring cell carcinoma of the urinary bladder is an extremely rare tumor, accounting for approximately 0.24% of all bladder malignances [1, 2]. Signet-ring cell carcinoma of the urinary bladder can be primary - arising from the bladder wall or urachus remnants or metastatic from tumors originating in the stomach, colon, or breast. Saphir first described primary signet-ring cell cancer of the urinary bladder in 1955. Less than 100 cases have been reported in the literature since then [1].

In cases of signet-ring cell adenocarcinoma of the urinary bladder, metastasis from the gastrointestinal tract is possible, although full evaluation to detect the primary site should be done [3]. Most cases of signet-ring cell carcinomas show a diffuse *linitis plastica*-like infiltration of bladder wall, while some may show discrete masses as well [4]. The prognosis of *linitis plastica* carcinoma of the urinary bladder is poor. One reason being the delay in presentation, because irritative LUTS are a more common presentation than hematuria [5]. The tumor does not respond to radiation therapy or combination therapy. Radical cystectomy in early stages of bladder cancer may bring forth some hope for the control of this lethal disease [1, 5].

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