CASE REPORT

Signet-ring cell carcinoma arising from the urinary bladder

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Introduction: *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.*

Case report: *We report a case of a 45-year-old woman who was admitted with gross hematuria. Cystoscopy revealed a necrotic tumor on the left bladder wall. A transurethral biopsy showed signet-ring cell carcinoma. The bladder tumor was diagnosed as the primary one. Radical cystectomy was performed with ureterosigmoidostomy (Mainz pouch II). Histological examination showed a primary signet-ring cell carcinoma of the bladder (pT3bN0M0). Following surgery, the patient received adjuvant chemotherapy with cisplatin and fluorouracil.*

Conclusions: *Primary signet-ring cell carcinoma of the urinary bladder is an extremely rare tumor, accounting for approximately 0.24% of all bladder malignances. Patients with this type of cancer generally have a poor prognosis. However, our patient is free of disease 5 years after radical cystectomy.*

Key Words: signet-ring cell carcinoma, urinary bladder

Introduction

Adenocarcinoma stemming from the urinary bladder accounts for 0.5% to 2% of all bladder cancers.[1] Pathological examination is used to distinguish signet-ring cell and clear cell cancers of the bladder.

Signet-ring cell carcinoma is characterized by the intercellular, mucous vacuole compressing the hyperchromatic nucleus onto the side of the cell.[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 from 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. Primary signet-ring cell carcinoma of the urinary bladder is an extremely rare tumor, accounting for approximately 0.24% of all bladder malignances.

Patient survival time is usually short. The mean 5-year survival rate is 27%.[3]

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Case report

In 2002, a 45-year-old woman was admitted to our hospital with hematuria, urinary frequency, abdominal pain, and irritable voiding. Past medical history revealed that she had undergone a Caesarian section and had developed an iliac vein thrombosis in the postoperative period. An abdominal ultrasound examination showed moderate dilation of the renal pelvis and a 2 cm thickening of the left wall of the bladder. Cystoscopy showed a tumor with a wide, necrotic surface.

We performed a transurethral bladder tumor resection. A histological examination of the tumor led to a diagnosis of signet-ring cell carcinoma. Since a gastrointestinal (GI) origin of the tumor was suspected, the patient underwent a gastroscopy and colonoscopy, but there was no evidence of GI abnormalities. Abdominal surgical exploration was negative for any evidence of contiguous tumor from the GI tract. We performed radical cystectomy during which we removed the patient's left ovary and uterus, excised the frontal wall of the vagina, and performed a lymphadenectomy of obturator fossa lymph nodes. Urinary diversion was accomplished by the Mainz II procedure with the ureters introduced into a pouch created from sigmoid flexure.

The histological examination revealed T3b stage signet-ring cell carcinoma infiltrating the perivesical fat tissue, which had not metastasized to regional lymph nodes or to the surrounding organs (pT3bN0M0). We administered four cycles of cisplatin and fluorouracil (5-FU) as adjuvant treatment.

One year later, a computed tomography (CT) scan did not show any lymphadenopathy, but showed what appeared to be an up to 4 cm solid mass in the right ovary. We carried out an abdominal surgical exploration and a right oophorectomy, but did not find any evidence of metastasis. The patient had temporal peripheral dysesthesia, which was likely a complication from chemotherapy, and which disappeared with conservative treatment.

The patient had a positron emission tomography (PET) scan performed 2 years after the diagnosis had been made, and it did not show any evidence of a malignancy. The patient is currently symptom-free 5 years after her diagnosis of primary signet-ring cell carcinoma of the bladder, Figures 1, 2 and 3.

Discussion

The pathogenesis of primary signet-ring cell carcinoma of the bladder has not been determined. Many experts suggest that mesonephric remnants of the trigone of the bladder might be the source of this cancer.

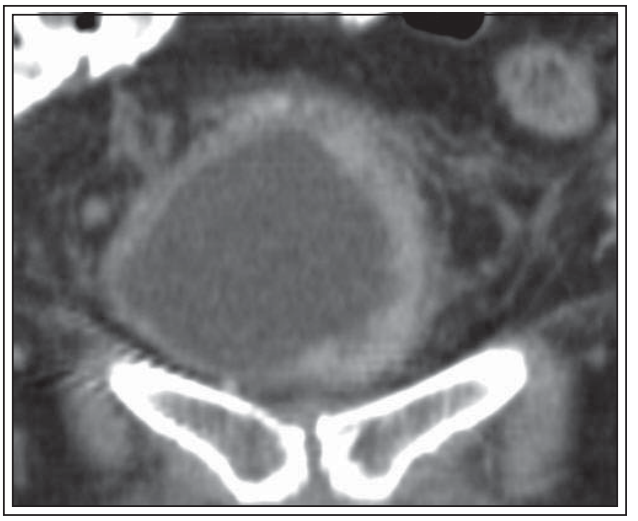

Figure 1. CT scan of the bladder showing the primary tumor.

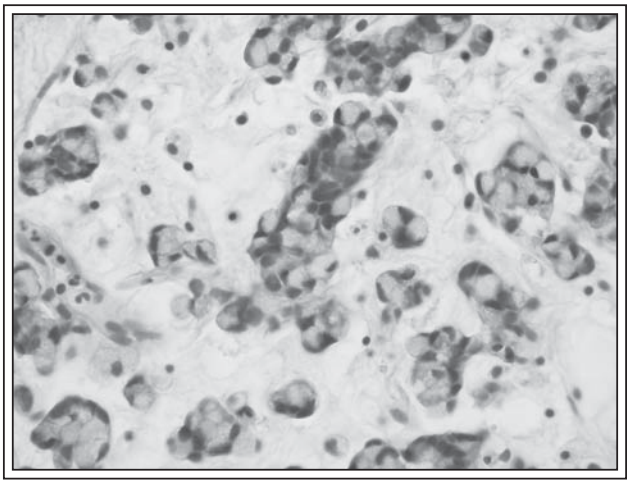

Figure 2. HE staining of signet-ring cell carcinoma of the bladder.

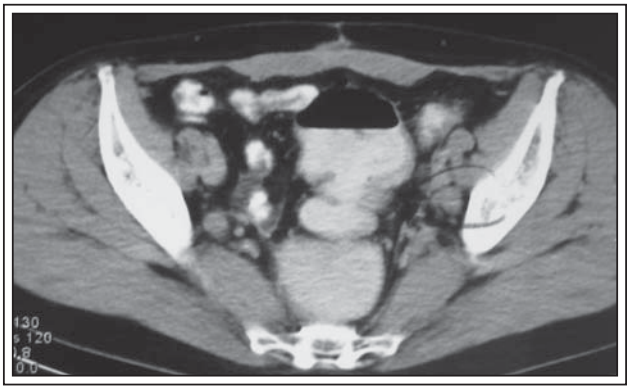

Figure 3. CT scan of the bladder after cystectomy shows possible local tumor recurrence.

However, this would not explain signet-ring cell carcinoma in other locations.[4,5] Others suggest that malignant transformation of urothelial metaplasia might be caused by bladder irritation. Some experts emphasize the pathologic role of pluripotent cells of transitional epithelium.[6]

Based on literature reports, primary signet-ring cell cancer is mainly found in individuals who are aged 28 to 83 years old, and 78% of cases are found in men. Patients present with nonspecific complaints. In 65% of cases, patients are admitted to hospital due to hematuria. The patient in the current case also reported abdominal pain and voiding irritation. A physical examination often reveals a lower urinary tract obstruction (caused by the tumor), a palpable abdominal mass, or a thickening of the inner wall of the rectum.

Findings from imaging tests are not specific either, since they usually show thickening of the bladder wall. An IV contrast CT examination can show the lack of filling and the dilatation of the upper urinary tract.

A cytological examination of urine might show signet-ring cells. The serum tumor marker carcinoembryonic antigen (CEA) levels may be elevated.

A diagnosis of signet-ring cell cancer of the urinary bladder is based on results from a transurethral biopsy. Following this diagnosis, examination of other organs such as the prostate, stomach, and colon is indicated.[1]

In signet-ring cell cancer of the urinary bladder, a histological examination typically shows cells containing mucin vacuoles. These vacuoles do not stain with hematoxylin-eosin (HE) stain, but are highlighted with periodic acid-Schiff (PAS) reaction. In one-third of cases, transitional or squamous elements can be observed in addition to adenocarcinoma.

The life expectancy for patients with signet-ring cell carcinoma arising from the urinary bladder is often short, since the cancer progresses rapidly. The disease usually presents in advanced stages because the patient may remain symptom-free for a long time. One-quarter of patients have distant metastases at the time of diagnosis and 60% of patients die within 1 year of diagnosis.

If distant metastases are discovered, palliative therapy consisting of a partial cystectomy and selective intraarterial chemotherapy might be considered. However, longer, disease-free survival time was recorded in patients who underwent radical cystectomy.[7]

Signet-ring cell carcinoma arising from the urinary bladder is not radiosensitive. In cases where the tumor contains transitional cell elements, the transitional cell component may be destroyed by radiation, but the remaining tumor grows more rapidly. Based on the treatment of signet-ring cell carcinomas arising from other organs, the patient is usually given a combination of cisplatin, fluorouracil, and doxorubicin. With bladder adenocarcinoma, the combination of cisplatin and mitomycin C appears to be the most effective treatment, however survival usually does not improve significantly, and the combination therapy does not result in therapeutic responses in patients in other histological subgroups.[8] □

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