Primary Leiomyosarcoma of the Ureter

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Leiomyosarcoma is a rarely seen neoplasm of the ureter, with less than 20 cases being reported in the literature to date. It is important to distinguish leiomyosarcoma from rhabdomyosarcoma, with the aid of immunohistochemical markers. We report the clinical features, histology, imaging and treatment of ureteral leiomyosarcoma in a female patient. [Asian J Surg 2008;31(4):191–4]

Key Words: leiomyosarcoma, ureter

Introduction

Malignant neoplasms of the ureter are typically transitional cell carcinoma with squamous cell carcinoma, and adenocarcinoma comprises nearly all of the remaining cases. Leiomyosarcoma is a rarely seen neoplasm of the ureter with fewer than 20 cases being reported in the literature to date.1,2 It is important to distinguish leiomyosarcoma from rhabdomyosarcoma, with the aid of immunohistochemical markers. Here we report a case of ureteral leiomyosarcoma in a female patient.3

Case report

A 47-year-old woman presented to our hospital with a dull pain in the right flank, which had persisted for 4 months. She did not complain of any history of urological calculus and no gross haematuria was noted.

Ultrasonography identified right moderate hydronephrosis with dilation of the proximal ureter. Computed tomography (CT) showed the presence of a soft mass (3 × 3 cm), which extended from the right ureters, with dilation of the proximal ureter and renal pelvis (Figure 1). The affected ureteral wall possessed surrounding incrassation, which formed a 3.5 × 3.5-cm soft-tissue mass. Furthermore, the lumen of the affected ureter appeared to be irregular in shape. After injection of contrast medium, the mass was greatly enhanced, with a clear portion in the nearby tissues. The distance of the mass from the renal pelvis was about 8 cm. Further examination identified microscopic haematuria, and ureteroscopy revealed a soft mass that obstructed the lumen of the ureter. A biopsy of the mass was performed, and pathological examination identified characteristics of leiomyosarcoma.

The patient was surgically treated by right nephroureterectomy, with retroperitoneal lymphadenectomy. The soft mass was 3.5 cm in diameter, with proximal ureter dilation (Figure 2). Pathological examination showed that the cancer was composed of fascicles of interlacing, moderately large, spindle-shaped cells, with abundant eosinophilic cytoplasm. Immunohistochemistry showed strong staining for smooth muscle actin (SMA) (Figures 3 and 4). A diagnosis of leiomyosarcoma of the right ureter was made, and three retroperitoneal lymph nodes were dissected and confirmed as negative for tumour by pathological examination. The cutting margin of the ureter near the bladder was negative and the kidney was intact. No adjuvant chemotherapy was given after surgery. The patient’s convalescence was uneventful at 6 months after surgery. However, the patient presented again with emaciation

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Figure 1. Computed tomography of leiomyosarcoma of the ureter. (A) A soft mass (3 × 3 cm) extending from the right ureter. The affected ureteral wall possessed surrounding incrassation. The mass was substantially enhanced with injection of contrast medium. (B) Dilation of the proximal ureter and renal pelvis.

Figure 2. Gross appearance of leiomyosarcoma of the ureter. (A, B) Grossly, the appearance was a solid, greyish, lobulated mass with a trabeculated surface. The affected ureteral wall possessed surrounding incrassation, and formed a 3.5 × 3.5-cm soft-tissue mass. The lumen of the affected ureter appeared to be irregular in shape. The collecting system of the affected kidney had moderate dilation.

Figure 3. Histological appearance of leiomyosarcoma of the ureter. (A, B) Typical architecture of leiomyosarcoma with fascicles of spindle cells arranged at right angles. Cells were non-tapered and may have blunt-ended nuclei; occasional paranuclear vacuoles were evident. Focal nuclear pleomorphism was seen. Tumours were mitotically active, and myxoid change incorporated nodules of the tumour (haematoxylin & eosin, 100×). (B) The tumour had no capsule and the cells encroached upon peripheral normal smooth muscle tissue (haematoxylin & eosin, 100×).
and a dull pain in the same position. CT showed involvement of the retroperitoneal lymph nodes, and consequent therapy was not carried out due to the patient’s personal wishes.

Discussion

Leiomyosarcoma is a rare soft-tissue tumour that accounts for 10–20% of soft-tissue sarcoma, is generally seen in middle-aged patients, and afflicts women more frequently than men. Its usual locations, in order of frequency are: retroperitoneal, intra-abdominal, cutaneous and subcutaneous.

Leiomyosarcoma is a highly malignant tumour with an extremely poor prognosis. The 5-year overall survival rate in patients with soft-tissue sarcoma of all stages remains poor, at only 50–60%, and 5-year disease-free survival is rare. In one study, the 5-year survival of 705 patients with leiomyosarcoma from 22 series was 27.8%. A literature review has revealed only 17 cases of primary leiomyosarcoma of the ureter, and metastasis is reported to have occurred in eight patients, with four having died from the disease. In our case report, the recurrence of node-negative leiomyosarcoma at 6 months after surgical excision is unusual.

An important aspect of the management of leiomyosarcoma is to differentiate the lesion from rhabdomyosarcoma and other spindle-cell neoplasms. This is especially difficult with poorly differentiated high-grade lesions. On immunohistochemistry, leiomyosarcoma is negative for epithelial markers (cytokeratins and epithelial membrane antigens). Positivity for desmin and SMA shows the smooth muscle origin of the tumour and gives a definitive diagnosis. Negative myoglobin, cytokeratin and S-100 help to rule out rhabdomyosarcoma, sarcomatoid carcinoma, and melanoma, respectively.

Leiomyosarcoma shows nodular aggregates and densely packed, interlacing bundles of smooth-muscle cells in the centre. At the margin, strands of smooth muscle cells extend between collagen bundles. Areas of lesser differentiation are present in all tumours to varying degrees, including irregularly shaped, anaplastic nuclei and atypical giant cells with bizarre nuclei. A high mitotic rate remains the main criterion for leiomyosarcoma, and other histopathological features of the tumour include atypical smooth-muscle cells with peripheral extension into the surrounding tissue, pleomorphism, giant cells, and necrosis of the tumour and peripheral tissue.

Only some rhabdomyosarcomas contain cells that are recognizable as rhabdomyoblasts, by virtue of their cytoplasm exhibiting cross-striation, which is best revealed by phosphotungstic acid-haematoxylin staining. Round cells of the embryonal variant tend to have cytoplasmic rims that are stained red by trichrome. The botryoid type is a variant of the embryonal type and is found mainly in mucosa-lined hollow organs. Cells with elongated eosinophilic cytoplasm may be suggestive but not necessarily diagnostic of rhabdomyosarcoma, and may be found in the pleomorphic type. A sclerosing variant has cords of small, round malignant cells embedded in a densely hyalinized matrix that has a chondroid and osteoid appearance.

Immunohistochemical staining of deparaffinized sections shows expression of desmin and muscle-specific actin in the great majority of leiomyosarcomas. Anti-desmin staining is found in 47–85% of superficial leiomyosarcomas, and is less common in higher-grade tumours. Immunohistochemical studies of deparaffinized tissue may show rhabdomyosarcoma cells to be positive for vimentin, which indicates a mesenchymal phenotype. More differentiated cells may be positive for desmin and myoglobin, which is indicative of muscular differentiation.

Wide local excision is considered the treatment of choice with radiation and chemotherapy being offered to patients with positive margins or nodes, or those with bulky disease. Radiotherapy can be considered for larger tumours and/or positive margins, if anatomically feasible. Adjuvant chemotherapy has not been proven to be effective and remains investigational. Treatment for metastatic...
disease is palliative. Active agents include doxorubicin, ifosfamide, gemcitabine and docetaxel.

Since there is no mode of definitive treatment for this rare disease, one must extrapolate from modes of treatment of leiomyosarcoma of the ureter and epithelial tumours of the ureter. In our case, radical surgical excision of the kidney, surrounding tissues, lymph nodes and ureter was accomplished.

We conclude that, although leiomyosarcoma is rarely seen, it should be listed in the differential diagnosis of ureter tumours.

References