Primary myxoid leiomyosarcoma of renal pelvis: Case report and literature review

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\begin{abstract}
Leiomyosarcoma arising from renal pelvis is a very rare disease. A patient was admitted to hospital with left renal colic due to nephrolithiasis, with a filling defect in renal pelvis that was considered to be a blood clot. Diagnosis of leiomyosarcoma was made after visual inspection and biopsy of the exophytic lesion. Laparoscopic radical nephrectomy was performed, histological and immunohistochemical investigation confirmed leiomyosarcoma with mixomatoid component. No adjuvant treatment was performed, the patient remains healthy 5 years after surgery without recurrence. Herein we provide literature review, discussion of the diagnosis and treatment scenario of the patient with renal pelvis leiomyosarcoma.
\end{abstract}

1. Introduction

The vast majority of ureter and renal pelvis tumors have epithelial origin (95%), mainly transitional cell carcinomas (TCC). Sarcoma ranges 0.8–2.7% of upper urinary tract malignancies, 50–60% of which are leiomyosarcomas. We report a case of a renal leiomyosarcoma with mixoid component in 73-year-old male.

2. Case presentation

Patient was admitted to hospital with complaints of left-sided renal colic and hematuria within 2 weeks in 2016. He had extracorporeal shock wave nephrolithotripsy and two ureterolithotripsies ipsilaterally several years before, denied radiotherapy or chemotherapy, was hemodynamically stable with blood pressure of 140/80 mmHg, had Bosniak I right renal cyst and hypertension stage II. Medications taken were valsartan, atorvastatin, captopril in standard dosages. Urinalysis revealed 40–50 rbc/hpf without any atypical cells. Alkaline phosphatase, liver function tests were normal. Blood creatinine 146 μmol/l. There were no signs of lymphadenopathy or metastases.

Ultrasound examination revealed two left kidney stones: in pelvis (15mm) and in lower calyx (16mm). Excretory urography revealed 18 × 14mm filling defect in the dilated pelvis. Computerized tomography (CT) with intravenous contrast administration revealed a filling defect above the stone, which contained round-shaped tissue with clear contours 15 × 10 × 9mm without contrast enhancement, with renal pelvic wall and proximal ureter thickening (Fig. 1).

The stones were removed during percutaneous nephrolithotomy (PCNL). A villous tumor 18 × 15mm with narrow base, originating from uretero-pelvic junction was visualized during nephroscopy. Pinch biopsy microscopic picture concluded tumor to be leiomyosarcoma. Additional Mallory staining colored bundles of elongated cells with orange, noted polymorphism of nuclei and nucleoli, coarse-lumpy structure of chromatin (Fig. 2).

After PCNL, laparoscopic radical nephrectomy was performed without any technical difficulties. It confirmed previous histological diagnosis: low-grade myxoid sarcoma with areas of hemorrhage and necrosis, reactive changes involving surface urothelial layer, peripelvic fat, spreading to the renal parenchyma and vessels - leiomyosarcoma with the myxoid component pT3N0M0G1 stage 3, negative surgical margin (R0) (Fig. 3). No evidence of recurrence appeared during 5-year follow-up period.

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Fig. 1. (a) Kidney ultrasound before PCNL. (b) CT-scan before contrast administration, (c) excretory phase, filling defect in renal pelvis. (d) KUB, (e) excretory urography 10 min, (f) 75 min.

Fig. 2. Histologic investigation. (a) hematoxylin-eosin stain, elongated cells located on myxomatous stroma forming disordered bundles, occurring giant multinucleated leiomyoblasts x10, (b) x40. (c) Mallory stain, noted polymorphism of nuclei and nucleoli, coarse-lumpy structure of chromatin x40.
3. Discussion

Renal pelvis leiomyosarcoma is extremely rare and less than 20 cases have been reported, is solitary tumor, develop from smooth muscle cells of renal pelvis, ureter, vessels, more common in females, twice more often on right side and in immunocompromised patients or in the presence of Epstein-Barr virus. It has relation with penile carcinoma, urolithiasis, xeroderma pigmentosum, and tuberous sclerosis. Cases of renal sarcoma were reported in children 9 months of age, but the highest incidence occurs from 4th to 6th decade.

These tumors are considered prognostically unfavorable. Due to few observations, 5-year survival rate ranges from 29-36% to 50–60%. For low-grade disease 5-year cancer-specific survival can be 90%, decreasing to 30% with high-grade malignancy. For tumor <5cm 5-year overall survival rate is 77%, with larger sizes - 42%. Favorable prognostic factors are tumor size <5cm, low-grade, organ-confined form, no regional lymphadenopathy, R0 after surgery.

Contemporary treatment strategy of leiomyosarcoma is based on radical surgery, following adjuvant chemotherapy with or without radiation therapy. Adjuvant chemotherapy is the subject of controversy: doxorubicin, anthracycline, ifosfamide, gemcitabine in various schemes. In our case we considered only active surveillance to be reasonable. Leiomyosarcoma usually present with flank pain, hematuria, or palpable abdominal mass, mimicking renal cell carcinoma (RCC) and is often characterized by complex of non-specific complaints such as frequent urination, decreased appetite and weight loss. Imaging features are nonspecific: eccentrically infiltrating ureter tumor, forming local thickening, without hyperdense areas on CT-scan. Filling defect can be seen in the collecting system, causing calyx deformation, hydronephrosis with renal insufficiency. Early contrast media enhancement with hypodense areas indicates necrosis and high tumor aggressiveness. Differential diagnosis should be with TCC, retroperitoneal lymphadenopathy, considering lymphoproliferative diseases, metastasis of urogenital tumors, specific infectious processes (tuberculosis), melanoma or lung cancer, retroperitoneal fibrosis, characterized by infiltration, ureter medialization and hydronephrosis.

Grignon suggested following criteria for diagnosis of primary renal sarcoma: there shouldn’t be sarcoma elsewhere to exclude metastatic consequence, tumor should have kidney origin rather than retroperitoneum, sarcomatoid RCC should be excluded.

The diagnosis is based on immunohistochemical staining: positive for smooth muscle antigen (SMA), desmin, calponin, h-caldesmon, components of basal membrane, such as laminin and type IV collagen, and negative for epithelial membranes antigen (EMA), antibodies to CK, S-100 protein, HMB-45 (human melanoma black 45) and CD 117 (CD-34).

4. Conclusion

Given the absence of characteristic radiographic features, postoperative histopathological investigation can be used to confirm the diagnosis of leiomyosarcoma. Contemporary treatment strategy is based on radical surgery. There is no generally accepted method of adjuvant treatment of this category of patients.

Declaration of competing interest

No conflict of interest.

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N/A.

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