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Primary upper pole liposarcoma of the kidney with invasion to inferior vena cava: A case report

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ABSTRACT

Genitourinary tract liposarcoma is considered as the second most commonly reported type of sarcomas. Renal liposarcoma with tumor invasion to inferior vena cava (IVC) is distinctly rare. This tumor has a relatively indolent clinical course with risk of local recurrences (20%–85% rate) after surgery. Angiomyolipomas (AML) are the most important differential diagnosis because both are large fat-containing lesions. Herein, a patient with upper pole kidney liposarcoma with tumor invasion to renal vein and IVC is presented. The optimal management is radical nephrectomy and IVC tumor resection. Chemotherapy and radiotherapy have not shown significant survival benefit.

Introduction

Liposarcoma accounts for at least 20% of all soft tissue sarcoma in adults. Genitourinary tract liposarcoma is considered as the second most commonly reported type of sarcomas. Renal liposarcoma with tumor invasion to inferior vena cava (IVC) is distinctly rare.[1] In this case-report we presented a patient with upper pole kidney liposarcoma with tumor invasion to renal vein and IVC.

Case presentation

A 62-year-old woman presented with intermittent gross hematuria, right upper quadrant dull pain and weight loss for the previous 6 months. Ultrasonography imaging revealed right upper pole kidney mass including echogenic components in favor of fat tissue. Urine analysis confirmed hematuria. Metastatic evaluations consisted of chest radiography, liver function test and serum alkaline phosphatase was normal. Abdominal computed tomography (CT) imaging and magnetic resonance imaging (MRI) (Fig. 1) demonstrated upper pole mass in the right kidney with fat component that invaded into the main renal vein and IVC up to the sub hepatic level. The patient underwent radical nephrectomy and IVC tumor resection using thoracoabdominal incision. The tumor of the main right renal vein and IVC was excised by making a longitudinal incision on the main renal vain. There was no need to reconstruct IVC. We detected no significant adhesion of viable tumor to the IVC (Fig. 2). Final pathology report confirmed primary upper pole well differentiated liposarcoma (3 × 3 cm) with extension to the main renal vein and IVC. Perinephric fat and right renal capsule were involved by tumor. Sections of tumor showed renal tissue which was involved by neoplastic tissue composed of lipoblast cell with sharp and round cytoplasmic lipid vacuole. The cells had hyperchromatic nucleus with eccentric location (Fig. 3). This pathological feature confirmed renal liposarcoma.

Discussion

Liposarcomas of the genitourinary tract are the second most commonly reported type of sarcomas. Liposarcoma accounts for at least 20% of all soft-tissue sarcoma in adults. The peak incidence of liposarcoma is in 5th or 6th decade of life and male predominance.[1] Renal liposarcoma with IVC involvement is distinctly rare.[1] Liposarcoma classifies in four subtypes: well differentiated liposarcoma, dedifferentiated, myxoid and pleomorphic.[1] This tumor has a relatively indolent clinical course with risk of local recurrences (20%–85% rate) after surgery.[3] well-differentiated liposarcomas can be subdivided morphologically into four main subtypes: adipocytic (lipoma-like), sclerosing, inflammatory and spindle cell.[2]

These tumors can primarily originate from renal capsule or renal sinus. The most important prognostic factors for survival is histologic grade that reflects the extent of differentiation and completeness of

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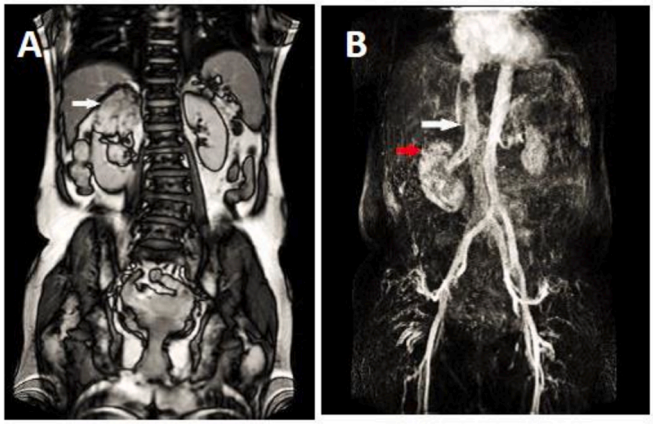

Fig. 1. Abdominal CT imaging and MRI of the case, showing upper pole mass in the right kidney.

resection.[3] Angiomyolipomas (AML) are the most important differential diagnosis because both are large fat-containing lesions. Defect in the renal parenchyma, enlarged vessels in the lesion and Human Melanoma Black-45(HMB-45) positivity are three major radiologic findings helping in differentiating AML from liposarcomas.[4] Early detection of retroperitoneal liposarcomas is difficult because the symptoms of these tumors appear late and are nonspecific. Retroperitoneal liposarcomas grow slowly in the retroperitoneal space and may reach a considerable size before being diagnosed.[1] Generally, the main treatment of liposarcoma of the kidney is radical nephrectomy and IVC tumor resection. In rare cases, a combination of chemotherapy and/or radiotherapy may be used. Other treatment options including adjuvant therapy and kidney preserving surgical procedures need to be developed. The prognosis

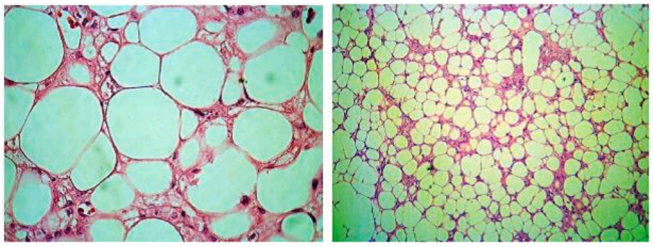

Fig. 3. Pathological feature, showing the cells with hyperchromatic nucleus and eccentric location, confirming renal liposarcoma.

after treatment depends on several factors including the size of the tumor and the histology classification of the tumor. Therefore, there are controversies about the usefulness of additional treatment for liposarcoma of the kidney after radical nephrectomy and its effect on survival of patients. It has been documented that the post-operative radiotherapy leads to reduction of recurrence rate.[5] However, a meta-analysis by Perez et al., failed to show significant radiotherapy-related survival benefits in the treatment of retroperitoneal liposarcoma.

In this case-report, we presented a kidney liposarcoma with direct invasion to renal vein and IVC up to the inferior surface of liver. Viable extended tumor was removed safely by an incision over the renal vain. We found no significant tumor adhesion and gross invasion to renal vein and IVC wall. This pattern of contagious tumor growth thorough renal pelvis, renal vein and IVC is extremely rare. The prognosis of renal sarcoma is especially poor, with a reported 5-yr survival rate of 29%.[1] Matsushita et al. reported that chemotherapeutic agents such as doxycycline and ifosfamide demonstrating activity against metastatic sarcoma, but such chemotherapeutic agents have not shown survival benefit.[1]

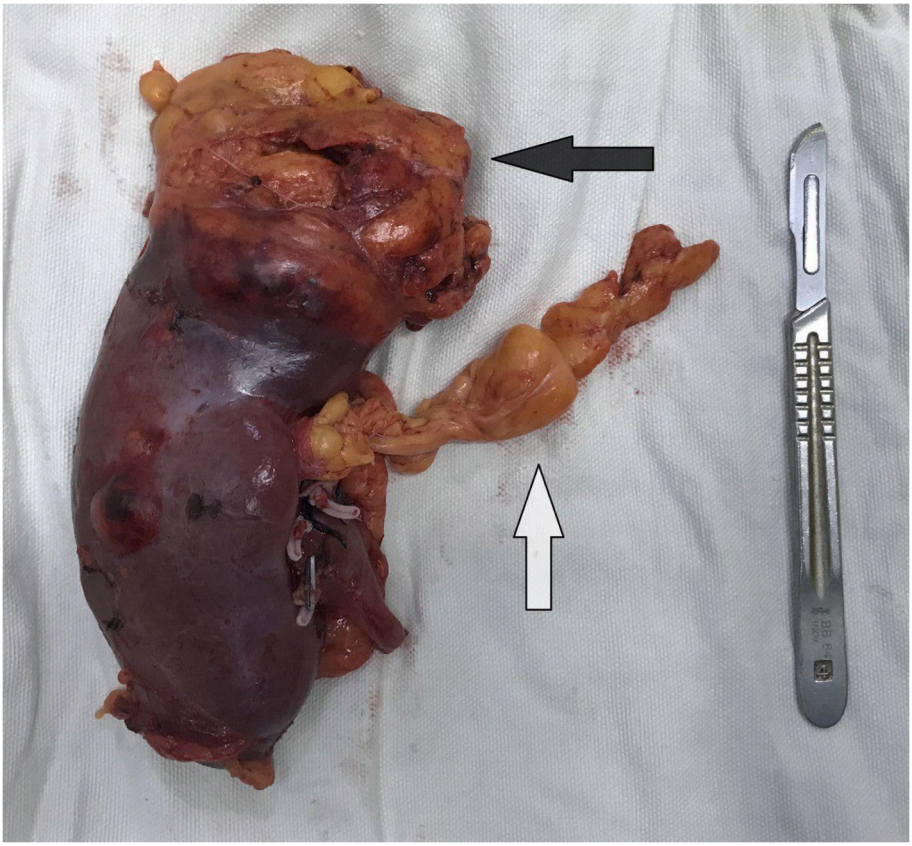

Fig. 2. The tumor which was removed from the patient's kidney.

Conclusion

Renal liposarcoma with IVC tumor involvement is extremely rare. The optimal management is radical nephrectomy and IVC tumor resection. Chemotherapy and radiotherapy have not shown significant survival benefit.

Conflicts of interest

None.

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