

Renal leiomyosarcoma with soft tissue metastasis: An unusual presentation

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Abstract

Primary renal sarcomas are exceptionally rare, constituting 1% of all malignant renal tumours. Since the prognosis for a renal sarcoma is particularly poor, differentiation from sarcomatoid renal cell carcinoma (RCC) is necessary. Histopathology and immunohistochemistry are the only modes of diagnosing these sarcomas as they have no specific features clinically and radiologically. We report a case of renal leiomyosarcoma which was clinically diagnosed as RCC. Histopathology revealed a spindle cell sarcoma and immunohistochemistry (IHC) confirmed a renal leiomyosarcoma. She also had a liver nodule and mass in the right side of neck and left arm. Biopsy from these sites revealed a similar morphology and IHC confirmed leiomyosarcoma. We report a case illustrating the rarity of this disease and its aggressive nature owing to fatal metastatic potential.

Introduction

Primary sarcomas constitute 0.8% to 2.7% of renal tumours in adults.^{1,2} Of these, leiomyosarcoma is the most common type (50%–60%), followed by liposarcoma, haemangiopericytoma, fibrosarcomas, malignant fibrous histiocytomas, and rhabdomyosarcoma.^{1,2} The clinical presentation mimics that of renal cell carcinoma (RCC) (i.e., flank pain, hematuria, and abdominal mass). These are highly aggressive neoplasms metastasizing commonly to lung, liver, and bone.³ To the best of our knowledge, this is the first report of soft tissue metastasis from primary renal leiomyosarcoma.

Case report

A 39-year-old female presented with a 4-year history of lump in the right lumbar region. The lump was gradually increas-

ing in size and was associated with dull aching pain in the abdomen. She also complained of lump in the right cervical region and left arm. An ultrasound of the abdomen revealed a large heterogeneous mass in the right lumbar region of the renal fossa measuring 10 × 7 cm. Magnetic resonance imaging revealed the presence of a large well-defined soft tissue mass from the upper pole of right kidney with evidence of small intraluminal component in the inferior vena cava and renal vein (Fig. 1, part A). Liver revealed multiple altered signal intensity lesions of variable size. A clinical diagnosis of RCC with metastases to the liver and soft tissues was made and the patient underwent right radical nephrectomy along with liver biopsy and excision of soft tissue masses. Grossly, the renal tumour was large, globular and encapsulated, measuring 11 × 7 × 6 cm at the upper pole of right kidney (Fig. 1, part B). Cut surface was grey white, firm, with a whorled appearance and areas of hemorrhage and necrosis. Soft tissue masses of arm and neck measured 5 × 3 × 3 cm and 3 × 2 × 2 cm, respectively.

The examined microsections showed a tumour composed of spindle cells arranged in interlacing fascicles with moderate nuclear pleomorphism, multinucleation, and focal areas of necrosis. Mitotic figures were 6-7/10 hpf. On extensive sampling, no definite epithelial component could be identified (Fig. 2).

On immunohistochemistry, the tumour cells were positive for vimentin, smooth muscle actin, and focally for desmin and negative for CK, RCC and CD10 (Fig. 3).

Therefore, a diagnosis of renal leiomyosarcoma was rendered. Sections from the renal pelvis, ureter, and adrenal were unremarkable. Sections from the liver nodule, right cervical, and left arm mass showed large foci of metastatic sarcoma (Fig. 4). Patient had an uneventful postoperative period and was given an option for chemotherapy which she refused. She was discharged in a stable condition after 10 days. The patient was well at the last follow-up 2 months after the procedure.



Fig. 1. A: Magnetic resonance image showing soft tissue mass at the upper pole of right kidney. B: Gross image showing a large, encapsulated tumour involving the upper and mid pole.

Discussion

Leiomyosarcomas are malignant neoplasms of smooth muscle origin. They are most commonly found in the uterus, stomach, small intestine, and retroperitoneum.⁴ Leiomyosarcomas of renal origin are very rare and constitute only 0.12% of all malignant renal neoplasms.⁵ First reported by Berry in 1919, these tumours are thought to originate from the renal capsule or the smooth muscle fibres in the renal pelvis or from the renal vessels.^{5,6} They most commonly affect the 50-to-60 age group with no gender predilection.⁷

These neoplasms present with the clinical triad of pain, hematuria, and abdominal mass mimicking the presentation of RCC. Computed tomography scan can sometimes be helpful in suggesting the diagnosis of these neoplasms and angiography would be helpful to confirm the diagnosis. If a well-differentiated neoplasm originates from the renal capsule or renal sinus and the tumour is hypovascular or avascular on angiograms, the diagnosis of renal sarcoma should be considered.⁸ The gross appearance of a white fibrous tumour with ill-defined edges and areas of degen-

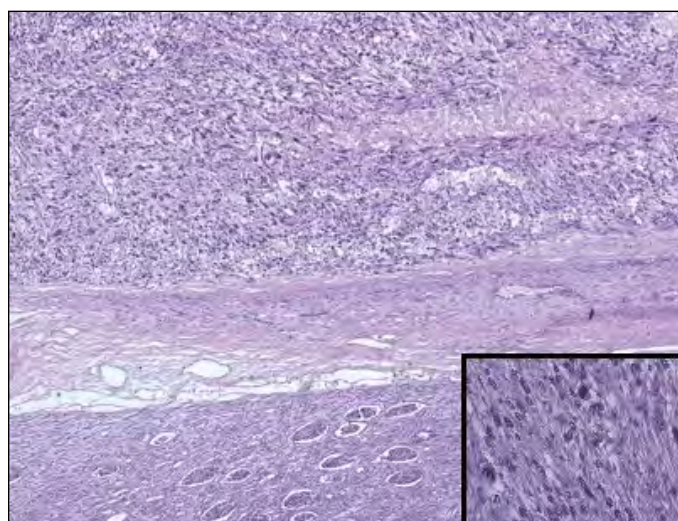


Fig. 2. Photomicrograph showing interlacing bundles of tumour cells; inset shows nuclear pleomorphism and mitoses (hematoxylin and eosin stain 20 \times).

eration also mimics RCC. Eventually, the histopathological examination and IHC help confirm the final diagnosis. Alternating fascicles of spindle cells with marked atypia and mitosis are part of the usual morphological picture shared by both leiomyosarcoma and sarcomatoid RCC.^{5,9} The absence of epithelial elements on extensive sampling and IHC clearly ruled out sarcomatoid RCC in the present case. Bulky tumour instead of small intraparenchymal lesions ruled out the possibility of metastasis.¹⁰

Renal leiomyosarcomas have an aggressive behaviour owing to rapid growth rate, frequent metastases, and high local and systemic recurrence rates.¹¹ One of the most common sites for metastasis is the liver as was seen in our case. Other reported sites were lung, bone, and stomach, but soft tissue metastasis has not been reported to date. Our patient presented with a renal mass with metastatic deposits in the liver and soft tissue, thus throwing light on the disruptive behaviour of these rare neoplasms. Despite the finding that soft tissue comprises about 55% of our body mass, hematogenous metastases to these areas are rare. Direct extension of a primary tumour to soft tissue occurs more often than distant soft tissue metastases. Several factors have been impli-

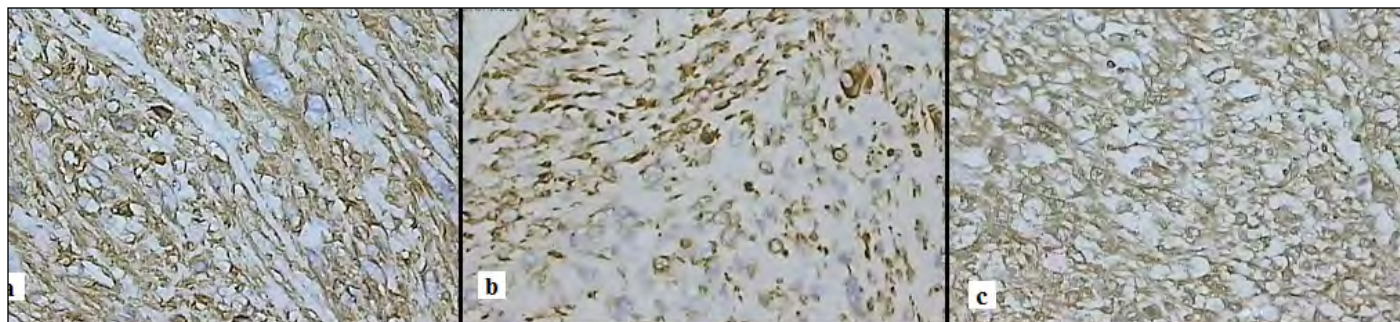


Fig. 3. Immunohistochemistry: SMA (a), desmin (b) and vimentin (c).

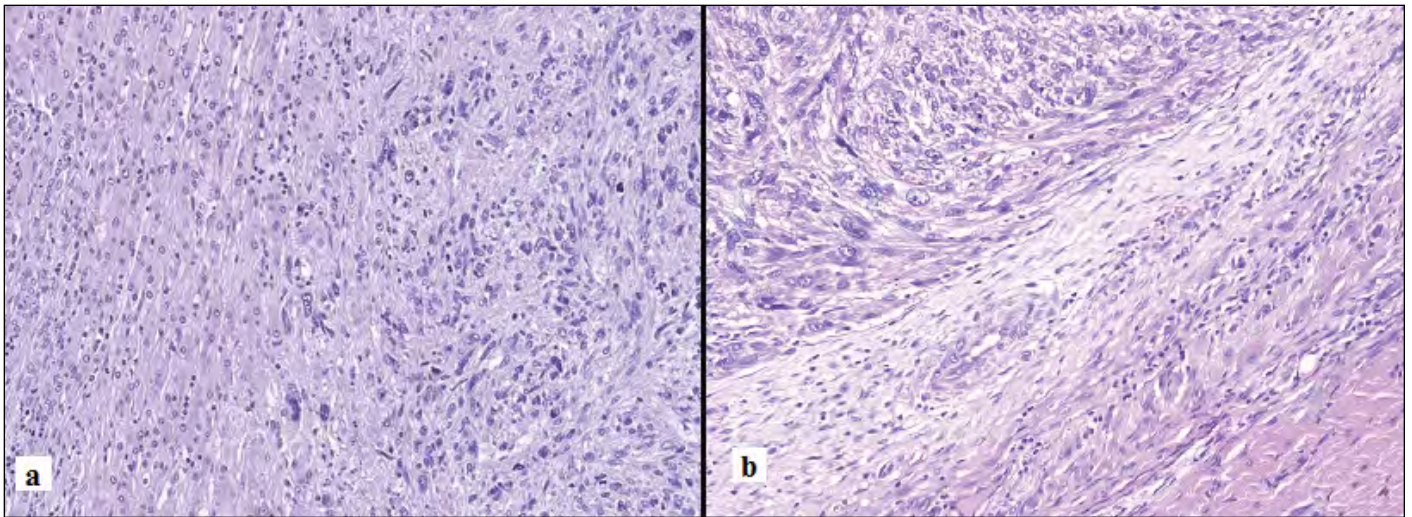


Fig. 4. A: Metastatic nodule in liver. B: Metastatic nodule in arm with adjacent skeletal muscle (hematoxylin and eosin, 100 \times).

cated in the rarity of this phenomenon, such as changes in pH, accumulation of metabolites, and the local temperature at soft tissue sites.¹² The most commonly reported malignancies in the literature to result in distant soft tissue metastases are lung, kidney, and colon carcinoma.¹³ Metastases to distant soft tissue sites from a primary soft tissue sarcoma are very rare. In a study done by Plaza and colleagues, they accounted for <10% of all cases of metastases to soft tissue. This figure may be underestimated due to the inherent difficulties in the histopathologic diagnosis of sarcoma, or due to difficulties in distinguishing clinically between metastases of sarcoma and multifocal disease.¹³

Management of high-grade sarcoma involves a multidisciplinary approach, including surgical resection, radiotherapy, and adjuvant chemotherapy. The role of adjuvant chemotherapy and/or radiotherapy is still disputed due to lack of data on the treatment of this rare renal neoplasm and variability of results from limited experience in the literature.^{1,5} The favourable prognostic factors are tumour size <4 cm, low grade, absence of nodal metastases, and radical surgical treatment. The 5-year survival rate is 29% to 36%, with most patients dying of disease within a year of diagnosis.⁸

Conclusion

Leiomyosarcoma is a rare tumour of the kidney. Although it has a propensity to metastasize widely, soft tissue metastasis is unknown. To the best of our knowledge, this is the first case of primary renal leiomyosarcoma presenting with soft tissue metastasis.

Competing interests: Authors declare no competing financial or personal interests.

This paper has been peer-reviewed.

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