

# Renal dedifferentiated liposarcoma with intra-caval tumor thrombus: A rare case

Charu Shastri, Jatinder Kumar<sup>1</sup>, Sushila Jaiswal, Anil Mandhani<sup>1</sup>

Departments of Pathology, <sup>1</sup>Urology and Renal Transplantation, SGPGI, Lucknow, Uttar Pradesh, India

## ABSTRACT

Combined penoscrotal incarceration is rarely reported in the literature. It is a urological emergency. Depending on the nature of the constricting object, duration of constriction, and with no defined treatment methods, inventiveness is usually required in removing difficult objects. This is a report of a penoscrotal incarceration by four closed steel rings, treated in Northern Ireland with a few minor points reiterated.

**Key words:** Dedifferentiation, IVC thrombus, liposarcoma

## INTRODUCTION

Sarcomas represent 1% to 2% of all malignant renal tumors in adults, with leiomyosarcomas being the most common and liposarcoma the second most common type.<sup>[1]</sup> Peak incidence is in the fifth decade of life.<sup>[1]</sup> Renal sarcoma is less common but more lethal than sarcoma of any other genitourinary sites, with a reported 5-yr survival rate of 29%.<sup>[2,3]</sup>

Renal liposarcoma is a rare neoplasm, with only few well-documented case reports in the literature. Like elsewhere, renal liposarcomas can exhibit various histological subtypes: well differentiated, spindle cell, pleomorphic, or dedifferentiated histology. Presentation with a vena caval tumor thrombus has not been described in a renal liposarcoma. Herein, we report a rare case of renal dedifferentiated liposarcoma with inferior vena caval tumor thrombus.

## CASE REPORT

A 65-yr-old man was referred to our hospital for

**For correspondence:** Anil Mandhani Department of Urology and Renal Transplantation Sanjay Gandhi Post Graduate Institute of Medical Sciences (SGPGIMS) Raebareli Road, Lucknow - 226014 (UP), India

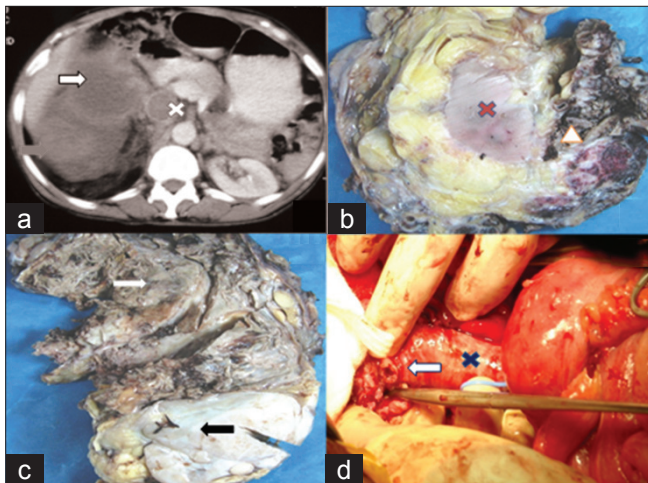
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complaints of abdominal lump and right flank pain for 4 months with no hematuria and lower urinary tract symptoms. He had a hard nodular mass, in right hypochondrium, epigastrium, and lumbar region. Imaging studies, i.e. ultrasound with Doppler study computed tomography (CECT) and MRI, showed an upper and mid polar renal mass measuring 20×16×10 cm, extending from the sub-diaphragmatic area to bifurcation of aorta with retroperitoneal lymphadenopathy and IVC thrombus, with some doubtful fat density [Figure 1a]. Intra caval thrombus was seen extending from intra hepatic portion of IVC to the right common iliac vein. A clinical diagnosis of renal cell carcinoma was made. Tumor thrombus extended from just inferior to liver till bifurcation of aorta. Thrombus was adherent to the IVC wall at renal vein and IVC confluence, therefore around 2 cm patch of IVC was resected and repair done [Figure 1d].

On examination, cut surface of nephrectomy specimens showed a large heterogeneous growth comprising of a distinct large necrotic tan brown area along with smaller fleshy yellowish area, together measuring 20×14×9 cm [Figure 1c] infiltrating the renal sinus and encasing the rest of kidney completely [Figure 1b]. Grossly normal looking kidney measured 11×6 cm and was easily shelled out from tumor except at the renal sinus, suggesting a possible origin from renal sinus.

Microscopy from the necrotic tan brown area revealed a malignant mesenchymal tumor, predominantly malignant fibrous histiocytoma (MFH) like high grade sarcoma with brisk mitosis (20-25/10 HPF) [Figure 2a]. Tumor in the white fleshy area showed a well-differentiated liposarcoma showing lipoblasts lying in myxoid and focally sclerosed background, well demarcated from the dedifferentiated area



**Figure 1:** (a) Pre-op CT scan showing large heterogeneous mass in renal area with large hypo-intense non-fatty dedifferentiated area (white arrow) and smaller hyper-intense fatty area (red arrow) along with IVC thrombus with largely non-fatty area (cross). (c) Mass comprised of large necrotic tan brown area (white arrow) with smaller fleshy white area (black arrow). (b) Kidney (marked by cross) was encased by the tumor and could be easily shelled out from mass except at renal sinus (arrow head). (d) Intra-operative removal of tumor thrombus (cross) at renal vein outlet (arrow)

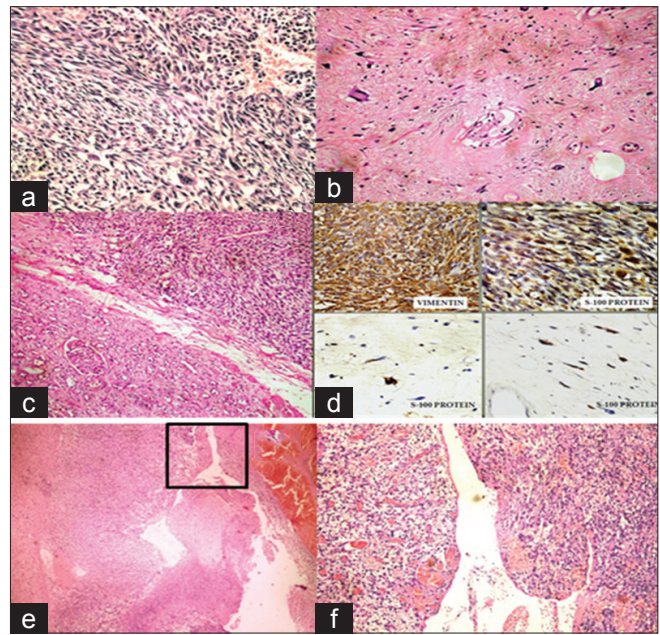
[Figure 2 b]. The tumor did not infiltrate kidney in any of the planes except renal sinus [Figure 2 c].

The sections from the IVC thrombus showed dedifferentiated areas of tumor only [Figures 2e, f]. The resection margins of ureter and renal artery were free from invasion. Diagnosis of dedifferentiated liposarcoma was made. Areas of epithelial differentiation or angiomyolipomatous morphology were not seen on multiple sectioning.

Immunohistochemistry was performed, and tumor cells were strongly positive for vimentin and S-100, and negative for pancytokeratin, epithelial membrane antigen (EMA), desmin, and HMB-45 [Figure 2d]. Post operatively, the patient was managed with doxorubicin and ifsofamide chemotherapy and patient had an uneventful 9 month follow-up.

## DISCUSSION

Liposarcoma arising in extremities or retroperitoneum affects middle-aged and old patients, and tend to follow a relatively indolent clinical course with local recurrences after resection and occasional distant metastasis, mainly to the lungs.<sup>[4]</sup> Liposarcoma of kidney is a rare tumor with only few well-documented reports in the literature, many of these being associated with tuberous sclerosis and probably correspond to angiomyolipomas (AML). It could be difficult to establish the exact origin of the tumor, which can arise from sinus fat, or from the renal capsule.<sup>[3]</sup> However, the presence of renal vein thrombus with a vena caval infiltration is a finding that strongly suggests renal origin of the sarcoma.



**Figure 2:** a) Sarcoma with storiform and fascicles, marked atypia, mitosis, and giant cells. (b) Fleshy white area with atypical spindle stromal cells, tumor giant cells and typical lipoblasts having central nucleus with indentation and abundant vacuolated to clear cytoplasm (Hand E  $\times 400$ ). (c) Dedifferentiated area with well separated normal renal parenchyma (Hand E  $\times 200$ ). (d) IHC: strong vimentin and S100 positivity in dedifferentiated and well-differentiated areas. Lipoblasts are strongly S100 positive ( $\times 400$ ). (e) Low and medium. (f) Power view of IVC wall with adherent thrombus and invasion into wall

Dedifferentiation is defined as a malignant transition from atypical lipomatous tumor/well-differentiated liposarcoma to non-lipogenic sarcoma of variable histological grade.<sup>[4]</sup> Most of the cases reported showed hypo or iso-intense non-fatty mass along with a fatty area typical of liposarcoma,<sup>[5]</sup> as in our case. It's a rather infrequent phenomenon, which can occur in up to 10% of all well-differentiated liposarcomas although the risk is higher in deep-seated locations. Approximately, 90% of cases occur de novo, whereas 10% develop in recurrences. Dedifferentiated liposarcoma, with its high-grade histology exhibits an aggressive clinical course compared to well-differentiated liposarcoma and is associated with an 83% local recurrence rate, 30% distant recurrence rate and 6 fold increased risk of death as compared to well differentiated histology.<sup>[6]</sup>

An important differential diagnosis is AML because both are large fat-containing lesions.<sup>[3]</sup> Three major imaging findings helping in AML diagnosis (defect in the renal parenchyma, enlarged vessels in the lesion, along with finding of typical areas of AML) and HMB 45 positivity are crucial for making the distinction. Liposarcomas arise in perirenal fat within Gerota's fascia or capsule and they displace, compress, and distort the kidney but usually do not invade the adjacent renal parenchyma. Therefore, liposarcomas do not cause a defect in the renal parenchyma, and interface of tumor with kidney is smooth unlike AML, as in our case. These findings are difficult to appreciate on radiology.

The most important prognostic factors for survival are histological grade and completeness of resection.<sup>[6,7]</sup> Visceral liposarcomas have a poorer prognosis than extremity/trunk lesions, and this has been attributed to frequent presentation with tumors of large size and multiorgan involvement that precludes complete resection. Approximately, 40% of dedifferentiated liposarcomas will recur locally, 17% will metastasize, and 28% of the patients will ultimately die as a result of tumor.<sup>[4]</sup> The standard treatment has been radical nephrectomy, with or without chemo-radiotherapy.<sup>[8]</sup> In our case, chemotherapy was given with six cycles of Doxorubicine and Ifosfamide postoperatively. At follow up of 9 months there was no evidence of recurrence.

In conclusion, the present case is a first reported case of renal liposarcoma with tumor thrombus in IVC that was successfully managed with nephrectomy and post-operative chemotherapy with uneventful follow up.

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