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Case Report

Radiology Section

A Rare Case of Huge Renal Leiomyosarcoma and Imaging Diagnostic Dilemma

AMOL MADANLAL LAHOTI, GURNIHAL SINGH CHAWLA, AVINASH DHOK, JITESH RAWAT, KISHOR NEMADE

ABSTRACT

Soft tissue sarcomas are rare mesenchymal tumours. Leiomyosarcoma is a rare malignant tumour of smooth muscle origin that generally arises from soft tissues and uterine tissue. On occasions, they may arise from smooth muscle of the vessel walls, more commonly from the veins. Renal leiomyosarcomas frequently originates from the smooth muscles of renal capsule, renal pelvis or renal vessels. Correct diagnosis and appropriate management is very essential in such cases.

We highlight the possibility of malignant soft tissue sarcoma like leiomyosarcoma in the kidney. We emphasize on the suspicion of rare possibility and clinicopathological correlation is required to identify such cases, which has pronounced prognostic implications as whether to do surgical resection or biopsy or treat such patient with chemotherapy or radiotherapy. Decision varies with diagnosis.

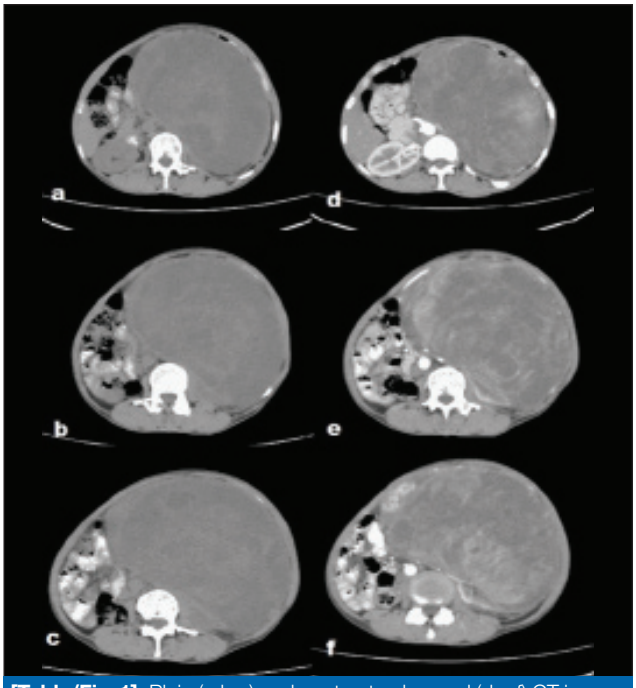
Keywords: Mesenchymal tumour, Neuroectodermal tumour, Sarcomas

CASE REPORT

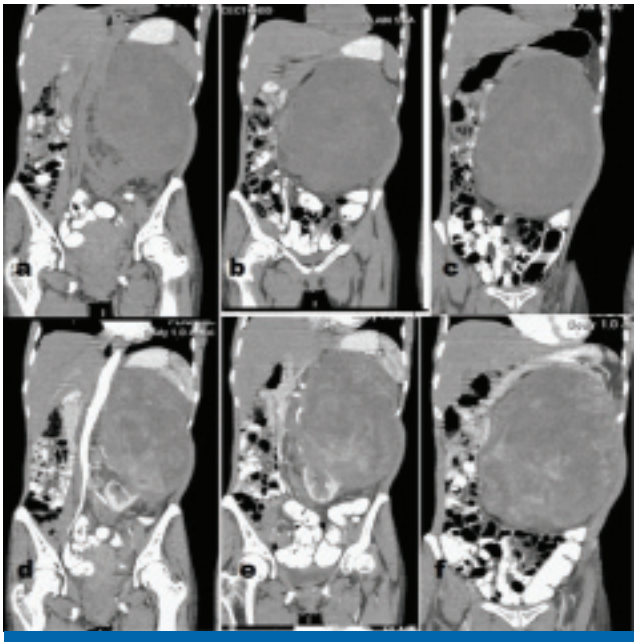
We present the case of 50-year-old woman who presented with chief complaints of progressively increasing lump in abdomen since eight months that has increased significantly in size since last two months, associated with pain in abdomen radiating to back since last five days, loss of appetite and weight since last two months.

On clinical examination, there was a large lump in abdomen almost occupying whole of the abdomen, more of the left side. No abnormalities were observed in routine laboratory examination, except an iron deficiency anemia (hemoglobin 9 g/dL).

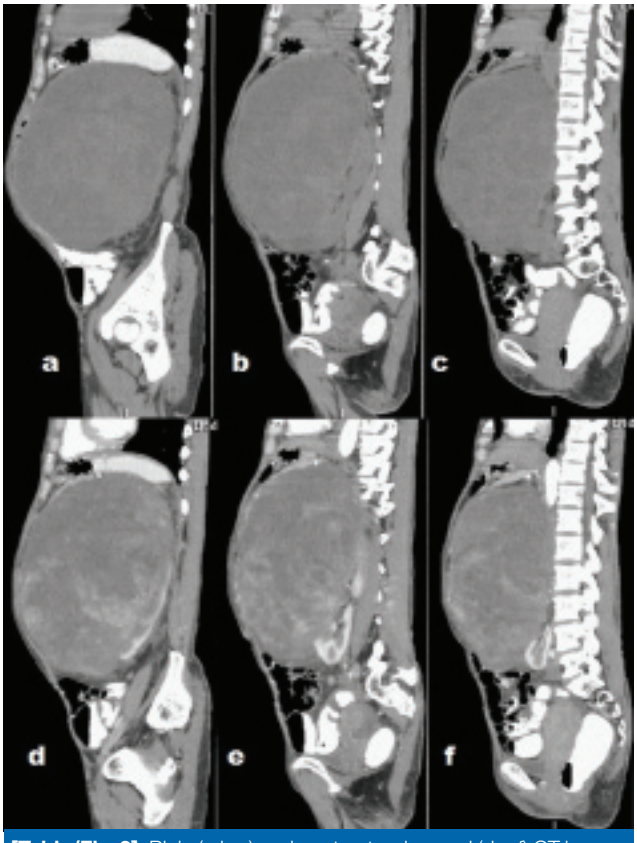
Ultrasonography demonstrated a large heterogenous predominantly hypoechoic mass lesion in the region of left supra renal region and adrenal could not be visualized separately likely suggestive of Renal Cell Carcinoma (RCC) or adrenal tumour. Multiple calcific foci are noted within this lesion on plain Computed Tomography (CT) scan. On contrast enhanced CT-scan imaging demonstrated huge mass of size16.4 x 19 x 22.6 cm (in AP transverse and CC dimensions respectively) with approximate volume of 3000 cc arising from left kidney and showed significant heterogeneous enhancement. Moderate adjacent fat stranding is seen. Medially, the mass was displacing the abdominal aorta, inferior vena cava small bowel loops and head of the pancreas on the right side [Table/Fig-1-3]. However, there is no evidence of thrombus or extension of the mass lesion in renal vein or inferior vena cava or adjacent structures. Adrenal gland was well visualized separately from the lesion. Patient did not



[Table/Fig-1]: Plain (a,b,c) and contrast enhanced (d,e,f) CT image, axial section shows a large heterogeneously enhancing lesion arising from the left kidney and mass effect secondary to large leiomyosarcoma without extension to adjacent organ.



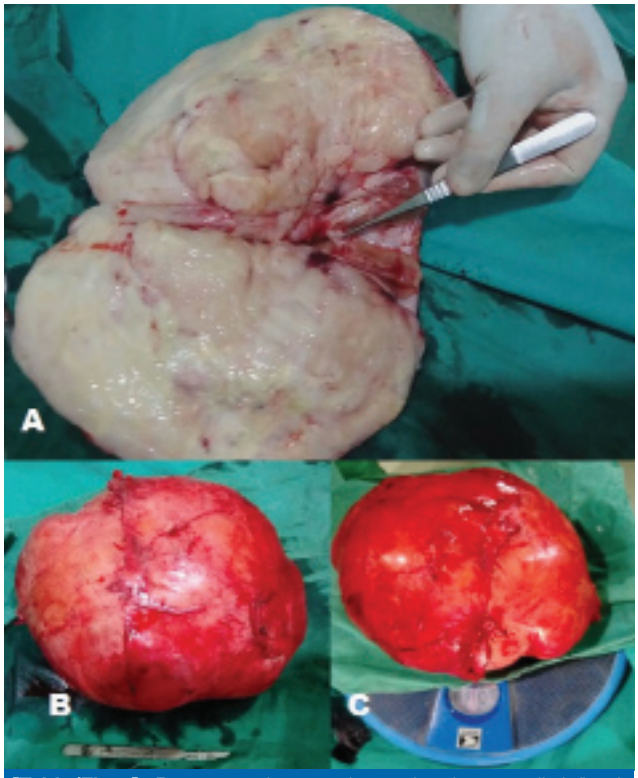
[Table/Fig-2]: Plain (a,b,c) and contrast-enhanced (d,e,f) CT image, coronal section shows a large lesion arising from the left kidney displacing the abdominal aorta, inferior vena cava small bowel loops and head of the pancreas on the right side.



[Table/Fig-3]: Plain (a,b,c) and contrast-enhanced (d,e,f) CT image, sagittal section shows a large lesion arising from the left kidney and mass effect.

complain of hematuria or fever (suggestive of RCC) or signs of raised or decreased blood pressure or primary elsewhere (suggestive of primary or metastatic adrenal tumour).

Open left radical nephrectomy by lumbotomy was performed. The surgical specimen revealed a large solid mass weighting of 3.5 kg [Table/Fig-4] which probably originates in the renal capsule. The excised specimen was sent for histopathogical examination through which the diagnosis of high grade renal leiomyosarcoma was confirmed. The tumour was infiltrating the renal capsule, however renal vein or adrenal gland, adjacent organs and pelvis were not involved. The hilar lymphnodes were excised and were also not involved. The immunohistochemical profile with smooth muscle actin was faintly positive. Focal immunostaining was demonstrated for vimentin, desmin and CD117 (KIT). No immunostaining was found for melan A, S-100 and HMB-45 protein.



[Table/Fig-4]: Post operation specimen showing a well-defined, smooth marginated mass, weighting 3.5 kg. On histopathological examination, leiomyosarcoma arising from kidney.

DISCUSSION

Soft tissue sarcomas are rare mesenchymal tumours. They have an incidence of 50/10, 00,000 [1]. The term leiomyomas as first described by Virchow in 1854 as rare mesenchymal tissue tumours of benign etiology originating from smooth muscle cells [2]. Leiomyomas and leiomyosarcomas may involve any organ of the genitourinary tract of which uterus is the most common site followed by the kidneys [3].

Leiomyosarcoma accounts for 50%-60% of all renal sarcomas and is of most common type amongst renal sarcomas [4]. It usually arises from the renal capsule, the renal parenchyma, smooth muscle fibers of the renal pelvis, or the main renal vein [4]. Both men and women are equally affected by the disease occurring around 40 and 70 years of age. Patients mainly come with complaints of flank pain, an abdominal mass, and hematuria [4,5]. Grossly, renal leiomyosarcoma is a large well-circumscribed encapsulated tumour which is soft-to-firm in consistency as compared to renal cell carcinomas which has irregular and poorly defined margins invading the adjacent structures and vessels. On histological examination, the tumour consists of spindle cells interspersed with a variable amount of connective tissues arranged in a plexiform, fascicular, or haphazard growth pattern.The presence of nuclear pleomorphism, mitosis, density, and necrosis differentiates leiomyosarcoma from a benign leiomyoma [4]. Primary sarcomas constitute from 0.8 to 2.7% of total renal tumours in adults [6].These tumours usually have an insidious presentation, with symptoms and signs occurring at late stages of the disease, like nonspecific abdominal pain, palpable mass, vomiting and hematuria and weight loss. Neither ultrasonography, CT-scan nor MRI can accurately differentiate between leiomyosarcomas and RCCs [7]. The major prognostic factor is total surgical resection [2], when it is completed, 5 years' disease free survival could be of 60%.Radical nephrectomy is the treatment of choice, which should be followed by postoperative radiation and chemotherapy [4]. In cases of small lesions (less than 4 cm), it is possible to opt for conservative surgery. A further option for smaller lesions is renal biopsy.

CONCLUSION

This case highlights the rarity of the disease and depicts the tumour's potential for assuming extreme proportions and causing pronounced displacement of adjacent major structures. In terms of size, it is one of the largest leiomyosarcoma of renal origin ever reported. It also concludes that clinical details and imaging alone cannot differentiate between leiomyosarcoma and RCC and only histopathology can provide the definitive diagnosis. Because the presenting symptoms and imaging results do not provide a sufficient ground for accurate and timely diagnosis, a high index of suspicion should always be maintained. Biopsy plays an indispensable role in diagnosis. Suspicious cases should be biopsied as early diagnosis implies excellent prognosis and increased patient survival. A high index of suspicion is necessary to arrive at the correct diagnosis.

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AUTHOR(S):
1. Dr. Amol Madanlal Lahoti
2. Dr. Gurnihal Singh Chawla
3. Dr. Avinash Dhok
4. Dr. Jitesh Rawat
5. Dr. Kishor Nemade

PARTICULARS OF CONTRIBUTORS:
1. Junior Resident, Department of Radiology, NKP SIMS and LMH, Nagpur, Maharashtra, India.
2. Senior Resident, Department of Radiology, NKP SIMS and LMH, Nagpur, Maharashtra, India.
3. Professor and Head, Department of Radiology, NKP SIMS and LMH, Nagpur, Maharashtra, India.
4. Senior Resident, Department of Radiology, NKP SIMS and LMH, Nagpur, Maharashtra, India.
5. Junior Resident, Department of Radiology, NKP SIMS and LMH, Nagpur, Maharashtra, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Gurnihal Singh Chawla,
F4/8, Vindhyachal Hostel, NKP Salve Institute of Medical Sciences and LMH, Nagpur-440019, Maharashtra, India.
E-mail: gurnihal.chawla1@gmail.com

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