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RENAL HEMANGIOPERICYTOMA : A RARE CASE REPORT WITH REVIEW OF LITERATURE

Radio-Diagnosis

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ABSTRACT

Objectives : Description and illustration of clinical and imaging findings of renal hemangiopericytoma. **Background :** A 56 year old female presented with left flank pain since 6 months. The patient was subjected to NECT and contrast enhanced CT following standard renal protocol and relevant MRI sequences were taken.

Imaging Findings:

- NECT revealed a relatively well circumscribed isodense mass lesion in middle pole of the left kidney with a small foci of calcification.
- On CECT, in the cortico medullary phase, there was intense homogeneous enhancement of the mass lesion, with few non enhancing areas and in the nephrographic phase, significant washout of the contrast compared to corticomedullary phase.
- On MRI , The lesion was isointense to the renal cortex on T1 and On T2, alternate areas of extremely low signal intensity and moderately high signal intensity were seen. There was no evidence of macroscopic fat seen with chemical shift imaging. On DWI , the lesion demonstrated diffusion restriction.
- The patient underwent left radical nephrectomy .The specimen was sent for histopathological and IHC examination , which revealed findings of renal hemangiopericytoma.
- **Final Diagnosis :** Renal hemangiopericytoma.

Conclusion :

- Solitary fibrous tumor (previously characterized as hemangiopericytoma) is a rare spindle cell neoplasm that is most often found in the pleura. To date, only 40 cases of solitary fibrous tumor of the kidney have been reported in the literature.
- Awareness of the various mesenchymal renal tumors and familiarity with their imaging findings permit optimal patient management.

KEYWORDS

Renal hemangiopericytoma, Solitary fibrous tumour, Renal cell carcinoma

INTRODUCTION:

- Solitary Fibrous Tumours (SFT) , previously calssified as hemangiopericytoma (HPC) can manifest anywhere in the body, but they are extremely rare in the kidney
- In clinical practice, it is easy to misdiagnose rSFT/HPCs as other tumors arising in the kidney, such as renal cell carcinoma (RCC), or as retroperitoneal tumors .
- Therefore, accurate pre-operative identification of rSFT/HPCs is important so clinicians can avoid extensive surgery, especially radical nephrectomy and regional lymph node dissection.
- However, the accurate pre-operative characterization of indeterminate rSFT/HPCs remains a challenge.

Background :

- A 56 year old female presented to the surgery out patient department with left flank pain since 6 months .The pain was dull aching and gradually progressive in nature.
- No history of fever /hematuria/burning micturition/loss of appetite or loss of weight .
- On physical examination , there was mild tenderness in the left lumbar region .
- The laboratory results, including complete blood counts, renal function parameters, serum electrolytes , and urinalysis were within normal range.

Imaging Findings:

- NECT revealed , a relatively well circumscribed isodense mass lesion with mean attenuation values of +35HU to +40HU, approximately measuring 3.6x3.5x4.1cm (APxTDxCC) involving the cortex and medulla of middle pole of the left kidney with a small foci of calcification (Fig 1).
- In the cortico medullary phase , there was intense homogeneous enhancement of the mass lesion (the attenuation value of +210HU-+230HU), with few non enhancing areas(Fig 2a) and in the nephrographic phase(Fig 2b) , there was significant washout of the contrast(attenuation values of +175HU to+180HU) compared to corticomedullary phase.
- The mass lesion appeared to partially obliterate the renal sinus and displace the renal pelvis anteriorly . No dilatation of the pelvis / collecting system was seen (Fig 4).
- Mild extra capsular extension of the lesion was seen posteromedially , abutting the psoas and the quadratus lumborum muscle.
- The lesion appeared to abut the left main renal vein and the segmental branch of renal artery. However there was no involvement of the renal artery / renal vein / IVC by the lesion(Fig 3).
- Normal contrast opacification of the renal pelvis and ureter was seen in the excretory phase(Fig4).
- There was no retroperitoneal lymphadenopathy.

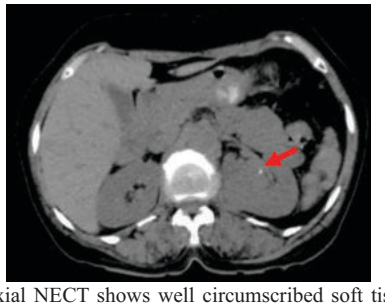

Figure1: Axial NECT shows well circumscribed soft tissue density mass lesion with foci of calcification (arrow)

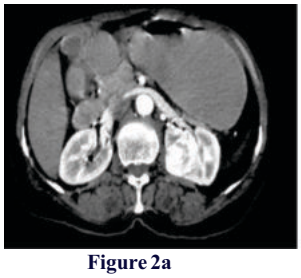

Figure 2a

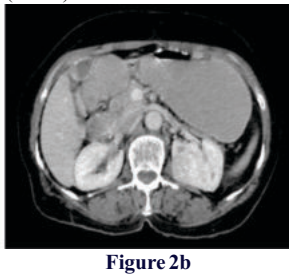

Figure 2b

Figure 2a&2b: Axial CECT image in the cortico-medullary phase(Fig 2a) , shows intense homogeneous enhancement of the lesion with few non-enhancing areas. In nephrographic phase (Fig2b),demonstrates significant washout of the contrast

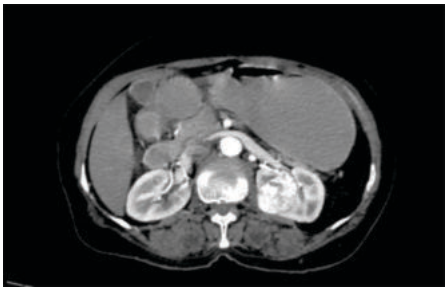

Figure 3 axial CECT image in corticomedullary phase : shows normal opacification of the left renal artery and vein .

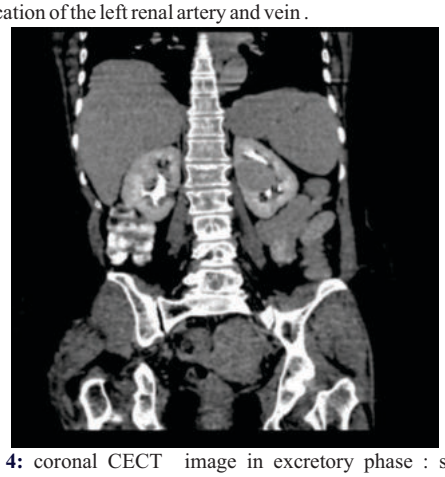

Figure 4: coronal CECT image in excretory phase : shows no dilatation of the pelvis / collecting system and normal opacification of the pelvis

- On MRI , The lesion was isointense to the renal cortex on T1-weighted images.
- On T2- weighted MR images(Fig 3), alternate areas of extremely low signal intensity and moderately high signal intensity were seen.
- There was no evidence of macroscopic fat seen with chemical shift imaging(Fig 4a&4b).
- On DWI , the lesion demonstrated diffusion restriction(Fig 5) .

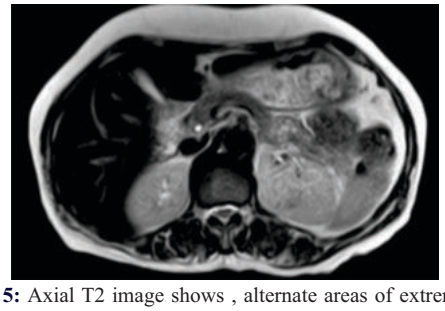

Figure 5: Axial T2 image shows , alternate areas of extremely low signal intensity and moderately high signal intensity within the lesion.

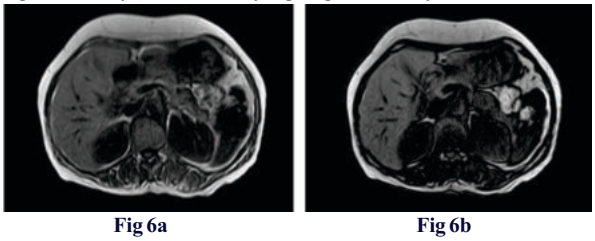

Fig 6a **Fig 6b**

Figure 6 a&b:Axial T1 In-phase GRE (Fig6a) and Out-phase(Fig 6b) shows no macroscopic fat within the lesion.

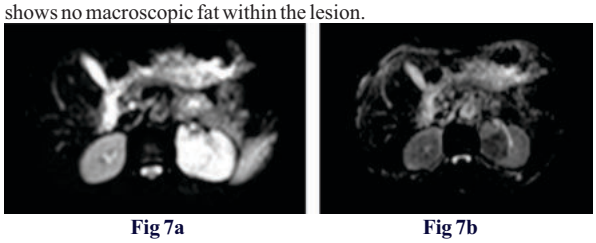

Fig 7a **Fig 7b**

Figure 7 a& b: DWI shows mild diffusion restrictionin the lesion (Fig 7a),with low ADC Values(Fig 7b)

Final Diagnosis: Renal hemangiopericytoma.

Differential Diagnosis : Renal cell carcinoma. RCC often shows an indistinct interface with the surrounding parenchyma and frequently has a lobulated or irregular outer margin. On contrast enhanced CT/MRI , RCC shows s enhancement on cortico medullary phase with no significant washout in the nephrographic / excretory phase.

Follow up : The patient underwent left radical nephrectomy .The specimen was sent for histopathological examination , which revealed renal tissue with well defined mesenchymal tumour and plenty of variable ectatic and copressed , thin walled branching vessels with gaping sinusoidal vessels ,suggestive of renal hemangiopericytoma. The findings were further confirmed on IHC where, the tumour cells were positive for CD34,CD99 and bcl2.

DISCUSSION:

Solitary fibrous tumor is a rare spindle cell neoplasm that is most often found in the pleura. Extrapleural solitary fibrous tumors are being increasingly diagnosed, including some located in the kidney. To date, only 40 cases of solitary fibrous tumor of the kidney have been reported in the literature[1].

Most tumors previously characterized as hemangiopericytomas have now been reclassified as solitary fibrous tumors. Within the kidney, solitary fibrous tumors may arise from the renal capsule, cortex, pelvis, or peripelvic connective tissue. Most patients are older than 40 years, with a slight female predilection. Clinical features include a palpable abdominal mass, flank pain, and hematuria[1].

The cross-sectional imaging findings of renal solitary fibrous tumors are attributable to the combination of variable cellularity, dense collagen content, and focal hemangiopericytomatous vascular pattern. On CT images, renal solitary fibrous tumor appears as a well-circumscribed lobulated soft-tissue mass in the region of the renal sinus or capsule

The tumors typically show relatively homogeneous enhancement despite the large size of the mass. Necrosis, hemorrhage, and calcifications are rare. At MR imaging, solitary fibrous tumors are isointense to the renal cortex on T1-weighted images. On T2- weighted MR images, alternate areas of extremely low signal intensity and moderately high signal intensity are distributed in a radial configuration.

Surgical removal is the treatment of choice. Most solitary fibrous tumors are benign; however, 10%–15% of extrapleural solitary fibrous tumors show malignant behavior in the form of recurrence or metastatic disease.

Awareness of the various mesenchymal renal tumors and familiarity with their imaging findings permit optimal patient management[2]

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