Case Report

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Pathology Section

Renal Leiomyoma: An Uncommon Differential Diagnosis of Renal Masses with a Clinical Relevance

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ABSTRACT

Renal leiomyomas are an extremely rare neoplasm which account for 1.5% of the benign lesions and 0.29% of all treated renal tumours, with autopsy evidence of 4.2% to 5.2%. Given their exceptional incidence, they are rarely included in the differential diagnosis of renal masses, despite their capacity to grow to a considerable size. The definitive diagnosis of a leiomyoma is only possible after histopathological examination, most of the times of a nephrectomy specimen. However, due to the rising number of diagnosis resulting from improved medical imaging and interventions, renal leiomyomas should gain importance in the differential diagnosis of renal masses, specialy with respect to kidney sparing surgery. Hereby, we report a case of 60-year-old male patient with a large well circumscribed left renal mass, diagnosed as Renal cell carcinoma on imaging.

Keywords: Carcinoma, Leiomyoma, Leiomyosarcoma, Renal masses

CASE REPORT

A 60-year-old male patient, known case of hypertension and diabetes mellitus presented with a non specific complaint of flank pain. All the haematological and biochemical parameters were within normal limits. Contrast Enhanced Computed Tomography (CECT) whole abdomen revealed a large well circumscribed left renal mass, involving the upper and mid polar regions of kidney. Perinephric extension and extensive calcification were also seen. The appearance was suggestive of Renal Cell Carcinoma on imaging. The patient was admitted and a planned Radical nephrectomy was done keeping in mind a malignant neoplasm as evident on CECT. Radical nephrectomy's specimen was sent for histopathological evaluation. A mass of 14 X 8.5 X 4 cm was received; it was circumscribed and encapsulated with a well defined limit between the thin rim of renal tissue and the lesion. The tumour was very hard in consistency, with the cut surface showing solid whorled white appearance [Table/Fig-1].

On microscopy, fascicles of long spindle cells showing a whorling pattern along with extensive areas of calcification were seen. Nuclei of the spindle cells were regular, oval, having bland chromatin. No tumour cell pleomorphism or immature elements were evident in multiple sections [Table/Fig-2,3]. At the periphery of the section scattered tubular cells without cytological atypia were present.

An initial diagnosis of benign spindle cell lesion was considered. In view of the associated epithelial component (tubular cells) the possibility of Mixed epithelial and stromal tumour (Adult Mesoblastic Nephroma) was kept as a differential diagnosis. However, on immunohistochemistry Smooth Muscle Actin (SMA) was positive [Table/Fig-4] whereas, Cytokeratin and HMB-45 were negative [Table/Fig-5], excluding the diagnosis of Adult Mesoblastic Nephroma and Angiomyolipoma (a common differential). Thus, confirming the diagnosis of Renal Leiomyoma.

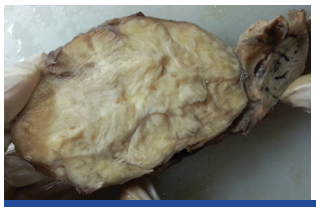
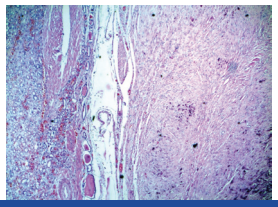
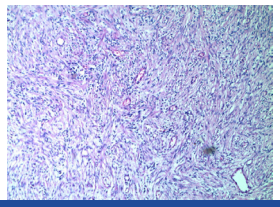
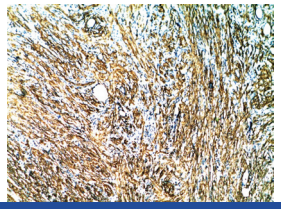
DISCUSSION

Leiomyomas are constituted under the Benign Mesenchymal neoplasms, with origin from the smooth muscle cells. In the genitourinary tract they are seen to involve any organ but are known to most commonly affect the kidneys, originating from smooth muscle cells of renal capsule, pelvis, calices and blood vessels [1,2].

The various other benign mesenchymal renal tumours are angiomyolipoma, hemangioma, lymphangioma, juxtaglomerular cell tumour, renomedullary interstitial cell tumour (medullary fibroma), lipoma, solitary fibrous tumour, and schwannoma. It may arise from smooth muscle cells of the renal capsule, the muscularis of the renal pelvis and cortical vascular smooth muscles. However, the renal capsule being the most common site [3].

Renal leiomyomas are uncommon neoplasms accounting for 1.5% of the benign lesions and 0.29% of all treated renal tumours [4]. The autopsy evidence has a much higher frequency than the clinical incidence [5]. The literature reviewed showed an autopsy evidence of 4.2% to 5.2% [6]. They are often detected incidentally. In a study conducted on nephrectomy specimens over a period of 10y, renal leiomyomas represented 1.5% of benign renal tumours and 0.3% of overall tumours treated [7].

There are no specific clinical symptoms (e.g., flank pain or flank tumour) or at times completely absent, making most of the leiomyomas an incidental finding during routine diagnostics [5].


[Table/Fig-1]: An encapsulated, circumscribed mass with a well defined limit between the thin rim of renal tissue and the lesion. The cut surface showing a solid whorled white appearance., **[Table/Fig-2]:** Microphotograph showing whorling pattern along with areas of calcification and at the periphery of the section scattered renal tubular cells (H&E,4X) **[Table/Fig-3]:** Section from tumor showing fascicles of long spindle cells having regular, oval nuclei with bland chromatin. (H&E,40X) **[Table/Fig-4]:** Smooth muscle actin: cytoplasmic positivity **[Table/Fig-5]:** HMB 45: negative

More than 50% cases reported in the literature presented with pain and a palpable mass, 20% presented with hematuria with a fair number of cases being incidental with no clinical signs or symptoms [6]. In this case as well the patient presented with a non specific complaint of flank pain.

These tumours are seen to have a female preponderance, specifically white women [3]. The average age at presentation ranges from 40-45 years [6]. Both the findings, not being in concordance with this particular case, make it an unusual presentation of renal leiomyoma.

On reviewing the literature for cases and clinical manifestations it was seen that Steiner and colleagues divided Renal Leiomyomas into two groups:

(i) Incidentally discovered at autopsy (very small lesions)

(ii) Clinically symptomatic lesions.

However, on further exploration it was found that Wagner and colleagues added a third group comprising of radiologically detected lesions without clinical signs and symptoms. With the new advents in radiology and imaging this third group encompasses a majority of leiomyoma cases [6].

In the first group, the average size is less than 5 mm, while in the second group of symptomatic leiomyomas an average size of 12.3 cm has been cited. Till date, the biggest renal leiomyoma encountered is 57.5 cm [8,9].

In the present case scenario the size was of 14x8.5x4 cm, a little larger than the average size mentioned in the literature studied.

On imaging Renal leiomyomas can present in a plethora of findings. On ultrasound they present as a solid mass, but cystic changes are not uncommon. On CT a well-defined mass is encountered. Leiomyomas of the renal capsule are occasionally attached to the cortex by a small stalk and irregular calcification may be seen. They are usually sharply demarcated without infiltration into surrounding tissue or evidence of metastasis [2]. The CT findings of this case were however suggestive of a Renal Cell Carcinoma of the left kidney.

On gross examination the renal leiomyomas have been described as well defined mass, solid and white or red in colour. The microscopic features comprise of fusocellular elements with absence of mitotic figures, pleomorphism, Hyperchromatism and, the absence of Perineural invasion. All of these features when present point towards the malignant counterpart of leiomyoma.

The differential diagnosis is mainly with Leiomyosarcoma the malignant counterpart possible only after nephrectomy on histopathological examination, as on radiology an affirmative diagnosis is not possible. Another important differential is of Angiomyolipoma (AML) of the kidney, which is mostly composed of a combination of thickened blood vessels, mature adipose tissue and smooth muscle cells. However at times only a smooth muscle component is most represented, which can then be confused with a spindle cell lesion. Therefore for its confirmation HMB45 a melanocytic marker is used [5]. In view of the microscopy of this case Adult Mesoblastic Nephroma was kept as first differential. However, on reviewing the immunoprofile for mixed epithelial and stromal tumours they were shown to be immunoreactive with antibodies to cytokeratin, especially cytokeratin 7 and show strong positivity in the epithelial elements [10].

Whereas, in the present case scenario Smooth Muscle Actin(SMA) was positive and Cytokeratin along with HMB-45 were negative, thus confirming the diagnosis of Renal Leiomyoma.

CONCLUSION

Renal Leiomyoma is an infrequent tumour that must be included in the differential diagnosis of renal masses. Laprotomy and nephrectomy are performed in most cases of suspected renal masses, as it is still the gold standard for their diagnosis. The definitive diagnosis of a leiomyoma is only possible after histopathological examination of the tumour. However, due to the rising number of diagnosis resulting from improved medical imaging and interventions, renal leiomyomas should gain importance in the differential diagnosis of renal masses prior to nephrectomy, with respect to kidney sparing surgery.

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