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A benign mimicker of renal cell carcinoma: A case report

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Abstract

We report a rare case of benign renal solitary fibrous tumor in a 40-year old male who presented with a vague fullness and dragging sensation on right lumber region. Computed tomography showed large heterogeneously enhancing solid cystic leision arising from upper pole right kidney for which radical nephrectomy was done. On histopathological examination the diagnosis of solitary fibrous tumor of kidney was made.

Keywords: Solitary fibrous tumor, benign, kidney

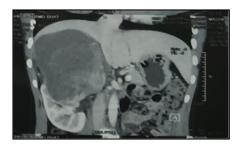
Introduction

Solitary fibrous tumor is an uncommon tumor of mesenchymal origin. It can occur at any site but most commonly arises from pleura. About 30% are found in soft tissues and other organs like kidney, prostate,, bladder, breast etc

Very rarely they can involve the kidney and on imaging solitary fibrous tumors can mimic renal cell carcinoma which can be ruled out by histopathological examination and immunohistochemistry confirmation. Mostly they are benign but may be aggressive and turn out to be malignant and hence, radical nephrectomy is always a preferred treatment.

Case Presentation

We present a case of 40 year old male patient who presented with pain and dragging sensation on right lumbar region intermittently. On physical examination a mass was palpable in the right lumbar region which was non tender and mobile. Urine examination was done which showed 30 to 40 RBCS /hpf along with few pus cells. Laboratory results show blood urea 20, creatinine 1.0 blood urea nitrogen, LFT and CBC were also within normal limits



 $\textbf{Fig 1:} \ CT \ view \ of the \ solitary \ fibrous \ tumor.$



Fig 2: Specimen of Solitary Fibrous Tumor on Cut

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CT abdomen (Figure 1) showed evidence of 12.9X 11.5 X15.8cms heterogeneous lesion showing solid to cystic component arising from upper pole of right kidney. The lesion showed calcifications. Perilesional fat stranding was seen with peri-renal collaterals. The lesion appeared

to cause compression of upper pole calyces, however no intra calyceal spread was seen. Based on the clinical and imaging procedures the patient was diagnosed as a case of renal cell carcinoma and was planned for right radical nephrectomy. The specimen was sent for histopathological examination. Grossly (Figure 2) the kidney measured 20x10x7 cms.oncut section it revealed a well circumscribed mass at upper pole measuring 12.5x12x11 cms. Cut section through the mass showed pinkish surface along with some grey white areas.

On histopathological examination (Figure 3) tumor showed hypercellular and hypocellular areas composed of spindle cells arranged in intersecting fascicles and in storiform pattern. Areas of thick ropy collagen, branching hemangiopericytoma like blood vessels and foci of calcifications were also identified. No atypical mitosis or areas of necrosis were seen.

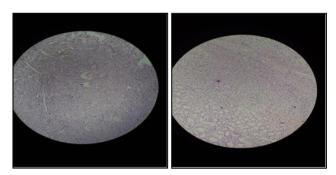


Fig 3: Solitary Fibrous Tumor (10X-HPE)

Discussion

Solitary fibrous tumor previously known as localised fibrous mesothelioma or hemangiopericytoma are rare mesenchymal tumors. Arising in age group of 4- 85 years [1-2]. There is no sex predilection [3]. Solitary fibrous tumors have been reported to arise from upper respiratory tract, orbit, meninges, breast and salivary gland [4]. But can occur at any site.

They are discovered incidentally but can be symptomatic. The patient can have hematuria, flank pain or abdominal fullness due to pressure effects. Solitary fibrous tumors can be associated with hypoglycemia due to production of IGF-1 [5]

Renal solitary fibrous tumors may arise from capsule [15%] or peripelvis [6%] and [3%] can arise from renal pelvis while 76% arise from unknown site $^{[6]}$. Till now 53 cases have been reported so far. Percentage of malignancy can range from 10-15% $^{[7]}$.

The malignant behavior is determined by presence of increased cellularity, pleomorphism and increased mitotic, activity greater than 4 mitosis per 10 high power fields, along with necrosis and hemorrhage ^[7]. Only three of the reported cases in literature had necrosis and hemorrhage ^[8]. In our case no mitosis necrosis or hemorrhage was seen. Grossly, solitary fibrous tumors are well circumscribed grey white firm tumors measuring 5 to 8 cm showing spindle shaped fibroblast like cells with a collagenous stroma on

microscopy. They contain pachy, hypo and hyper cellular areas. Cytoplasm is ill-defined. Solitary fibrous tumors are highly vascular tumors ^[6]. 10 to 15% of extra plural solitary fibrous tumors are malignant and aggressive ^[5]. Only one case of pulmonary metastases has been reported till now ^[9]. There are no reports of local recurrence after nephrectomy ^[10-11] but single case of metachronous solitary fibrous tumor of contralateral kidney was reported hence it is necessary that the patient be kept under imaging surveillance postoperatively ^[12].

On imaging CT may be non specific and may show areas of calcification, necrosis and enhancement. MRI detects fibrosis and dense collagen as low signal intensity on T2 weighted images. Some tumors may contain fat [13]. Some of the features may coincide with renal cell carcinoma, hence solitary fibrous tumors may be misinterpreted as renal cell carcinoma on imaging procedures.

Solitary fibrous tumors are strongly positive for CD34, bcl2, CD 99 and vimentin. CD34 and CD99 positivity is seen in in 92- 95% and 70- 75% of cases respectively ^[8]. 22% show positivity for bcl2 and 90% to 95% cases are strongly positive for vimentin ^[14]. They also show STAT-6 positivity which is more sensitive and specific..

Most solitary fibrous tumors do not recur or metastasize and most of them have a good prognosis [15]. 10% of extra plural and 10 to 15% of intra plural tumor are malignant [16]. Radical nephrectomy is the preferred treatment of choice and patient should be kept under long term follow up as it is difficult to predict the aggressiveness of the tumor and its potential to recur.

Conclusion

Renal solitary fibrous tumor is a closed mimicker of renal cell carcinoma on USG and CT scan as well as clinical features do not help to differentiate the two conditions. Only histopathological examination can make a definitive diagnosis of solitary fibrous tumor and help to differentiate from renal cell carcinoma. The prognosis of solitary fibrous tumor is better than renal cell carcinoma as most of the cases are benign, however, close follow-up is mandatory to look for any recurrence or change to any aggressive behavior.

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