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A Rare Case of Primary Retroperitoneal Schwannoma: A Case Report

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ABSTRACT
Schwannomas are tumors which arise from Schwann sheath of nerves and are very rare in the retroperitoneal region. They are commonly seen in the head and neck location. We report a case of retroperitoneal schwannomain a 50 year old man who came with complaints of right flank pain. The patient was evaluated with computed tomography (CT) and magnetic resonance imaging (MRI)of abdomen. Surgical resection was done and histopathology confirmed the diagnosis of schwannoma. In this report we illustrate the clinical and radiological features of this common disease in uncommon location.
Key Words: *Retroperitoneal Schwannoma, Rare Retroperitoneal Tumors.*

INTRODUCTION

Schwannomas (also known as neurilemmoma) are tumors that arise from schwann sheath of the peripheral or cranial nerves except I and II. They can occur anywhere within the neural tissue but more commonly seen in the head and neck region and rarely seen in the retroperitoneal region. They usually occur between third and sixth decades of life, with an equal predilection for men and women. Primary tumors of the retroperitoneal region are rare and schwannomas include only 1-10% of them [(1)].The majority of retroperitoneal schwannomas are benign in nature though malignant ones have also been reported[(2,3)]. A retroperitoneal involvement accounts for 0.3-3.2% of all primary schwannomas and 0.3-6.0% out of all retroperitoneal tumors[(4)].Here, we describe a case of retroperitoneal schwannoma which was histopathologically proven.

CASE REPORT

A 50-year-old man was initially evaluated in outside hospital with complaints of right-sided flank pain. Physical examination revealed abdominal tenderness in right lumbar quadrant. Laboratory tests were unremarkable. The patient

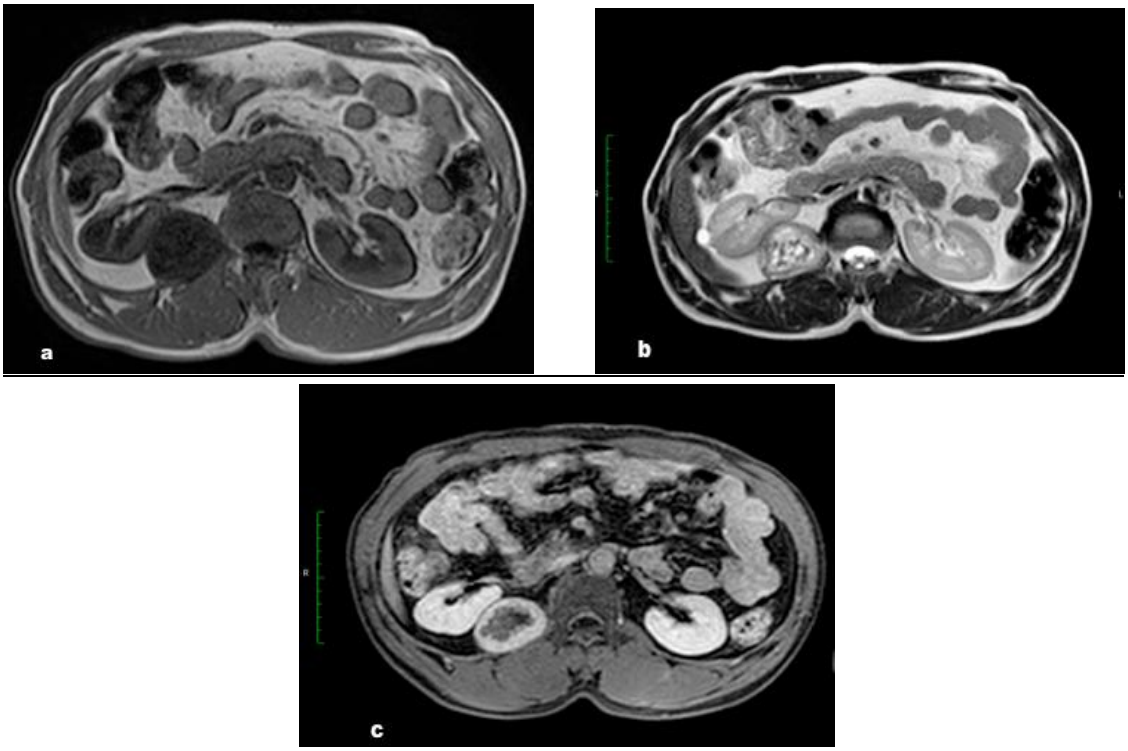
was clinically suspected for a renal colic and hence referred for a CT abdomen plain study which was done in outside hospital. CT images did not reveal renal or ureteric calculi but revealed a large round shaped, isodense soft-tissue mass in the right side of retroperitoneum in right posterior para renal space. There were a few punctate hyperdensities within the center of the lesion. Patient was referred to our institute for further management.

Patient was clinically reevaluated in our hospital. CT plain abdomen images done outside was reviewed. To further characterize and evaluate the lesion, contrast enhanced magnetic resonance imaging (MRI) of abdomen was performed with a 1.5 Tesla scanner (Philips Multiva) with Torso coil in our hospital. MRI revealeda large lesion located in right posterior para renal space. The lesion measured 5.5 x 4.2 x4.4cm. The lesion appeared hypointense on T1 and heterointense on T2 weighted images [Figure-1a,b]. Center of the lesion shows multiple small T1 hypointense and T2 hypointensities, suggestive of micro calcifications, which was reviewed on the plain CT images. No evidence of fat signal intensity was seen within the lesion. On post contrast images, lesion showed peripheral enhancement with central non-enhancing area [Figure-1c]. Central non enhancing area is suggestive of necrosis or cystic degeneration. The lesion was deep to right crus of diaphragm and superficial to right psoas and quadratuslumborum muscle. No obvious invasion was seen into adjacent right psoas and quadratuslumborum muscle [Figure-2]. The lesion was seen causing mass effect on right kidney midpole cortex. Fat plane was preserved between the lesion and right kidney. Right 12^{th}rib was adjacent to the lesion which did not show abnormal marrow signal or destruction, to suggest invasion. No definite neural for aminal extension of the tumor into spinal canal was seen. Adjacent vertebra was unremarkable. Inferior vena cava was separately seen. On dynamic contrast enhanced images, gradual enhancement pattern was seen in time signal intensity curve [Figure-3]. No evidence of diffusion restriction seen. No significant enlargedparaaortic lymph nodes were seen. Based on the imaging characteristics to the location and signal characters, diagnosis of retroperitoneal benign soft tissue tumor with necrosis and micro calcifications was made. Incidental right renal simple cysts were seen.

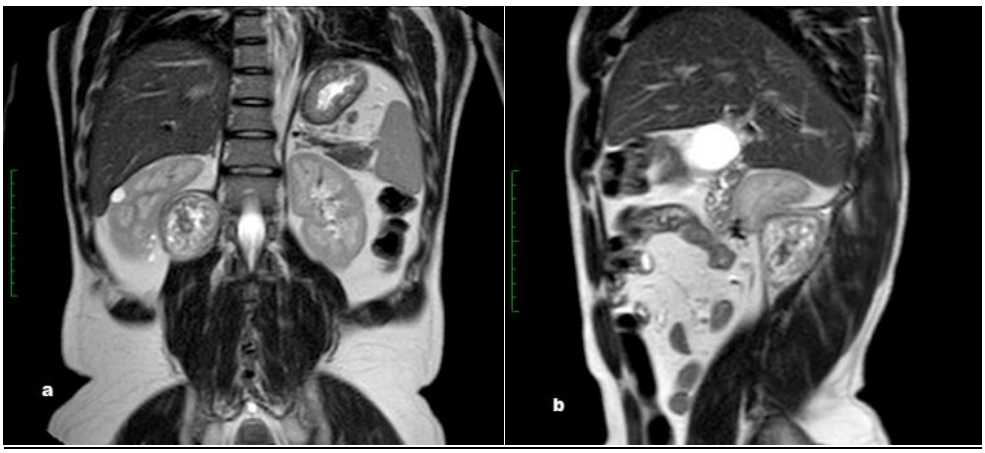
Surgery was planned, midline laparotomy incision was made, bowel loops were mobilized, peritoneum dissected. Globular grey white firm to hard mass was localized adjacent to right kidney just above the right psoas muscle, lateral to the inferior vena cava. Mass was in close proximity to the 12^{th} rib and the adjacent neural foramen. Mass was excised in toto by blunt and sharp technique [Figure-4].

Excised mass was sent to for histopathology examination. On microscopy, sections showed an encapsulated neoplasm composed of alternating hypo and hypercellular areas [Figure-5]. The hypercellular areas are composed of spindle cells in palisades with verocay bodies in myxoidstroma. Hypocellular areas contain areas of hyalinization, cystic degeneration, calcification, areas of ossification and hemorrhage. Histopathological diagnosis of ancient schwannoma was made.

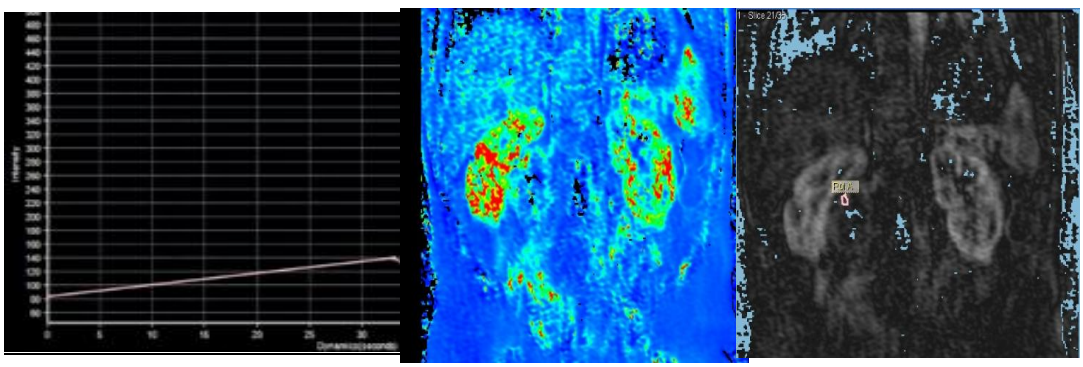
The patient is doing well and has been followed-up till date for a period of 6 months and there is no evidence of any symptoms or recurrence.



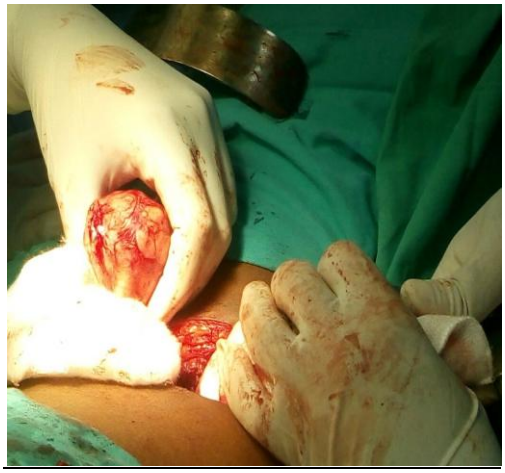
[Figure-1] : AxialT1 (a), T2 (b)and T1 fast suppressed contrast (c) images showing T1 hypointense, T2 heterointense, peripheral enhancing retroperitoneal mass in right posterior pararenal space.



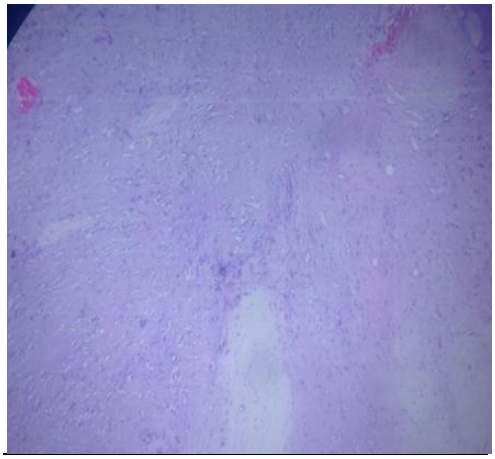
[Figure-2]: T2 coronal (a) and sagitta l (b) images showing T2 heterointense retroperitoneal mass deep to right crus of diaphragm and superficial to right psoas and quadratuslumborum muscle.



[Figure-3]: Dynamic contrast enhanced MRI study showing gradual enhancement pattern in time signal intensity curve.



[Figure-4]: Intraoperative image showing the globular right retroperitoneal mass.



[Figure-5]: Histopathology images showing encapsulated neoplasm composed of alternating hypo and hypercellular areas, suggestive of ancient schwannoma.

DISCUSSION

Schwannomas are classified as nerve sheath tumors that are mostly benign in nature. These neoplasms are usually seen third and sixth decades of life. Symptoms of benign schwannomas are highly nonspecific and depends on the location and size of the lesion.

In the retroperitoneal region, the presence of an schwannoma is a rare entity except in patients with Von Recklinghausen's disease[1]. The malignant change usually takes place when it is associated with Von Recklinghausen's disease. In this patient, there was no association of Von Recklinghausen's disease. Localization in deeper structures such as the posterior mediastinum and retro peritoneum is unusual, accounting for only 1-3% of all schwannomas and only 1% of all retroperitoneal tumors[5].

Schwannomas are located typically eccentric in relation to the nerve of origin. They have a true capsule, which is composed of epineurium. Calcification has been reported in about 23% in retroperitoneal schwannomas, which has been observed in our patient on unenhanced CT images.

In general, MRI is regarded as the modality of choice for the evaluation of retroperitoneal tumors. They allow better evaluation of the origin, extent, and internal composition of these lesions. There are some imaging characters for schwannomas, which are "target sign and fascicular sign". However, these signs are not seen frequently in retroperitoneal schwannomas. The "fascicular sign" denotesthe appearance of bundles, a general property of neurogenic tumors. On the other hand, the "target sign" is the occurrence of hypointense center and hyperintense periphery on T2 sequence of MRI. Cystic degeneration has been reported frequently in retroperitoneal schwannomas with incidence of about 66%.

On histopathology, two main microscopic patterns are recognized: A highly cellular component (Antoni A cells) and a myxoid component (Antoni B cells). The predominance of one or other reflects the heterogeneous findings at CT and MRI. They appear as alternating hypo and hypercellular where hypo represents hyaline cells and hyper represents hyaline cells. Most of the schwannomas exhibited intense immunohisto chemical staining for S100 protein, confirming its neuroectodermalorigin[5].

The differential diagnosis for retroperitoneal schwannomas involves sarcoma, other neurogenic tumors such as paraganglioma and pheochromac-ytoma and malignant fibrous histiocytoma. In addition if the retroperitoneal schwannoma contains significant amount of cystic degeneration, retroperitoneal masses such as hematoma and lymphangioma should be ruled out.

Although rare, malignant counterparts of schwannomas do exist. Detection of a malignant schwannoma is very important, since it will affect the treatment strategy. Malignant schwannomas tend to appear with an irregular contour and have a propensity for invasion to the adjacent structures.

Complete surgical excision is the treatment for both benign and malignant retroperitoneal schwannomas[6]. Radiotherapy and chemotherapy provide no added value[7]. Benign schwannomas have a local recurrence rate of about 10-20%, which may be sometimes due to an incomplete surgical excision and hence an enucleation is not sufficient[8]. In one study, it was evident that two patients undergoing surgery developed complications[7]. One patient developed L2-L4 root injury and the other patient developed L5-S1 nerve impairment, which well recovered following physiotherapy. There were no complications post operative in our patient, the symptoms reduced and he has been followed-up till date for a period of 5 months without evidence of any recurrence.

For surgical treatment of retroperitoneal schwannomas, the current approach is by endoscopic assisted mini laparotomy. Aggressive surgery is not often indicated for benign retroperitoneal schwannomas. Though local resection is generally enough, metastatic cases have been reported in literature after resection [1,2].

CONCLUSION

To conclude, we present common tumor in a rare location, which was detected incidentally in evaluation of a patient with renal colic. Rare tumoral lesions with a benign course such as schwannoma can be detected incidentally. The preoperative differential diagnosis of retroperitoneal schwannoma is useful for meticulous dissection and complete excision of the tumor. Complete excision and follow-up is mandatory to prevent recurrence. Physicians should bear in mind the possibility of benign schwannoma in retroperitoneal mass lesions.

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