

Case Report

Retroperitoneal schwannoma: case report

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ABSTRACT

Retroperitoneal space is a rare location for schwannomas accounting for about 1-3% of all schwannomas with wide variety of symptoms on presentation. We herein report a case of retroperitoneal schwannoma in an adult who suffered from left lower abdominal pain for 1 year. We also highlight the chain of investigations, surgical management that was undertaken and the postoperative complication that the patient developed.

Keywords: Retroperitoneal mass, Schwannoma, Ultrasound guided biopsy

INTRODUCTION

The occurrence of retroperitoneal schwannoma is very rare, this location accounts for 1-3% of all schwannomas and 1% of retroperitoneal neoplasms.¹

Most schwannomas are benign and usually arise as solitary tumors from the peripheral nerves. The malignant form is much more uncommon.² Most of the schwannomas are asymptomatic and found accidentally.³

CASE REPORT

We report a case of a 35 years old female who had presented to us with complain of lower abdominal pain for 1 year, it was episodic, dull and aching, more in left lower quadrant, radiating to leg. The clinical examination showed a positive straight leg raise test.

Abdominal CT scan was performed revealing a lesion in left para-aortic region measuring 6.7×6.3×10.0 cm with small focus of calcification, causing mild lateral displacement of left kidney and compressing the left proximal ureter resulting in mild left sided hydronephrosis and proximal hydroureter, the lesion was also abutting the

left renal vein, aorta and small bowel loops with intact fat planes. U/S guided biopsy was done which showed cells that stained positive for S-100 and SOX10 suggestive of spindle cell neoplasm with neural differentiation. Intrabdominal schwannoma and malignant peripheral nerve sheath tumor (MPNST) were differentials based on histopathology.

Patient underwent a midline laparotomy. Peroperatively the mass seemed to be arising from left paravertebral space, had multiple feeding vessels from aorta. Colonic mesentery was splayed over it, left ureter was adherent to posterolateral aspect of the mass, left renal vein was in close contact with the superior surface of the mass. There were large retroperitoneal lymph nodes that were removed with the tumor.

Histopathology showed circumscribed well encapsulated spindle cell neoplasm with no infiltrative edges. Postoperatively, the patient had developed bilateral hydronephrosis and acute kidney injury due to perop dissection and manipulation of ureters, which was managed by bilateral percutaneous nephrostomy tube placement, after which the renal functions improved. She was then discharged home on 8th postoperative day.

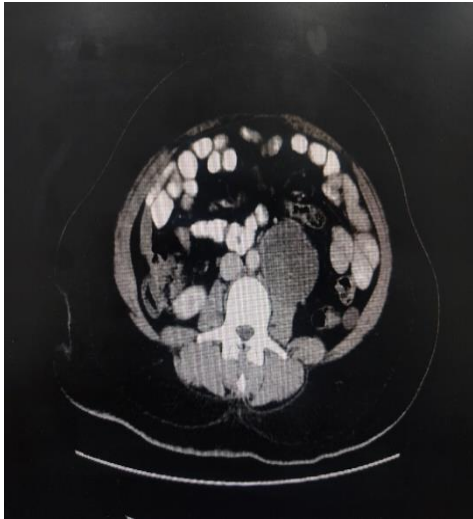


Figure 1: Axial view of abdominal CT showing left retroperitoneal mass.



Figure 2: Mass abutting the left kidney and causing mild hydronephrosis.



Figure 3: Axial view of CT abdomen after removal of retroperitoneal mass.

DISCUSSION

Schwannomas are neurogenic tumors deriving from the Schwann cells of nerve sheaths, affecting predominantly females and mostly found in patients aged 20 to 60 years.⁴

The diagnosis of retroperitoneal Schwannoma is late and difficult preoperatively because none of the clinical signs are pathognomonic to this tumor. Benign Schwannomas do not invade nearby organs, the symptoms are due to the compression and displacement of the retroperitoneal structures. In a series of 82 patients with Retroperitoneal Schwannoma, the majority of patients had abdominal distension with no other associated signs (30.5%) or the diagnosis was incidental (34.1%), with the remaining patients having abdominal pain (20.7%), low back pain (6.1%) or digestive problems (6.1%).^{1,2} In our case lower abdominal pain radiating to leg was the predominant symptom, suggestive of nerve compression.

Radiologic examination plays a major role in the diagnostic approach. CT scan with contrast shows classically a well-defined tumor with slightly low density compared with soft tissue due to the high presence of myelin and fat. CT scan done in our patient showed faintly enhancing lesion with a small focus of calcification (Figure 1 and 2).⁴

MRI is considered the first-choice imaging tool for the exploration of retroperitoneal tumors, as it allows a better visualization of the origin of the lesion, its extent, its internal and vascular architecture and the involvement of other organs.¹ Schwannomas are commonly hypointense on T1 and hyperintense on T2-weighted MR images. The target sign (a hypointense center with a hyperintense periphery) and fascicular sign (bundles) are two well-known characteristic features of neurogenic tumors on MRI; however, they are not seen frequently in Retroperitoneal Schwannomas. Percutaneous biopsy is a matter of debate. Some physicians skipped it due to the possibility of biopsy-related complications (bleeding, infection, tumor seeding) while other physicians managed patients conservatively due to benign biopsy results.⁴ We opted to biopsy the lesion at the expense of all the expected complications to characterize the histological nature of the mass to decide on any neoadjuvant therapy.

The final diagnosis is solely based on histopathologic examination of the specimens. RS is histologically characterized by alternating antoni A and antoni B areas. In antoni A tissue, the cells are arranged in an organized compact pattern, while in antoni B tissue, the cells are scattered loosely in an edematous matrix immunohistochemically, positive expression of S-100 and negative expression of CD-34 is valuable for diagnosis.^{3,4} Despite recent research on the therapeutic strategies against RS, complete surgical resection appears the only potentially curative approach. Subtotal resection may be performed to minimize surgical risk and preserve surrounding vital structures and some surgeons believe

that simple enucleation or partial excision is sufficient since not all patients are candidates for complete excision.⁵ In our patient the tumor was adherent to left ureter, which was preserved by preoperative ureteric stenting, it was also closely abutting renal vessels and aorta which were preserved by careful hand dissection and using liga clips for hemostasis and hence safe resection without a capsule compromise was ensured. Alternative therapies, such as radio- and chemotherapy often proved insufficient.⁵

The prognosis of benign schwannomas is good and the most frequent complication is recurrence, probably due to incomplete excision, which is reported in 5-10% cases.³

CONCLUSION

Retroperitoneal schwannomas are rare tumors. The diagnosis is often delayed in most cases due to vague presenting symptoms. Percutaneous biopsy, although controversial, can guide the management. Complete surgical resection remains the standard treatment.

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