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Case Report

Ancient schwannoma: a rare retroperitoneal tumor

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ABSTRACT

Schwannoma is a tumor of soft tissue, originating from Schwann cells which rarely appears in the retroperitoneal region. Retroperitoneal ancient schwannomas are rare tumors, more usually found in the head, neck and flexor surfaces of the extremities. Ancient schwannomas are a subtype of classic schwannoma with a predominance of degenerative changes like calcifications, hemosiderin deposition, interstitial fibrosis, vascular hyaline degeneration. We report a case of 60 years old female with complaints of vague pain in upper abdomen with discomfort since 6 months. Following imaging studies, retroperitoneal mass was found. The patient underwent surgery and excision of mass was done. Histopathological examination showed degenerative changes which was consistent with ancient schwannoma. No evidence of recurrence appeared during follow up period.

Keywords: Retroperitoneal tumor, Retroperitoneal schwannoma, Ancient schwannomas

INTRODUCTION

Schwannoma is a soft tissue tumor arising from Schwann cells of the peripheral nerve sheath. hence, it can appear in any Schwann cell of the nervous system, including trunks and extremities. It rarely occurs in the retroperitoneum region (only 0.5%–5% of all schwannomas).[1] Schwannomas account for 1% of all retroperitoneal tumours.[2]

Ancient schwannomas are a rare variant of peripheral nerve sheath tumors, or schwannomas. The term "ancient" refers to the histological degenerative features, which are acquired with increasing age in these tumors. They are slow growing and may produce vague local symptoms, but are usually diagnosed incidentally. They are an uncommon cause of retroperitoneal mass and classically encapsulated, highly vascular and have a distinctive radiological appearance.[1,3] Complete surgical removals with safe limit is the standard treatment, recurrences are rarely reported.

In this report we discuss a case of retroperitoneal schwannoma displacing the right kidney and placed between abdominal aorta and inferior vena cava, which was operated in our hospital.

CASE REPORT

A 60 years old female came to surgery opd with complaints of vague upper abdominal pain with abdominal discomfort for 6 months. Patient gave history of fullness in upper abdomen. Patient had no comorbidities. Patient's generalized condition was fair and vitals were within normal limit. Pallor was present.

Per abdominal examination had mild tenderness in the right upper abdomen. Rest of the examination was normal.

On investigating further haematological and biochemical investigations were within normal limits.

Ultrasonography (USG) abdomen and pelvis suggestive of well-defined thick-walled oval lesion of size 8.3×8.8×8.2 cm seen in sub hepatic region with echogenic material within having minimal vascularity and few hyperechoic thick septae within seen in retroperitoneum with

maintained fat planes with surrounding structures likely s/o retroperitoneal mass of neoplastic etiology.

Contrast enhanced computed tomography (CECT) A+ P s/o well defined retroperitoneal lobulated solid cystic mass lesion of approximate size of 9×9.3×11 cm (AP×TR×CC).

Solid component showed mild heterogeneous post contrast enhancement.

It was compressing and displacing IVC, right renal vein anteriorly. It was also displacing head of pancreas, 2nd and 3rd part of duodenum anteriorly closely abutting them.

Posterolaterally it is seen closely abutting the right kidney. Also related to ureter without causing hydroureter. Posteromedially related to abdominal aorta, right renal artery.

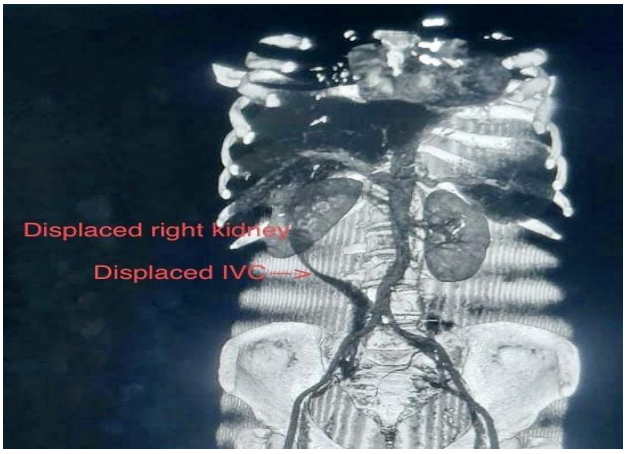

Figure 1: A large solid cystic tumor compressing and displacing IVC, right renal vein anteriorly.

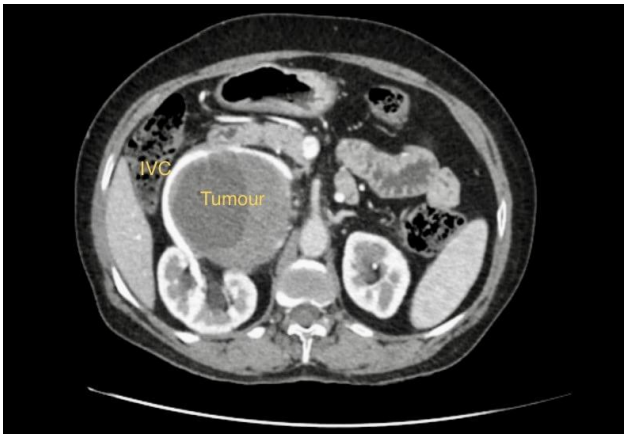

Figure 2: Abdominal CT showing tumor occupying retroperitoneal space.

After this patient underwent laparotomy. Right colon reflected and retroperitoneum reached E/o tumor that was displacing and compressing the IVC. Displacing right kidney and right renal vein. IVC was stretched over the tumor. Distal IVC control was taken and then dissection was slowly progressed and tumor was separated from IVC, right renal vein, right kidney. The tumor was excised. The specimen was sent for histopathological examination. Intraoperative and post-operative period was uneventful.

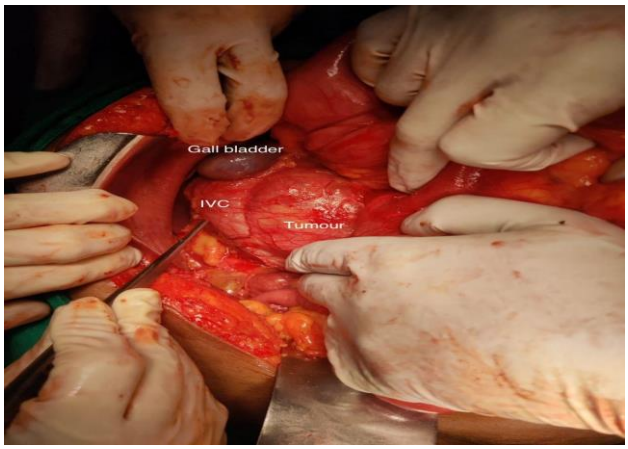

Figure 3: Intraop image showing tumour compressing and displacing IVC.

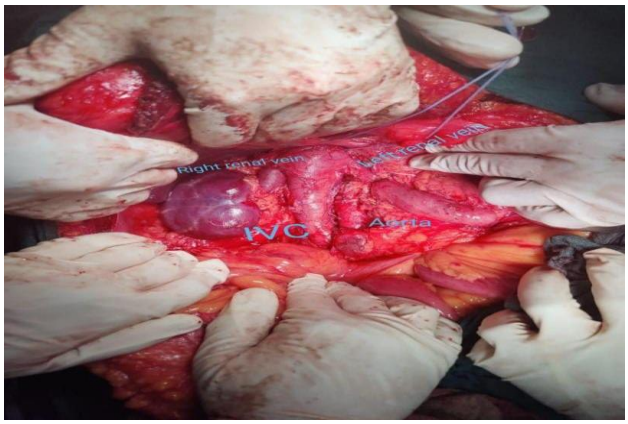

Figure 4: Intraop image showing structures after removal of tumour.

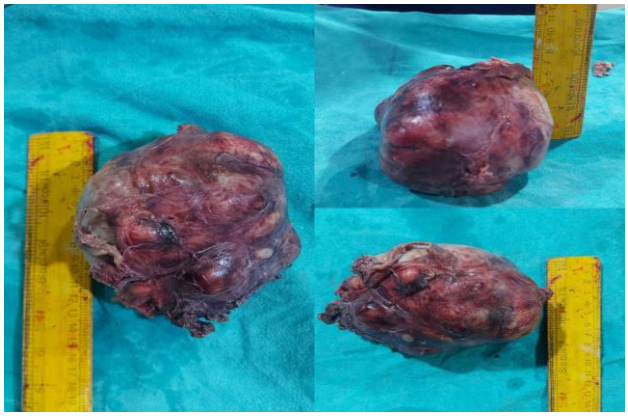

Figure 5: Gross specimen of retroperitoneal schwannoma of size 14×7×7 cm. Externally capsulated greyish brown lobulated tumour.

Histopathology report s/o well encapsulated tumor showed hyper and hypocellular areas. At places cellular atypia, cystic degeneration, hemosiderin deposition and vascular hyalinisation noted. Areas of necrosis and hemorrhage noted. No e/o mitotic activity. Histological features s/o ancient schwannoma. Patient was followed up for 6 months and was free of recurrence.

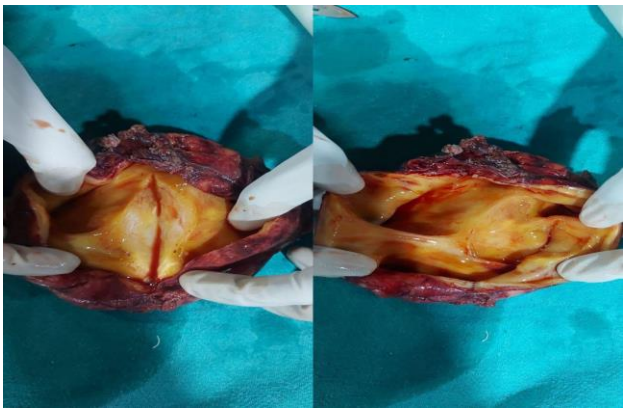

Figure 6: Macroscopic section of tumour showing solid and cystic areas.

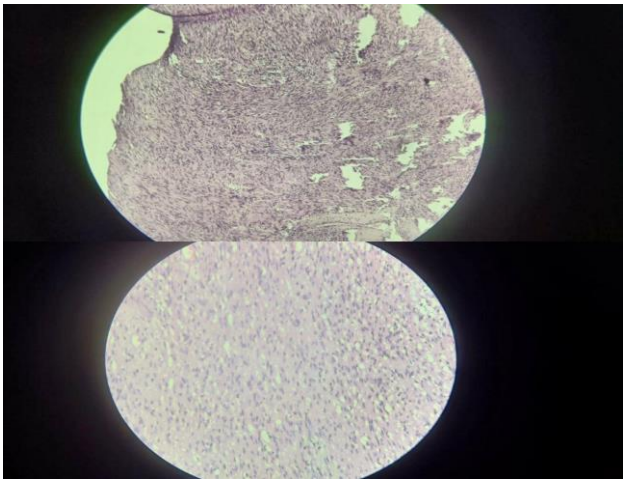

Figure 7: Microscopic image showing the tumour is composed of spindle cell proliferation with hyper and hypocellular areas.

DISCUSSION

Retroperitoneal schwannoma is a solid, encapsulated tumor which is benign in nature and arises from the paravertebral region. Macroscopically, schwannoma is solitary, well-defined mass, with a soft surface. Histologically, schwannoma consists of Schwann cells with hypercellular and hypocellular regions known as Antoni A and Antoni B with positive diffusion of S100 protein. The presence of degenerative changes, such as hemorrhages, cyst formation calcification and hyalinization, is a sign of ancient schwannoma which is a subtype of retroperitoneal schwannoma.[4] Ancient schwannoma is a rare variant of schwannoma first described by Ackerman and Taylor in 1951.[5] The term 'ancient' verbalize characteristic hypocellular and degenerative areas which is believed to occur with the long term progression of the tumour.[1] These cases are typically diagnosed in patients between 40 and 60 years of age, with a male-female ratio of 2:3.[6] Diagnosing retroperitoneal schwannomas is difficult. One study reported that symptoms were nonspecific, and neurologic symptoms were rare.[4] Symptoms include vague abdominal pain, flank pain, hematuria, headache, secondary hypertension, and recurrent renal colic pain.[7] Preoperative diagnosis is difficult because of the absence of pathognomonic features.[4] Total excision (including adjacent tissue if needed) is the best therapy for retroperitoneal schwannoma.[4,6,8,9] Because of the benign nature of most of the retroperitoneal schwannomas, some authors have recommended simple intralesional enucleation of the tumor and laparoscopic piecemeal excision.[10,11] Complete surgical resection with preservation of the surrounding structures mostly ensures cure in these patients.

The prognosis is good, and the most common complication is recurrence, possibly by incomplete excision of it being reported in 5–10% of cases.[8]

In our case patient was followed up for 6 months postoperatively. On follow up ultrasonography of abdomen and pelvis was performed, there was no evidence of any recurrence.

CONCLUSION

Ancient schwannoma is a rare subtype of retroperitoneal schwannoma, it is clinically often undetected or misjudged for other diseases. Imaging studies, followed with postoperative histopathological examination is used to confirm the diagnosis.

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