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Atypical Presentation of a Retroperitoneal Schwannoma Mimicking Gastrointestinal Stromal Tumor (GIST): A Case Report

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ABSTRACT

Schwannoma is a rare benign tumor that comes from the Schwann cells that sheath the peripheral nerves. We report the case of a 30 years old female patient who presented with non-specific abdominal pain and swelling in the left lumbar region for one year. CT scan findings revealed large soft tissue mass at left mid abdomen related to small bowel possibly Gastro-intestinal stromal tumor (GIST). USG findings revealed intra-abdominal cystic mass. After diagnosis patient was prepared for surgical removal. Patient was underwent complete surgical excision with an uneventful postoperative course. The histopathological study confirmed the nature of Schwannoma. Complete surgical excision remains the gold standard for the management of these tumors. The preoperative diagnosis is usually difficult due to their rare and non-specific clinical and radiological features; however, the definitive diagnosis is based upon histopathological examination and immune-histochemical.

KEYWORDS: Retroperitoneal Schwannoma, GIST, Histopathology, Complete surgical excision

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INTRODUCTION

Schwannomas are encapsulated tumors arising from the neural sheath of peripheral nerves. They are usually present in the head and neck or in the upper extremities, but may appear in the posterior mediastinum and more rarely in the retroperitoneum.[1]Retroperitoneal schwannomas are a very uncommon variety, making up about 3% of all schwannomas and 4% of retroperitoneal tumors. The sporadic form of retroperitoneal schwannomas is the most common, and predominantly affects females between the second and fifth decades of life.[2] The rarer malignant form is often associated with neurofibromatoses (von Recklinghausen disease).[3] Retroperitoneal schwannomas are mostly asymptomatic and found incidentally unless they grow large enough to cause compressive symptoms. The retroperitoneum is non-restrictive, so that benign tumours, such as schwannomas, are often able to grow to a large size before causing symptoms, which are usually vague and non-specific.[4] The lack of specific symptoms sometimes makes it difficult to accurately diagnose pre-operatively.[5] CT and MRI findings show characteristic features such as a well-demarcated, homogeneous, spherical, solitary mass, but none are specific.[4] Here, we report a case of a 30-year-old young female with retroperitoneal schwannoma. Written consent was obtained by participant in this study.

CASE SUMMARY

A 30 years old normotensive, non-diabetic lady presented with left sided lower abdominal lump for one year with occasional diffuse dull aching pain for six months. There was no history of fever, weight loss, night sweat, anorexia or any obstructive urinary symptoms. On physical examination general survey was essentially normal having no accessible lymph nodes palpable. Per abdominal examination revealed a non-tender, non-pulsatile, approximately 5cm X 4cm lump occupying left lumbar region of abdomen with ill defined margin, firm in consistency, mostly fixed but free from overlying skin with no expansile cough impulse. Her digital rectal, per vaginal and other systemic examinations revealed no abnormality. Abdominal ultrasound showed a thick walled non pulsatile cystic lesion measuring 6.4cm X 6.3cm was noted in left side of the umbilical region. A single septation with low level internal echo was noted within the cyst. CT scan revealed a lobulated heterogeneous enhancing 6.6 cm X 7.4 cm X 6.6 cm (AP X TD X CC) soft tissue density mass lesion occupying left mid abdomen with necrotic area noted within the lesion. The lesion was separated from left kidney and pancreas abutting the aorta and mildly compressing left mid ureter but related to small bowel loops; possibly GIST (Fig.1). No ascitis or paraaortic lymphadenopathy detected.

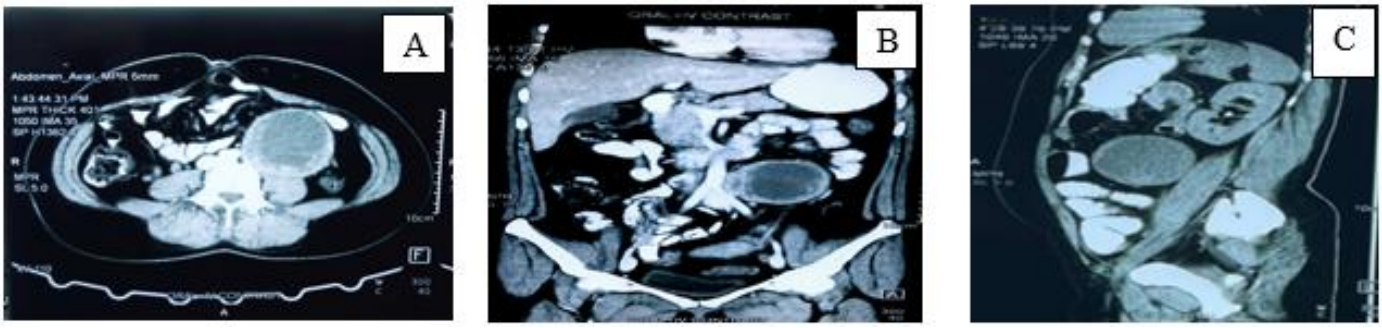

Fig.1. Computed Tomography (CT) of the abdomen in Axial (A), Coronal (B) and Sagital (C) sections. A large lobulated heterogeneously enhancing soft tissue density mass lesion noted at left mid abdomen, abutting the aorta and mildly compressing the left mid ureter.

Laboratory tests including tumor markers were normal. The patient underwent laparotomy and a partly cystic mass found on left lower abdomen arising from retroperitoneum pushing left ureter anteriorly (Fig.2).

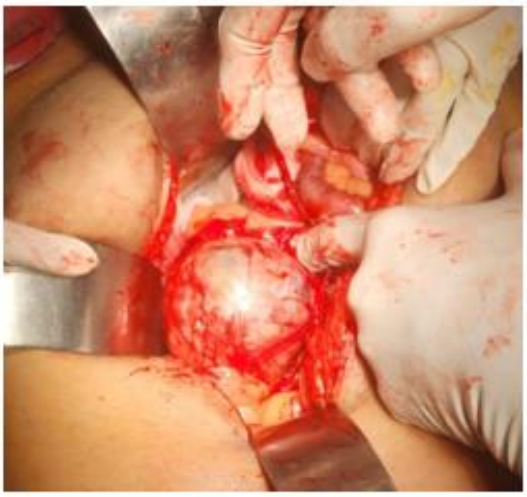

Fig.2. Intra-operative view showing a well-circumscribed mass in the left retro-peritoneum. Mass is being carefully mobilized and excised with preserving left ureter.

The mass was completely removed (Fig.3) preserving left ureter and keeping two drain tubes (one for retroperitoneal space and another on pelvis) in situ.

Fig.3. Excised retroperitoneal mass shows: A well encapsulated measuring about 7cm X 6 cm spherical lesion with smooth, glossy surface and evidence of vascular markings.

Fig.4. Gross cut section shows: Areas of cystic degeneration and necrosis, along with solid portions, some of which are brownish with hemorrhagic components.

Postoperative period was uneventful. Histopathological examination of the retroperitoneal mass along with Immunohistochemistry report revealed Schwanoma. Gross cut surface shows cystic and hemorrhagic fluid (Fig.4). Histologically, the cells were composed of hypercellular (Antoni A) and hypocellular (Antoni B) areas (Fig.5), no evidence of pleomorphism, mitotic figures or malignancy was observed. Immunohistochemical staining was strongly and diffusely positive for S-100 and SOX10 (Fig.6), while CD34, DOG1, SMA were negative. Ki-67 index was 1-2% immunoreactive, indicating low proliferative activity and confirming the benign nature of the tumor. Patient was discharged on her 5th postoperative day with instructions for regular follow-up. Patient's condition remained satisfactory with no sign and symptoms of any recurrence.

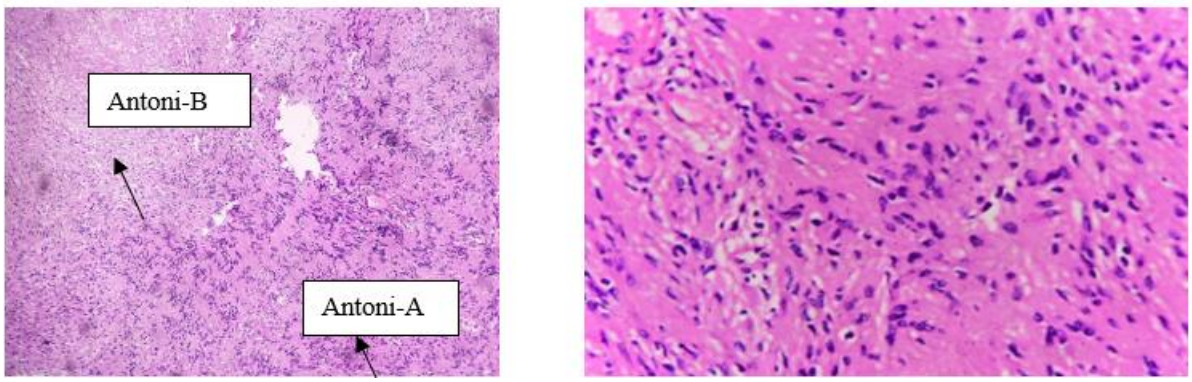

Fig.5. Histopathology report revealed mass composed of spindle cells with a focal nuclear palisading pattern composed of hypercellular areas which express Antoni A fibers and hypocellular areas which express Antoni B fibers.

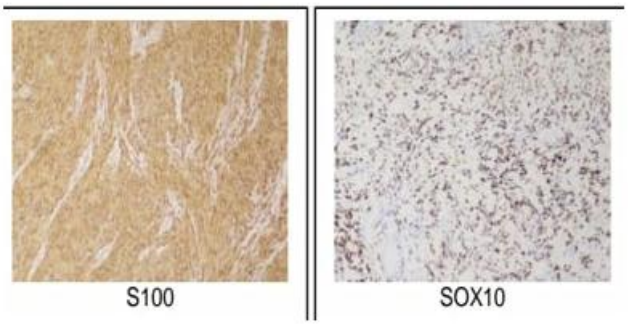

Fig.6. Immunohistochemical staining was strongly positive for S-100 and SOX10. Revealed Schwannoma.

DISCUSSION

Retroperitoneal schwannoma is a solid, encapsulated benign tumor that arises from the paravertebral region. Macroscopically, schwannoma is solitary, well-bordered mass, with a soft surface.[6] Preoperative diagnosis of retroperitoneal schwannoma is often difficult. There are no characteristic ultrasound, CT or MRI features to differentiate schwannoma from other differential diagnoses such as malignant peripheral nerve sheath tumors, sarcoma, lymphoma, neuroendocrine tumors (pheochromocytoma, paraganglioma), vascular tumors (hemangioma, cystic lymphangioma), fatty tissue tumors (angiomyolipoma, myelolipoma, lipoma), rhabdomyoma, or extragonadal tumors (teratoma or seminoma), among others [2]. However, the definitive diagnosis is based upon histopathological examination and immune-histochemical. Histologically on gross inspection, retroperitoneal schwannomas are solitary, well-defined and firm tumors. Microscopically they consist of spindle cells in areas with high (Antoni A) and low (Antoni B) cellularity.[7] The hallmark pattern of the benign variants is an alternation of these Antoni A and B areas, with a diffuse positivity for S100 protein in the cytoplasm of the tumor cells. [8] Schwannomas are resistant to radiotherapy and chemotherapy; therefore, surgical intervention is the only treatment option available. Benign schwannomas have a positive prognosis, but the most common complication is tumor recurrence, which occurs in 5– 10% of cases and is likely due to incomplete removal. To minimize the surgical risk and preserve adjacent vital tissues in benign types, it is recommended to perform subtotal resection.[9] Endoscopically assisted mini-laparotomy as an advanced approach to retroperitoneal Schwannoma. Post-operative follow-up plays an important role in management, considering the risk of recurrence.[10]

CONCLUSION

In conclusion, we report a case of a retroperitoneal schwannoma mimicking a GIST with illustration of the imaging findings. Despite the rare location of schwannoma in the retroperitoneum, they should be considered in cases of retroperitoneal abdominal masses.

CONFLICT OF INTEREST: None.

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