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Case Report

Postural limb abnormality: an unusual presentation of retroperitoneal Schwannoma

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ABSTRACT

Schwannoma is a tumor of soft tissue originating from Schwann cells that rarely appears in the retroperitoneum. Presentation depends on the site of disease leading to mostly pressure symptoms, sometimes with distal manifestations. Because of its complex location, it has varied presentations and is always a diagnostic dilemma. We reported a rare case of retroperitoneal schwannoma having mild difficulty in walking due to flexion deformity of left hip joint as the only initial presenting symptoms. Following imaging studies, a retroperitoneal mass was found. Partial excision of the tumor was performed, with the histology finding consistent with the retroperitoneal schwannoma. Patient was followed up for the last 2 years and had no evidence of recurrence.

Keywords: Retroperitoneal Schwannoma, Retroperitoneal tumor, Limb deformity, Partial excision

INTRODUCTION

Schwannomas are rare tumors of neurogenic origin that arise from Schwann cells of peripheral nerve sheaths. Retroperitoneal location is even rarer accounting for about 0.3-3% of all primary schwannomas.[1] They are usually benign, but malignant transformation can occur. Presentation depends on the site of disease leading to mostly pressure symptoms, sometimes with distal manifestations.[2] Because of its complex location, it has varied presentations and is always a diagnostic dilemma.

We report a rare case of retroperitoneal schwannoma having mild difficulty in walking due to flexion deformity of left hip joint as only initial presenting symptoms. The aim of this case report is to emphasize on the overall understanding of the diagnosis, treatment and prognosis of retroperitoneal schwannoma.

CASE REPORT

A 35-year-old gentleman came with mild difficulty in walking due to flexion deformity of his left hip joint. His sensori-neural and bony examination was normal. Abdominal examination revealed a retroperitoneal lump in the left lower quadrant. Ultrasonography abdomen showed a well-defined hypoechoic lesion of 25×11×8.5 cm in left lumbar region with multiple calcific foci and cystic areas. Computed tomography (CT) showed an encapsulated mass with variegated consistency, posterior to the left psoas which was mildly inflamed. Fat planes around the mass was maintained except in the medial region. He underwent exploration with removal of the mass except the medial 2cm which was attached densely to the retroperitoneum.

The histopathology of the mass revealed the schwannoma to be benign. It showed a well-encapsulated tumor with

many calcific, congested and hemorrhagic areas. The tumor showed cores of spindle cell lesion with moderate cellularity. No mitosis was seen. Immunohistochemical staining showed cellular positivity for S-100 marker.

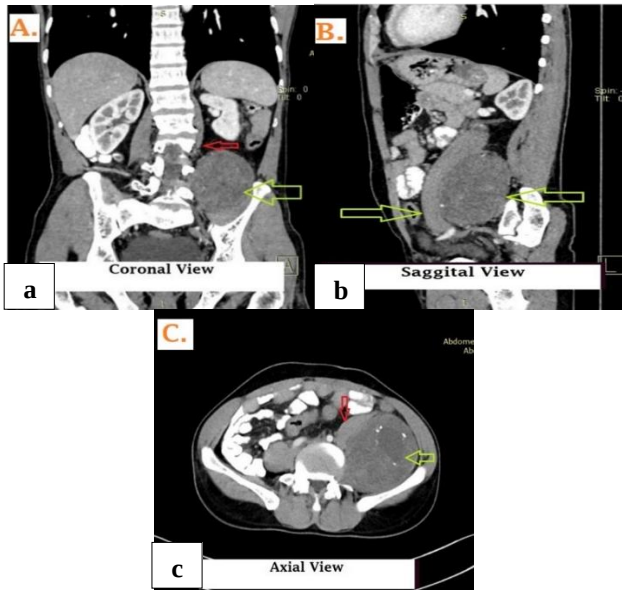

Figure 1: Radiological features of a retroperitoneal schwannoma, (a) coronal section showing a well-defined heterogeneously enhancing soft tissue density arising from lateral aspect of left psoas at level of L4 vertebra; (b) sagittal view showing abutment of the left psoas; and (c) axial view showing a retroperitoneal mass of size 8.6×8.8×11.3 cm having coarse calcific areas and few non enhancing areas suggestive of necrosis.

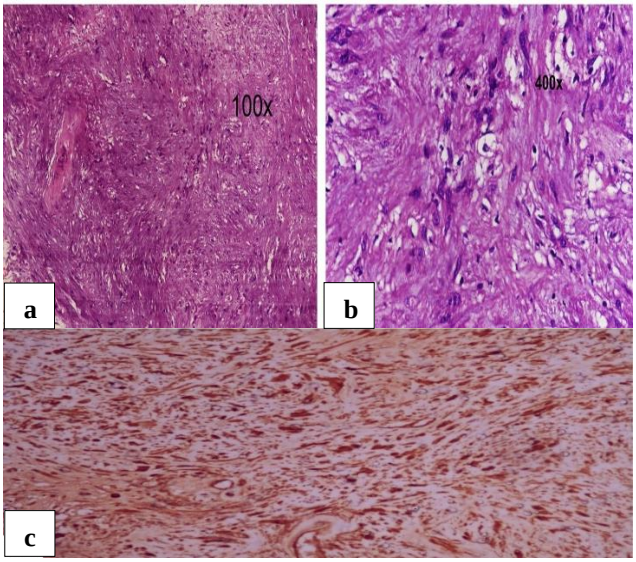

Figure 2: Photomicrographs of the tumor; (a) and (b) spindle-shaped cells in a typical palisading pattern and areas of myxoid and degenerative tissue with fewer cells (H&E; x100 and x400); and (c) immunohistochemical staining showed positivity for S-100.

DISCUSSION

Retroperitoneal schwannoma was first reported by Stallworthy in 1944 and most of the cases are reported in 3rd to 6th decade in life, with equal predilection for men and women.[3]

The retroperitoneal schwannoma is usually detected in the advanced stage with symptoms of flank pain and abdominal mass owing to the high flexibility of the retroperitoneal space. In the early stage it is usually detected incidentally on CT or magnetic resonance imaging (MRI) done for the vague symptoms.[4] In our patient, he had mild difficulty in walking due to gradual onset flexion deformity at the left hip joint. The patient when examined clinically revealed an intraabdominal, retroperitoneal lump in the left lower abdomen which gave us the clue for the diagnosis before imaging.

In spite of the recent advancement in the imaging modalities, the diagnosis of a retroperitoneal schwannoma can be challenging.[5] On computed tomography, the mass is usually well defined with low or mixed attenuation and having cystic, necrotic and calcific areas. MRI provides more accurate preoperative diagnosis as compared to CT. It appears hypointense on T1 weighted sequence while hyperintense on T2 weighted images. The intensity of the signal in T2 is inversely proportional to the cellularity of the tumor as compared to the non-cellular and necrotic zones which accentuate the T2 hypersignal. The malignant cases usually exhibit mixed intensity on both T1- and T2-weighted images. Although these findings are characteristic, they are not specific to make the preoperative diagnosis of retroperitoneal schwannoma.[6]

The differential diagnoses with schwannomas include fibrosarcoma, liposarcoma, ganglioneuroma, paraganglioma and neurofibroma which have similar findings on CT and MRI scan.[7] In addition, preoperative image-guided biopsy of a retroperitoneal lesion is not recommended due to the risk of great vessels injury, chance of tumor seedling and infection.[8] Complete surgical resection with negative soft tissue margin is considered as the treatment of choice. However, controversies still exist about whether excision of adjacent tissue and viscera. Some authors suggested that simple enucleation or partial excision without removal of adjacent organs was sufficient because of the benign nature for most cases and loss of adjacent organs may influence the prognosis.[9] In our case, we performed partial surgical excision as the tumor was densely adhered to the left psoas and retroperitoneum. Excising the left psoas can further affect the gait of the patient and can alter the prognosis.

Schwannomas are usually solitary, well encapsulated, firm with smooth surface. They frequently undergo secondary changes including calcification hemorrhage and cysts. On histopathology they appear in two distinct areas namely Antoni A and Antoni B areas. Clusters of compact elongated bipolar spindle cells arranged in a palisading

pattern are contained in Antoni A areas, whereas Antoni B areas are characteristic by loosely arranged cells in a myxoid background. However, in large retroperitoneal and pelvic schwannomas, uniform spindle cell appearance without independent Antoni A and Antoni B areas have also been reported.[10]

On immunohistochemical staining, they show cellular positivity for S-100 marker which is confirmatory for the diagnosis of schwannoma. The diagnosis of malignant retroperitoneal schwannoma may be suggested if mitotic figures, nuclear atypia and blood vessel infiltration are observed histologically.[11]

A benign retroperitoneal schwannoma has an extremely good prognosis and low recurrence rate. If recurrence occurs, still surgical resection is treatment of choice. Close follow up is necessary after removal of benign or malignant retroperitoneal schwannomas.[12] In recent years, laparoscopic excision and robotic resection have emerged as promising surgical approaches, but the location and the size of tumors may affect the choice of surgical approaches.[13,14]

CONCLUSION

Retroperitoneal causes of left hip joint deformity should always be kept in mind. However, it requires a very high index of suspicion to diagnose a retroperitoneal schwannoma causing a limb deformity. Thorough clinical examination and radiological imaging can be a useful adjunct. Histopathological examination confirms the diagnosis while surgery remains the treatment modality of choice in these cases.

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