Case Report

Silent giant: A case report of retroperitoneal schwannoma

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ABSTRACT

Schwann cells of the peripheral nerve sheath give birth to soft-tissue tumors known as schwannomas (neurilemmomas). Schwannomas of the retroperitoneum are uncommon. Diagnosis is frequently delayed in view of unspecific clinical symptoms and non-typical radiologic findings. The preferred course of therapy is total surgical excision. The prognosis is favorable; however, further monitoring is required due to the possibility of recurrence and malignant change. In this case report, we describe a case of retroperitoneal schwannoma in a 44-year-old male and examine the pertinent research.

Key words: Retroperitoneal, Schwannoma, Soft-tissue tumors

Schwann cells of the peripheral nerve sheath give birth to soft-tissue tumors known as schwannomas (neurilemmomas). They typically affect the flexor surfaces of the extremities, the head, and the neck. However, schwannomas can also develop in the posterior mediastinum and, less frequently, in the retroperitoneum; these lesions make up around 3% of all schwannomas [1]. Most patients either have no symptoms at all or display general symptoms including stomach pain, discomfort, constipation, and deep vein thrombosis. Pre-operative diagnosis might occasionally be challenging due to the lack of particular symptoms. Although schwannomas are normally benign, they can occasionally develop into malignancy.

Due to the rarity of this tumor, there are limited data about the prognosis and treatment protocol in these patients. Hence, we are reporting one such case of retroperitoneal schwannoma with a complete history and treatment.

CASE REPORT

A 44-year-old male presented with a 2-year history of abdominal and back pain since 2017 that was localized in the left side without radiation. He also has complaints of diarrhea/constipation on and off for more than 6 months. In 2019, the patient underwent a workup. On physical examination, we found a painless palpable mass in the left hypochondrium, measuring approximately 12 cm from the left costal margin to the left iliac fossa. There were no associated ascites, venous engorgement, or hepatomegaly. The abdominal computed tomography (CT) scan in 2019 showed the presence of a multiple hypodense lesion in the left lumbar and left iliac fossa, the largest being 14 cm × 12 cm with calcification and non-necrotic enhancement. It was maintaining a well flat plane with the left kidney and multiple retroperitoneal lymph nodes were positive. A renal diethylenetriamine pentaacetic acid (DTPA) scan was done to assess the status of the left kidney in view of the pressure applied by the tumor. DTPA scan showed a hydronephrotic left kidney with a thin cortex and poor parenchymal function. The patient was planned for biopsy and further management but defaulted in view of personal family reasons.

The patient visited again in 2021, and contrast-enhanced CT abdomen showed a 12 × 10 × 16 cm soft-tissue mass in the left retroperitoneal space extending from D11 to L5 level with non-enhancing necrosis. The left kidney was displaced anteromedially and cranially till midline. The fat plane was maintained with the left kidney and the ureter was normal. A repeat DTPA scan showed a hydronephrotic left kidney with a thin cortex, poor parenchymal function, and no deterioration in compression to the 2019 DTPA scan. Ultrasound (USG)-guided biopsy was done and the diagnosis came as schwannoma, which was further confirmed by S100 positivity in immunohistochemistry (IHC). The patient planned for resection but defaulted again.

Finally, the patient visited again in 2023 and worked up for surgery. On the CT abdomen (Fig. 1), the lesion size increased to 18.8 × 10.8 × 18.8 cm with multiple foci of internal calcification and necrosis. The lesion was encased by a lumbar branch of the aorta. Anteriorly, it was displacing the left kidney and ureter anteromedially, pancreas body, and tail superoanteriorly, with anterior displacement of the splenic flexure. Medially, displacing the abdominal aorta and its branches to the right side. Laterally, it

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maintained a fat plane with the spleen and abdominal wall. Lesion extension was from D11 to L 5 vertebra.

The patient underwent exploratory laparotomy and mass resection. Post-operative histopathology showed that the tumor had biphasic components in the form of Antony A and Antony B areas. IHC later showed S100 positivity (Fig. 2).

DISCUSSION

Neurilemmomas, also known as schwannomas, are soft-tissue tumors that develop from Schwann cells in the peripheral nerve sheath and are most common in women between the ages of 20 and 50 years [2]. Only a tiny percentage of schwannomas, which make up to 6% of all primary retroperitoneal tumors, are detected in the retroperitoneum, and they are typically found in the paravertebral or presacral area. Due to the long latency of tumor evolution, diagnosis often becomes late. Retroperitoneal schwannoma's patients typically do not have any symptoms or show generalized symptoms such as constipation, deep vein thrombosis, abdominal pain, and discomfort [3].

Retroperitoneal schwannomas are a diagnosis of exclusion because there are no identifiable symptoms. Abdominal ultrasonography, CT, and magnetic resonance imaging (MRI) are imaging modalities for retroperitoneal schwannomas. An efficient, quick, and affordable initial diagnostic tool is abdominal ultrasonography. It details the schwannoma's solid and cystic characteristics [4,5]. A solid tumor with a cystic component that is well-limited is typically visible on an abdominal CT scan. MRI gives the same information, but due to its improved diagnostic predictability compared to ultrasonography or CT, it is widely recognized as the preferred imaging technique for the identification of the majority of soft-tissue tumors [2,4]. A hyposignal on T1 and a heterogeneous hypersignal on T2 are characteristics of schwannomas [6]. However, an MRI cannot tell a benign lesion from a malignant one. Uneven outlines, variability, size (>5 cm), rapid growth rate, and cystic components are a few characteristics that have been associated with a malignant nature [7]. The presence of Antoni A and Antoni B tissue, which are characterized by high and low cellularity, respectively, is used to make a definitive diagnosis and determine the kind of schwannoma [8]. The majority of authors do not advocate pre-operative CT- or USG-guided biopsy due to the challenges in interpretation and the risks of bleeding, infection, and tumor seeding. As a result, post-operative histology should be used to provide a firm diagnosis of schwannoma.

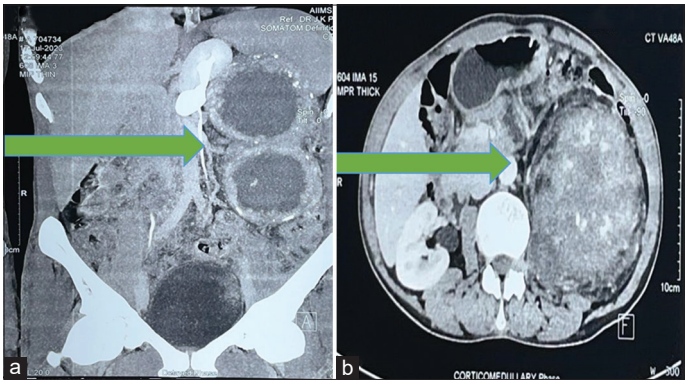

Figure 1: On contrast-enhanced computed tomography (abdomen) lesion size was 18.8 × 10.8 × 18.8 cm with multiple foci of internal calcification and necrosis. Anteriorly it was displacing the left kidney and ureter anteromedially, pancreas body, and tail superoanteriorly, with anterior displacement of splenic flexure. Medially, displacing the abdominal aorta and its branches to the right side. Laterally, it maintained a fat plane with a spleen and abdominal wall. Lesion extension was from D11 to L 5 vertebra

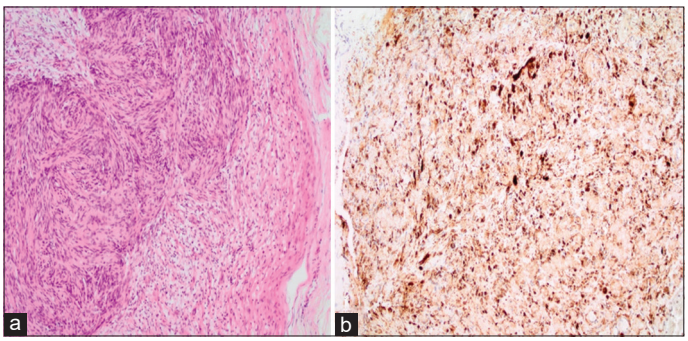

Figure 2: (a) Post-operative histopathology showed that the tumor had a biphasic component in the form of Antony A and Antony B area (hematoxylin-eosin); (b) S100 positivity

Radiotherapy and chemotherapy have no effect on schwannomas; the only effective treatment is total surgical removal [9,10]. The requirement for negative soft-tissue margins, particularly when surrounding tissue or viscera must be sacrificed, is up for debate. Others consider that simple enucleation or partial excision is sufficient, despite the fact that some recommend complete excision due to the danger of recurrence and malignancy that cannot be excluded before surgery. The likelihood of benign schwannomas turning cancerous is exceptionally low. Recurrence is the most common consequence and is described in 5–10% of patients, the most likely as a result of insufficient excision. These patients need long-term follow-up care, which includes a clinical evaluation and CT scan at 6 and 12 months following surgery, as well as once a year for the following 5 years [4].

CONCLUSION

Schwannomas of the retroperitoneum are uncommon. At the stage of a big tumor, the diagnosis is frequently delayed in view of unspecific clinical symptoms and non-typical radiologic findings. The preferred course of therapy is total surgical excision. The prognosis is favorable; however, further monitoring is required due to the possibility of recurrence and malignant change.

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