Case Report

A Rare Case of Retroperitoneal Schwannoma: A Case Report

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ABSTRACT

Abdominal schwannomas are a very rare entity and retroperitoneal schwannomas even more so. Here, we present the case of a 59-year-old male. The patient presented with undiagnosed intermittent pain abdomen. Workup and imaging revealed a well-defined solid cystic mass in the retroperitoneal lumbar region compressing the inferior vena cava. There was no evidence of disseminated disease. Laparotomy was done and the mass was excised. Histological examination revealed a benign schwannoma. The patient had an uneventful post-operative period and was discharged and is currently on follow-up.

Key words: Neurogenic tumor, Retroperitoneal malignancy, Schwannoma

Primary tumors of the retroperitoneal region are quite rare, and retroperitoneal schwannomas even more so. Schwannomas originate from Schwann cells of the peripheral nerve fibers and are usually located in the head, neck, and flexor aspects of the extremities [1]. The majority of retroperitoneal schwannomas are benign in nature although malignant ones have also been reported [2]. The rarity of a retroperitoneal schwannoma merits its recording for the benefit of clinicians to consider this as a differential while assessment of a retroperitoneal mass. It is important to exclude a malignant retroperitoneal tumor to choose the correct treatment.

CASE REPORT

A 59-year-old male came to our hospital with complaints of intermittent pain in the right upper abdomen for the past 2 months. The pain had no aggravating or relieving factors and was non-radiating in nature. There was no history of fever, vomiting, jaundice, bleeding per rectum, melena, hematemesis, or hematuria. The patient was a diabetic for the past 16 years and hypertensive for the past 10 years which was well-controlled with medications. The patient had no addictions or allergies. Bowel and bladder habits were normal.

The general survey was unremarkable with normal vitals. Examination of the abdomen revealed a palpable mass of 6 × 5 cm in the right lumbar region extending to the right iliac fossa. The lump was retroperitoneal in origin, firm, non-tender with a smooth surface and ill-defined margins, immobile, non-ballotable, and not moving with the respiration. There was no evidence of free fluid in the abdomen. Examination of the genitals and digital rectal examination was normal. Other systemic examinations were normal.

Biochemical analysis revealed hemoglobin to be 11 gm%, total leucocyte count was 6,500/mm^3, platelets were 2,45,000/dl, prothrombin time was 13 s, international normalized ratio (INR) was 1.08, urea was 35 mg/dl, creatinine was 0.8 mg/dl, total bilirubin was 0.9 mg/dl, serum glutamic oxaloacetic transaminase (SGOT) was 64 U/L, serum glutamic pyruvic transaminase (SGPT) was 56 U/L, and alkaline phosphatase was 98 U/L. Urine analysis was normal.

Chest X-ray revealed no abnormalities. Ultrasound (USG) of the abdomen showed a distended gallbladder with a solitary 13 mm calculus in the lumen. Common Bile Duct was 4 mm. There was no other abnormality. No comment was made regarding the retroperitoneal mass. Computed tomography (CT) of the abdomen showed a normal liver with distended gall bladder with 14 mm calculus in the lumen. The retroperitoneum contained a well-defined, smooth marginated, solid cystic mass lesion measuring about 4.5 × 4.2 × 5.1 cm seen in the right paravertebral location of the lumbar region with mild post-contrast enhancement of solid component. The lesion shows a mass effect with the displacement of inferior vena cava (IVC) with luminal narrowing and no calcification of fat components. These were suggestive of a neurogenic tumor.

Due to the diagnostic dilemma, magnetic resonance imaging (MRI) was done. MRI of the abdomen revealed mixed cystic and solid lesion of size 4.5 × 3.7 × 4.3 cm in the retroperitoneum inseparable from the IVC wall, suggestive of either a

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leiomyoma or neurogenic tumor. There was no retroperitoneal lymphadenopathy. To rule out the metastasis, we did a positron emission tomography-CT (PET-CT) scan. This showed a globular mixed density lesion of size 3.77 × 3.95 × 5.42 cm with well-defined margins in the retroperitoneal region at the right paracaval area with compression of IVC showing heterogeneous fluorodeoxyglucose (FDG) uptake.

After the workup and preparation of the patient, a diagnostic laparotomy was performed. On mobilizing the right colon, there was a 5 × 3 cm retroperitoneal growth in the right lumbar region abutting the IVC and appeared to be arising from the sympathetic trunk. The growth was dissected and excised preserving the surrounding structures and cholecystectomy was done (Figs. 1 and 2).

Histopathology of multiple sections studied from retroperitoneal mass showed a benign lesion with features of Schwannoma. They showed the typical Antoni A and Antoni B appearance. No evidence of malignancy is seen. The final diagnosis was thus confirmed as a retroperitoneal benign schwannoma based on the features in histology. The post-operative period was uneventful and the patient was subsequently discharged. The patient is currently on follow-up.

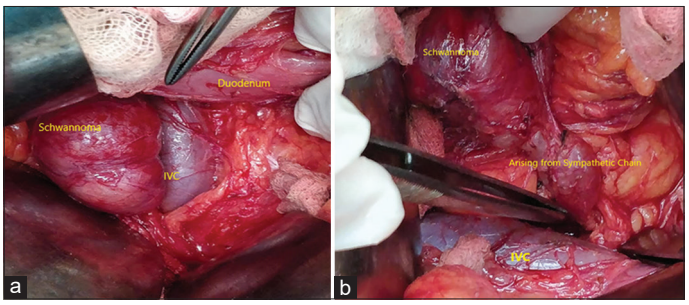

Figure 1: Intraoperative picture showing the (a) schwannoma and its relations; (b) schwannoma arising from the sympathetic chain

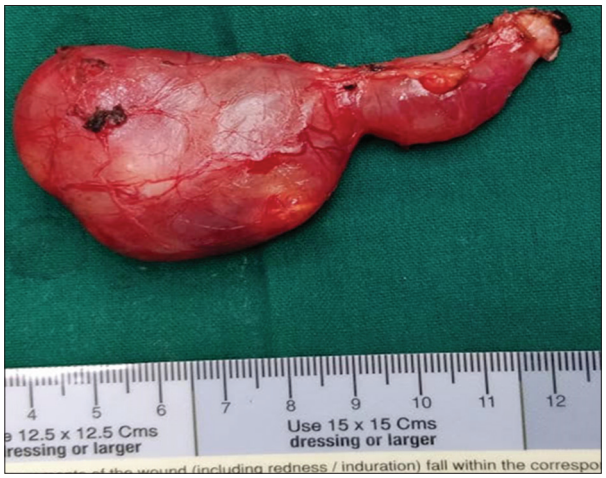

Figure 2: Post-operative specimen

DISCUSSION

Schwannomas are neurogenic tumors that are developed from the Schwann cells of nerves except for cranial nerves I and II, where Schwann cells are absent. They are commonly seen in middle-aged females [3]. The most frequent areas are the cephalocervical region (44.8%) and the limbs (32.6%). The benign tumor is more frequent in the spinal cord (20–30%). Visceral areas for this tumor include the stomach, liver, pancreas, kidney, heart, and brain [4,5]. It may be associated with Cushing syndrome and Von Recklinghausen's disease (5–8%) [6].

Retroperitoneal schwannoma represents 0.3–5% of all primary schwannomas and 1–4% of all retroperitoneal tumors [3,7-9]. Retroperitoneal schwannomas are usually discovered incidentally because retroperitoneal space is flexible and the diagnosis is often delayed; therefore, the lesion reaches the significant size at the time of diagnosis. The symptoms are vague and non-specific, such as vague abdominal pain and dull ache [10,11]. Atypical presentations are very rare and include flank pain, hematuria, headache, secondary hypertension, and recurrent renal colic pain [10]. The hallmark pattern of the benign variants in histopathology is an alternation of Antoni A and B areas, with a diffuse positivity for S100 protein in the cytoplasm of the tumor cells [10,11].

In general, MRI is regarded as the diagnostic modality of choice in the evaluation of retroperitoneal tumors. There are a few well-known imaging characters for schwannomas which are mainly "target sign" and "fascicular sign." The "fascicular sign" stands for the appearance of bundles which are a general property of neurogenic tumors. On the other hand, the "target sign" is the presence of hypointense center and hyperintense periphery on T2-weighted MRI. The lesion in this manuscript did not exert any of the above-mentioned typical diagnostic signs. Hence, we believe that for the pre-operative diagnosis of retroperitoneal schwannomas, a high index of suspicion is mandatory, especially in the absence of characteristic imaging features, as in our patient.

The management of non-functional masses depends on the size of the lesion. The majority of lesions remain stable, 5–25% increase and 3–4% decrease in size. Watchful waiting is appropriate for lesions lower than 4 cm and appearing benign on imaging. Lesions greater than 4 cm should be excised. After surgical resection, some authors recommended follow-ups of 6, 12, and 18 months [3]. Curative treatments have a favorable prognosis. In asymptomatic cases or patients where surgery poses a high risk, radiologic surveillance or radiofrequency ablation could be an option of therapy [10]. Strauss *et al*. reported a study of eight patients undergoing a biopsy with conservative treatment. None of them have changed after 32 months of follow-up [8].

CONCLUSION

Retroperitoneal schwannomas are very rare and very few have been reported in the medical literature. These rarely present with symptoms and are mostly diagnosed incidentally. One must have very high suspicion to diagnose them pre-operatively. One may keep them as a differential diagnosis in cases of retroperitoneal mass. The investigation of choice is MRI. The treatment is surgical excision although selected cases can be put on follow-up.

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