



Retroperitoneal Schwannoma: A case Report and Review of the Literature.

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ABSTRACT

In clinical practice, retroperitoneal schwannoma is uncommonly seen. This is a rare benign retroperitoneal tumor. Often unexpected preoperatively, diagnosis is based on post-operative histopathologic examination of surgical specimen. At an incidence estimated at 4%, benign and malignant schwannomas represent about 0.6% and 1.8% respectively of schwannomas in the retroperitoneum region. Both sexes are equally affected and the patient's age is often between 30 and 60 years. Clinical signs are nonspecific. Patients present unexplained lumbar or abdominal pain due to compression of neighboring organs. Total tumor excision has a therapeutic effect and a good prognosis. The case of a 45-year-old female patient in whom was discovered incidentally at ultrasound a left prerenal tissue mass measuring about 6 cm is reported. Further imaging notably, computed tomography revealed this mass in juxtaposition though non-adherent to the left kidney, aorta and the renal pedicle. She underwent a total excision of the tumor by open surgery. Histopathology of the surgical specimen revealed retroperitoneal schwannoma. The patient was discharged five days after surgery with a good prognosis with a follow-up of 15 months.

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Introduction

Retroperitoneal primitive tumors in adults are rare. Schwannoma is a benign tumor developed from Schwann's nerve sheath. Retro-peritoneal schwannoma (RPS) is a rare form [1]. Diagnosis is often unexpected preoperatively, and is only confirmed after histological examination of the surgical specimen of the retroperitoneal mass.

We here-in report a clinical case of a 45-year-old female patient in whom was discovered incidentally at ultrasound a left prerenal tissue mass measuring about 6 cm. We equally review data reported in literature, in terms of epidemiological, diagnosis and therapeutic aspects of this rare pathology.

Case report

A 45-year-old female patient presented with a history of vague left flank pain. Physical examination was unremarkable. Haemogram and biochemical analysis were normal. Ultrasonography revealed a 6-cm heterogeneous expansive mass in left adrenal topography. A computed tomography scan confirmed a 6.5 x 5.0 x 4.3 cm enhancing heterogeneous mass with necrotic central areas arising from left adrenal topography. This mass was in juxtaposition though non adherent to the left kidney, aorta, the renal pedicle and the adrenal gland. Due to the location and signal characters, the presumptive diagnosis was a neurogenic or a fibrous tumor.

Surgery was done via a midline abdominal incision; the abdomen was explored. The mass was localized just above the right psoas muscle, lateral to the aorta and inferior to the upper pole of the left kidney, the mass was resected (Figure 1)

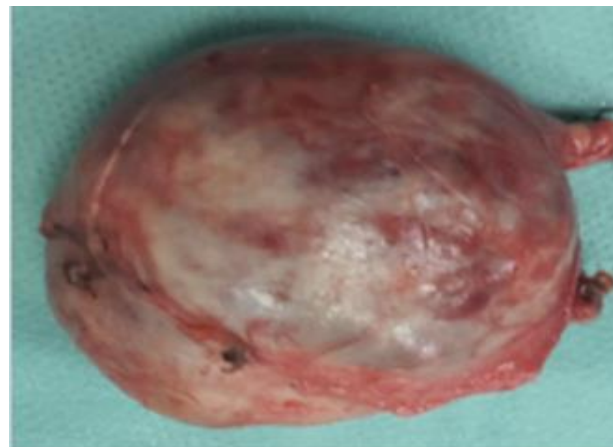


Figure 1. Resected retroperitoneal schwannoma surgical mass of about 6 cm in diameter.

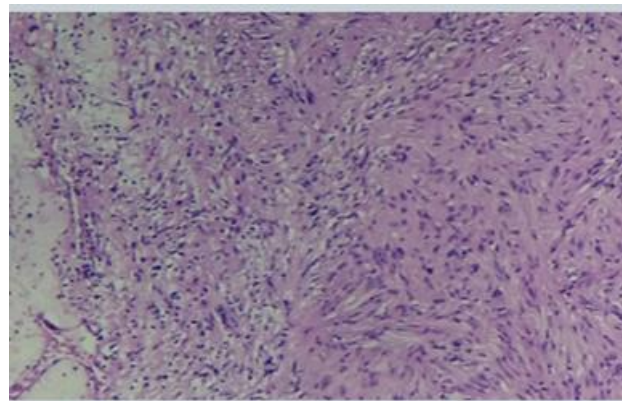


Figure 2. Neuroid differentiation zone: intersecting bundles with palisadic peripheral arrangement with cells that were positive for S100 protein on immunochemistry.

Macroscopic examination revealed a capsulated mass of 5 x 4 x 4.5 cm, with cystic and solid bright-yellow areas. On microscopic examination, the solid mass consisted of a proliferation of fusiform cells that formed a palisade pattern in some regions (Antoni type A). There were regions composed of gelatinous substance, myxoid and degenerative tissue with less cells (Antoni type B) (Figure 2). The cells were diffusely and strongly positive for S100 protein. The tumor was diagnosed as a schwannoma.

Postoperative period was uneventful. At the 15-month follow-up examination, the patient remained asymptomatic and there was no evidence of recurrence.

Discussion

Retroperitoneal schwannoma (RPS) (also termed neurilemmomas or neurinomas) is a rare entity comprising only 0.5% to 12% of all retroperitoneal tumors [2].

Most schwannomas are found in peripheral nerve fibers in the limbs, head, and neck. In the retroperitoneal region, they occur most commonly between 30 and 60 years of age, with a male/female ratio of 2:3 [3].

The retroperitoneal space is rather large and flexible, the diagnosis of RPS in the retroperitoneal region is difficult, and a large and deeply situated tumor is usually present before patients have any symptoms. The symptoms are vague and nonspecific, such as vague abdominal pain and distension [4, 5]. Atypical presentations are very rare and a variety of symptoms include flank pain and haematuria, portal thrombus, headache, and secondary hypertension and recurrent renal colic pain [4]. Evolution can be completely asymptomatic. Diagnosis is fortuitous or either during routine clinical or ultrasound examination [6].

Radiological studies are fundamental in the diagnostic evaluation. Computed tomography (CT) scans typically show well-defined low or mixed attenuation with cystic necrotic central areas. Cystic changes occur more commonly in RPS (up to 66%) than in other retroperitoneal tumors [7]. Other degenerative changes, such as calcification, haemorrhage, and hyalinization, can also be present.

Magnetic resonance imaging (MRI) findings of schwannomas have been reported as masses of low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. These findings are characteristic but not specific of schwannomas and have been reported present in only 57% of the cases in previous studies [8], hindering the correct diagnosis.

Additionally, the signal intensity on T2-weighted images may vary depending on cell density. Tumors with microscopic findings of hypercellular Antoni type A tissue have intermediate signals, while tumors with Antoni type B tissue have a bright signal on T2-weighted images. Additionally, other neurogenic tumors such as ganglioneuroma, appear more commonly in the retroperitoneum than schwannomas and have similar findings on CT and MRI [9]. Therefore, misdiagnosis of retroperitoneal schwannomas is not uncommon and preoperative diagnosis as hepatic tumors, pancreatic cystic tumors and psoas abscess have been reported [9].

Histologically, schwannomas consist of compact cellular lesions (Antoni type A tissue) and loose hypocellular myxoid lesions with microcystic spaces (Antoni type B tissue) [10, 11]. On gross appearance, schwannomas are usually solitary, well circumscribed, firm, smooth-surfaced tumors. The hallmark pattern of the benign variants is an alternation of the Antoni A and B areas, with a diffuse positivity for S100 protein in the cytoplasm of the tumor cells.

Malignant degeneration of schwannomas is extremely rare but when present, they act as high-grade sarcomas with a high likelihood of producing local recurrence and distant metastasis. The diagnosis of malignant peripheral nerve sheath tumor lacks standardized diagnostic criteria but features dense fascicles in a "marble-like" pattern consisting of asymmetrically tapered spindle cells. Malignancy is usually suggested histologically by mitosis, pleomorphism, and blood vessel infiltration [12].

The differential diagnosis include benign cystic lymphangioma, which is a combination of solid and cystic mixed mass associated with fatty plaques. Other neurogenic tumors such as paraganglioma and pheochromocytoma as well as liposarcoma and malignant fibrous histiocytoma should also be considered. In addition, if the retroperitoneal schwannoma contains considerable amount of cystic degeneration, retroperitoneal cystic masses such as hematoma should also be included in the diagnostic checklist.

CT-guided core biopsy and fine needle aspiration have been found to be unreliable for the diagnosis of RPS [2]. They may be helpful only if the sample contains enough Schwann cells to visualize microscopically. However, in areas of degeneration, the cellular pleomorphism can hinder the diagnosis and degenerative cells may be misinterpreted as malignancy. One also runs the risk of haemorrhage, infection, and tumor seeding; thus, many authors do not recommend CT-guided biopsy [5,11].

Surgical resection therefore is the only accurate approach for pathologic evaluation to enable diagnosis of retroperitoneal schwannoma. Recent advances in laparoscopic instruments and skills have made laparoscopy an excellent approach for biopsy and even surgical resection of benign retroperitoneal tumors, aiding the diagnosis of these lesions. Even though the best management of retroperitoneal neural sheath tumors is complete excision, considerable controversy exists over negative soft tissue margins [5]. It is argued that complete surgical excision is necessary and may include sacrifice of adjacent tissue and viscera though others believe that because this is a benign mass, a simple enucleation or partial excision of the tumor is sufficient and have reported no increase in the size of schwannoma during a 6 and 14-year period [13]. The argument here is that the morbidity associated with resection of adjacent tissue would not be justified in the treatment of a benign lesion [13].

Complete surgical resection so as to attain negative margins is recommended [5,14] because malignancy cannot be excluded accurately preoperatively or even intra operatively with frozen section analysis. This complete excision is the only valid treatment for schwannomas; since schwannomas are not sensitive to radiotherapy and chemotherapy [15].

In our case, the retroperitoneal mass was easily isolated from adjacent structures, and the lesion was completely resected with negative margins.

Most schwannomas are benign. Prognosis is extremely good, though malignant transformations have been reported in 5% to 18% of cases [16], usually associated with von Recklinghausen's disease.

A few cases of metastases such as hepatic, pulmonary, bone and subcutaneous tissue [17] that occurred after resection of a histologically benign schwannoma have been reported. Recurrence of malignant schwannoma after resection of a benign schwannoma has also been reported [18].

In view of recurrence or malignant transformation, it is recommended that careful monitoring by imaging is necessary after removal of benign retroperitoneal schwannomas.

Conclusion

Retroperitoneal schwannoma is a tumor of good prognosis in its benign form. There is often late diagnosis due to nonspecific clinical symptoms and imaging is less contributive though essential in exploring the retroperitoneal region. Diagnosis is based on post-operative histopathologic examination and immunohistochemistry of surgical specimen. Complete surgical excision is the mainstay treatment. Recurrence although rare after surgery imposes post-operative monitoring by CT scan.

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