



## Oncology

## Retroperitoneal Schwannoma: Case report

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## ABSTRACT

Schwannoma is a rare benign tumor that comes from the SCHWANN cells that sheath the peripheral nerves. Retroperitoneal location is even rarer than the frequency of this neoplasia. The diagnosis is immunohistochemical and the treatment is surgical. We report the case of a 36-year-old patient with abdominal pain and transit disorders for 4 months. An abdominal CT scan was found a rounded tissue mass of 10 cm long with well-defined borders. The patient was prepared for surgical removal. Per-operatively, a mass was found in the sub hepatic area, pushing the right kidney backwards. Anatomic-pathological examination was in favor of Schwannoma.

## Introduction

Schwannomas are rare ectodermal benign tumors that arise from Schwann cells, which cover the axons of peripheral nerves. The occurrence of retroperitoneal schwannoma is very rare, this location accounts for 1–3% of all schwannomas and 1% of retroperitoneal neoplasms.<sup>1</sup> The uni-focal and sporadic form is the most frequent; in its multifocal and familial form, it's associated with Von Recklinghausen's disease.

Retroperitoneal Schwannoma is most often asymptomatic and complete surgical removal with safe limits is the standard treatment, recurrences are rarely reported.

## Case report

Mr A.G., 36 years old, reports since 4 months the progressive installation of a feeling of abdominal heaviness associated with a pulsatile pain and transit disorders. The clinical examination found a patient in good condition, correct vital constants and a rounded mass of the right hypochondrium. The rest of the physical examination was without abnormalities, in particular without skin signs in favor of Von Recklinghausen's disease.

An abdominal CT scan was performed, revealing a large tumor mass measuring 10 cm, with well-defined contours, rising heterogeneously after injection of contrast agent, located in the sub-hepatic region, outside the spine and large vessels, pushing the right kidney up and back

(Fig. 1).

Surgery was performed through a right sub costal approach, allowing the removal of a sub hepatic well encapsulated mass. Macroscopically, the mass was nodular and well capsulated, measuring 10 × 7x5 cm and weighing 121 g with myxoid areas of soft consistency and white areas of firm consistency (Fig. 2).

Histopathological examination showed areas with variable density, sometimes myxoid with few cells, sometimes dense. The cells are arranged in short bundles, they are spindle-shaped with a wavy nucleus and pale cytoplasm. The immunohistochemical study shows that tumor cells express high levels of S-100 protein (Fig. 3).

The post-operational follow-up was simple, and the hospitalization length was 72 hours. No recurrence was detected on the control scans at 3 and 12 months postoperatively.

## Discussion

Schwannomas or neurilemmomas are rare benign tumors that develop from Schwann cells of the cranial, peripheral and autonomic nerves. In a review of 303 patients with Schwannomas, all locations included, only 2 (0.7%) were located in the retroperitoneal area. Retroperitoneal Schwannoma mainly affects patients between 20 and 50 years old with a female predominance.<sup>1,2</sup>

In general, the retroperitoneal space is wide and flexible, which makes the diagnosis of Retroperitoneal Schwannoma late and difficult

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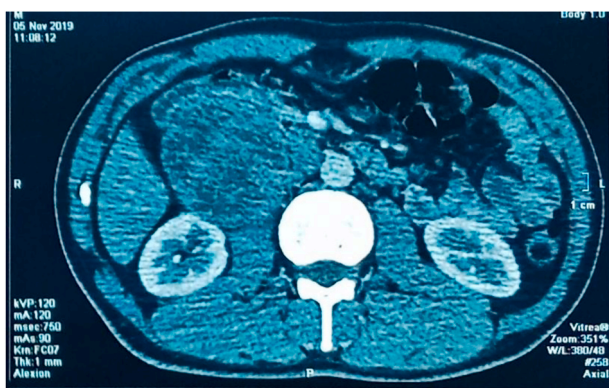


Fig. 1. Cross-section showing a heterogeneous retroperitoneal mass.

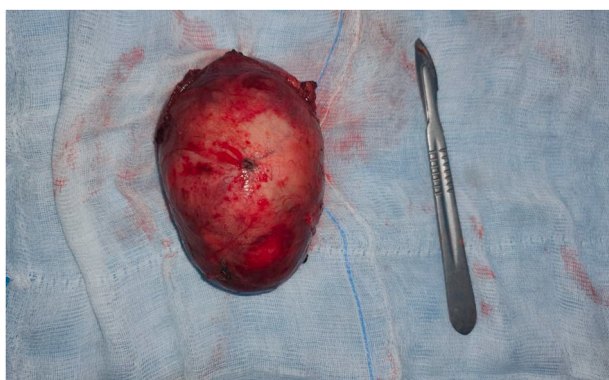


Fig. 2. Image of the mass after its removal.

preoperatively because none of the clinical signs are pathognomonic to this tumor. Benign Schwannomas do not invade nearby organs, the symptoms are due to the compression and displacement of the retroperitoneal structures. In a series of 82 patients with Retroperitoneal Schwannoma, the majority of patients had abdominal distension with no other associated signs (30.5%) or the diagnosis was incidental (34.1%), with the remaining patients having abdominal pain (20.7%), low back pain (6.1%) or digestive problems (6.1%).<sup>3</sup> However, cases with secondary hypertension, hematuria or renal colic have been reported. The clinical examination is generally poor, it can rarely detect a solid retroperitoneum-fixed mass.

Usually, the diagnosis is usually suspected in pre-operative care thanks to radiological diagnostic means. Ultrasonography is a reproducible, inexpensive and operator-dependent tool for diagnosis. It can reveal a semi-solid mass or a mass with cystic regions. The scanner generally shows low attenuation or well-defined mixed attenuation with cystic and necrotic core zones.<sup>2</sup> MRI is considered the first choice imaging tool for the exploration of retroperitoneal tumors, as it allows a better visualization of the origin of the lesion, its extent, its internal and vascular architecture and the involvement of other organs. Retroperitoneal schwannoma appears as a tumor with regular borders that are isointense with the muscle in T1, T2-weighted sequences show areas of low intensity that correspond to Antoni A areas and areas with excessively high intensity that correspond to Antoni B areas.<sup>4</sup>

The diagnosis of certainty can therefore only be made on histology. Biopsies are discussed on a case-by-case basis due to the risk of hemorrhage, infection and the risk of tumor spreading. Surgical excision is recommended in consideration of the possible histological heterogeneity of retroperitoneal tumors. Histologically, retroperitoneal Schwannomas are encapsulated and present 2 distinct areas: Antoni A zones which are compact cellular regions in the form of a palisade or turbine; Antoni B zones which are loose, hypocellular, myxoid regions with sometimes the presence of a gelatinous substance. Immunohistochemically, retroperitoneal Schwannoma is typically positive for the marker S-100, vimentin and NSE (Neuron-specific enolase). It responds to negative labelling for desmin, SMA (Smooth muscle actin), HHF35, CD34 and CD117.<sup>5</sup>

Retroperitoneal Schwannoma can degenerate to malignancy and present radiological and histological changes, it's characterized by the appearance of necrosis, stromal edema, fibrosis, perivascular hyalinization, calcifications, as well as the presence of nuclear degeneration with pleomorphism, lobulation and hyperchromatosis.<sup>2</sup>

The standard treatment is complete surgical removal of the tumor since Schwannomas don't respond to radiotherapy or chemotherapy. However, it may have an intimate contact with vessels or noble organs, at this time an incomplete removal can be performed. Other authors have described 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.

## Conclusion

Retroperitoneal Schwannoma is a rare tumor, which is difficult to diagnose preoperatively due to a borrowed symptomatology and little contributive imaging. Its diagnosis is often late. Complete resection is the standard treatment. Recurrences, although rare, require long-term monitoring by CT scan.

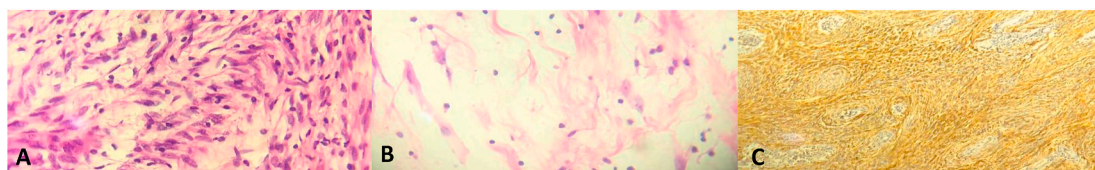


Fig. 3. Histological study of the tumoral mass showing dense areas (A), loose areas (B) responding positively to SP-100 marker(C).

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