

# Giant posterior pararenal schwannoma: A case report and review of literature

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**Abstract.** Schwannomas, arising from Schwann cells of peripheral nerve sheaths, are primarily benign tumors. They are rarely found in the retroperitoneal space. To date, ~30 cases on giant retroperitoneal schwannomas have been reported. Those with a location of pararenal space are even rarer. Clinically, they are often misdiagnosed as malignancies. In the present study, a case of a 35-year-old woman with a giant schwannoma measuring 13 x 8.5 x 6.5 cm in the posterior pararenal space of left retroperitoneum was presented, which was thought to be a malignant tumor from the result of computed tomography scan. Later postoperative pathology and immunohistochemistry were consistent with benign schwannoma. To the best of our knowledge, this was the largest posterior pararenal schwannoma reported in literature. It is significant to report this case, and summarize the characteristics, diagnosis, treatment and prognosis of the similar cases. Furthermore, a brief review of the previously published cases in literature is provided.

## Introduction

Schwannomas are mostly benign tumors. They are often seen in the head, neck and extremities. Studies have shown that only 0.7 to 2.6% of schwannomas are found in the retroperitoneum.

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Those in pararenal space are even rarer. Although most schwannomas are <5 cm in diameter, retroperitoneal schwannomas are able to grow into a large size in such a non-restrictive space (1). They are mostly asymptomatic until they grow too large to cause compression, which is usually ambiguous pain. In the present study we introduce a case of a 35-year-old female with a giant posterior pararenal schwannoma. To the best of our knowledge, this is the first time to report a posterior pararenal schwannoma in such a large size.

## Materials and methods

The surgical specimen were fixed in 10% formalin at room temperature overnight and embedded in paraffin. Later, the specimen were cut into four-micrometer-thick sections. Before staining, the sections were dewaxed in xylene, rehydrated through decreasing concentrations of ethanol, and washed in PBS. After that, the sections were stained with hematoxylin at 37°C for 15 min, rinsed under running water for 1 min and then left standing in water for 5min. Similarly, Staining with 0.5% eosin was executed for at 37°C for 3 min, followed by rinsing under running water. After staining, sections were dehydrated through increasing concentrations of ethanol and xylene. Rehydration was performed by continuous immersion in xylene, graded concentrations of ethanol, and tap water. Slides were kept in citrate buffer (pH 9.0) and two cycles of heat retrieval were performed in oven at 99°C for ten and five minutes.

Slides were washed in Tris buffer (pH 7.8). To blocking endogenous peroxidase, all sections were incubated with hydrogen peroxide for 10 min. Next, sections were washed thrice in Tris buffer, followed by 30 min incubation with primary antibodies. Primary antibodies used were S100 protein (cat. no. PIPA516257; rabbit polyclonal antibody, dilution 1:300), CD34 (cat. no. PIMA516924; mouse monoclonal antibody; QBend/10; dilution 1:100), SMA (cat. no. PIMA511547; mouse monoclonal antibody; clone 1A4; dilution 1:200), and Ki-67 (mouse monoclonal antibody; clone 4A1) (all from Invitrogen, Waltham, MA, USA). Secondary antibody including goat anti-mouse IgG (cat. no. PIA32723; dilution 1:500) and goat

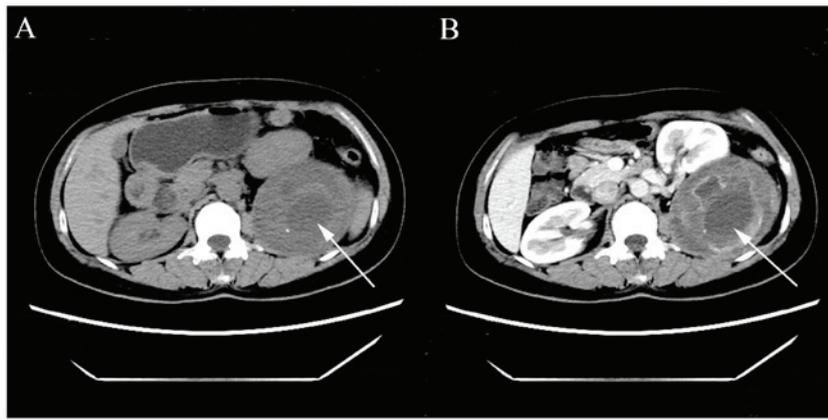


Figure 1. (A) Non-enhanced axial computed tomography image revealing a 89 x 81 x 104 mm, 13-48 HU, well-demarcated, cystic solid mass with mixed density in the left posterior pararenal space (arrows), adhering with adjacent organs. The left kidney was pushed to inferior. (B) Contrast-enhanced axial computerized tomography image showing apparent heterogeneous contrast enhancement inside the mass. Arrows indicate the location of the giant schwannoma.

anti-rabbit IgG (cat. no. A11034; dilution 1:500) (both from Invitrogen) was added correspondingly after washing with Tris buffer for 40 min. At last, chromogen diaminobenzidine (DAB) was added for 10 min, followed by counterstaining with hematoxylin for 2 min, sequential immersions in xylene and alcohol and mounting with distyrene plastisizer xylene (DPX). The results were observed under light microscope.

### Case report

A 35-year-old female was admitted to Peking University Shenzhen Hospital (Shenzhen, China) with an incidental finding of a left retroperitoneal mass 1 month earlier. Except for dull pain in left waist, no other symptoms showed up. On physical examination, a large, firm, smooth, non-tender mass was palpable. Ultrasound examination revealed a predominant solid mass measuring 109 x 89 x 86 mm between the left kidney and spleen, with multiple oval dark areas. Small amount of blood flow could be seen. CT scan revealed a giant cystic solid mass with mixed density in the left posterior pararenal space, adhering with pancreas, spleen, kidney and psoas major muscle. The left kidney was pushed to inferior with vascular circuitry. The lesion was predominantly low density, 13-48 HU, well-demarcated, smooth with capsule, and measured 89 x 81 x 104 mm with punctiform calcification inside it. On enhanced CT, apparent heterogeneous contrast enhancement could be seen inside the mass. The left kidney and adrenal gland had a normal morphology and uniform density, showing no abnormality on enhanced CT. No swollen lymph nodes were found (Fig. 1). According to the results above, adrenal neoplasms needed to be distinguished, including pheochromocytoma, cortical adenoma, and aldosteronoma. The blood pressure of the patient was in normal range. Later hematologic examination showed that the secretion of ACTH and cortisol (at OMN, 8 a.m. and 4 p. m.) had a normal circadian rhythm and was within normal range. Catecholamine, methoxy adrenalin, quantitative test of 24 h VMA (vanilmandelic acid) were within normal limits. Angiotensin, renin, ALD (aldosterone) in blood showed no abnormalities in both erect position and clinostatism. Basically, adrenal neoplasms could be ruled out. However, a preoperative diagnosis was still challenging.

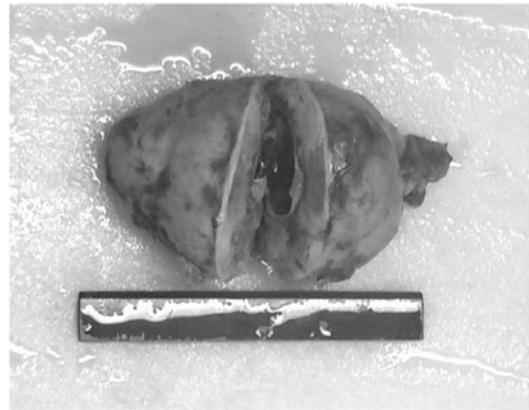


Figure 2. Gross examination showing a cystic solid mass measured 13 x 8.5 x 6.5 cm with intact smooth capsule.

Later, the patient underwent surgery. Intraoperatively, a cystic solid oval mass was found behind the kidney in the left retroperitoneum, with obvious adhesion with adjacent tissue. No obvious change in blood pressure of the patient was seen when touching the mass. The mass has an intact capsule and a complete resection was executed.

Gross examination showed a solid mass measuring 13 x 8.5 x 6.5 cm, with cystic degeneration (4.0 cm in diameter). It was wrapped with an intact smooth capsule (Fig. 2). On microscopy the mass was composed of spindle cells, with regions of high and low cellularity (Antoni A and B areas, respectively). Hemorrhagic and cystic degeneration could be seen in central area (Fig. 3).

Immunohistochemical staining revealed positivity for S100 and negative for CD34 and SMA. The Ki-67 proliferation index was about 10%. Based on these, a diagnosis of schwannoma was suggested. During the follow-up, there was no evidence of recurrence.

### Discussion

Schwannomas, arising from Schwann cells of peripheral nerve sheaths, are mostly benign tumors. They are often found in

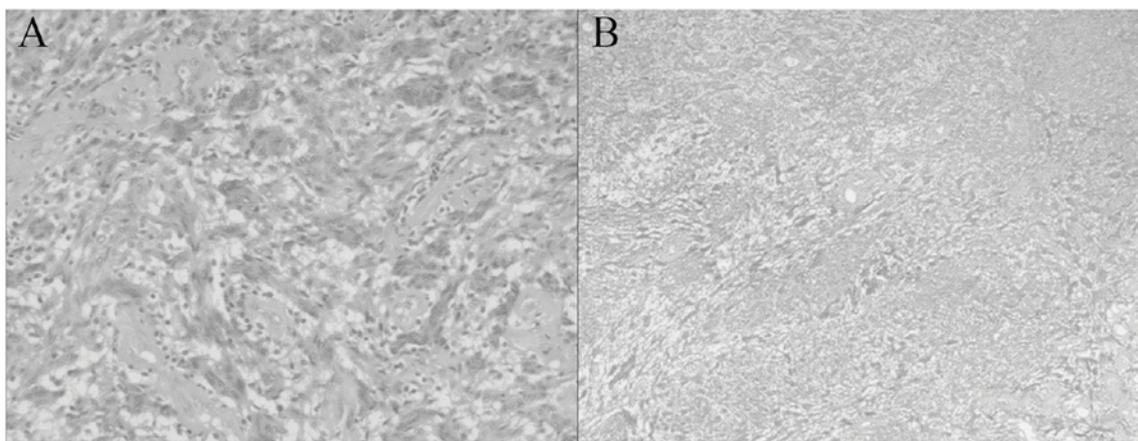


Figure 3. On microscopy the mass was composed of spindle cells, with regions of (A) high and (B) low cellularity. Hemorrhagic and cystic degeneration could be seen in central area. Magnification, x200.

the head, neck and extremities in the 4th and 6th decades of life (2) Studies have shown that only 0.7-2.6% of schwannomas are found in the retroperitoneum. Generally, schwannomas are less than 5cm in diameter. But retroperitoneum is a non-restrictive space which allows the tumors to reach a large size over a long time of growth (1). According to our search on Pubmed, we found out about 30 cases on giant retroperitoneal schwannoma. Yet, there have never reported a posterior pararenal schwannoma larger than 10cm in diameter.

According to a literature published by G. Lannaci in 2016, twenty-one cases of primary renal schwannoma have been reported in literature (3). Schwannomas originating in the pararenal space are extremely rare. To date, only a few cases have been reported. Miyagi T reported a posterior pararenal schwannoma with a diameter of 3.0 x 2.5 cm in 1986 (4). In 2010, a patient suffering from Neurofibromatosis Type 2 found a schwannoma posterior and inferior to the left kidney, measuring 9.5 x 4 x 4 cm (5). In 2013, Liu *et al* mentioned eight patients with retroperitoneal schwannomas in the anterior pararenal space (6). Here in, we show a giant posterior pararenal schwannoma measuring 13 x 8.5 x 6.5 cm, which we believe to be the biggest one that has been reported in this location by far. This is the first time to report a pararenal schwannoma with a size more than 10 cm in diameter.

Schwannomas are mostly well-demarcated round or oval masses on CT and MRI. Cystic and hemorrhagic degeneration can be seen in large retroperitoneal schwannomas, which appear as inhomogeneous low-density masses and homogeneous to heterogeneous contrast enhancement on enhanced CT. MRI images can reminder the origin and the exact location of the mass. The intensity of the masses can help with the inference of their properties. However, these changes are non-specific. We are not able to make a final diagnosis through these radiographic results.

Blood biochemical examination is conducive to exclude adrenal neoplasms. The high concentration of certain hormones in blood can reveal the existence of functional adenoma. However, for non-functional adenoma, the hormones in blood could be normal, which means that blood biochemical examination only plays a limited role in diagnosis. Fine-needle aspiration biopsy is a way to get the pathological diagnosis

of retroperitoneal masses. Yet it is suffered from controversy due to its low accuracy and potential complications. Still, preoperative diagnosis is challenging and a surgical removal is required for diagnosis.

For retroperitoneal masses larger than 4 cm, especially those with clinical symptoms, surgical removal is regarded as the most rational way, with the benefit of both diagnosis and therapy. An incomplete excision may increase 5-10% of local recurrence (7). Moreover the possibility of malignancy is considerable. Therefore, a complete excision is necessary.

To make a final diagnosis, we need to combine pathology and immunohistochemistry. Histologically, there are two growth patterns in schwannomas. The Antony A pattern is characterized by a cell-rich structure with cell nuclei running parallel to each other while the Antony B pattern has a loose distribution of the cells with varying degrees of myxoid and hyaline degeneration. Immunohistochemically, S100 is an important marker for the diagnosis of schwannomas (8).

The prognosis of retroperitoneal schwannomas are mostly good. The most common complication is recurrence, possibly caused by incomplete excision. Malignant schwannomas has poor prognosis and a high rate of recurrence though the malignant transformation of retroperitoneal schwannomas appears to be extremely rare (9).

In the present study, we introduced a giant posterior pararenal schwannoma which was believed to be the largest one reported in this location by far. There is a dilemma on the preoperative diagnosis of pararenal schwannoma. It's important to include schwannomas during the differentiation of pararenal masses, especially for those with cystic and hemorrhagic degeneration since the treatment and prognosis among them may differ a lot.

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#### Availability of data and materials

The datasets used during the current study are available from the corresponding author on reasonable request.

#### Authors' contributions

XP and ZL performed data collection, interpretation and drafted the manuscript. LiaZ and LiwZ contributed to the study design and acquisition of data. XW, YY, SY and YC participated in the study design, data collection, analysis of data and follow-up of the patient. YL contributed to the study design, and the analysis and interpretation of data.

#### Ethics approval and consent to participate

Written informed consent to participate was obtained from the patient.

#### Patient consent for publication

Consent for publication of any associated data and accompanying images was obtained from the patient.

#### Competing interests

The authors declare that they have no competing interests.

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