

# Large retroperitoneal schwannoma: a rare cause of chronic back pain

Journal of International Medical Research

2018, Vol. 46(8) 3404–3410

© The Author(s) 2018

Article reuse guidelines:

[sagepub.com/journals-permissions](http://sagepub.com/journals-permissions)

DOI: 10.1177/0300060518776474

[journals.sagepub.com/home/imr](http://journals.sagepub.com/home/imr)



Milan Radojkovic<sup>1</sup>, Dragan Mihailovic<sup>2</sup>,  
Miroslav Stojanovic<sup>1</sup> and Danijela Radojković<sup>3</sup>

## Abstract

Schwannomas are tumours that arise from Schwann cells of the peripheral nerve sheath and rarely occur in the retroperitoneum. We report a 45-year-old woman who presented with a 2-year history of continuous progressive right-sided lower back and dull flank pain radiating into her posterolateral thigh. Abdominal magnetic resonance imaging showed a homogenous soft-tissue tumour with thick capsular lining, which lay in the right retroperitoneum. The tumour was removed at surgery. A histological examination confirmed the diagnosis of benign encapsulated cellular schwannoma. Complete tumour excision should be regarded as the treatment of choice for benign retroperitoneal schwannomas. Successful treatment of these tumours requires thorough preoperative planning and a multidisciplinary approach.

## Keywords

Benign retroperitoneal schwannoma, lower back pain, tumour, encapsulated mass, magnetic resonance imaging, histological examination, S100 protein

Date received: 12 February 2018; accepted: 21 April 2018

## Introduction

Schwannomas (neurilemmomas) are soft-tissue tumours that arise from Schwann cells of the peripheral nerve sheath and predominantly occur in women aged between their 2nd and 5th decades. These tumours are typically solitary and encapsulated, and are slow growing and noninvasive. Although schwannomas can occur

<sup>1</sup>Surgery Department, University of Nis Medical School, Bul. Dr Z. Djindjica 81, Serbia

<sup>2</sup>Pathology Department, University of Nis Medical School, Bul. Dr Z. Djindjica 81, Serbia

<sup>3</sup>Internal Medicine Department, University of Nis Medical School, Bul. Dr Z. Djindjica 81, Serbia

### Corresponding author:

Milan Radojkovic, Surgery Clinic, Clinical Center, Bul. Dr Zorana Djindjica 48, Ul. Sestara Bakovic 14/28, 18000 Nis, Serbia.

Email: [mida71@mts.rs](mailto:mida71@mts.rs)



anywhere in neural tissue where Schwann cells are present, they are usually found in the head, neck, and flexor surfaces of the extremities, and rarely found in the posterior mediastinum.<sup>1</sup> Schwannomas are not commonly larger than 5 to 6 cm in diameter. The histological nature of schwannomas is determined by the presence of high and low cellularity areas (Antoni A and B tissue, respectively).<sup>2</sup> The majority of schwannomas are benign, although malignant tumours can occur usually in the presence of Von Recklinghausen's disease (in up to 60% of cases).<sup>3</sup>

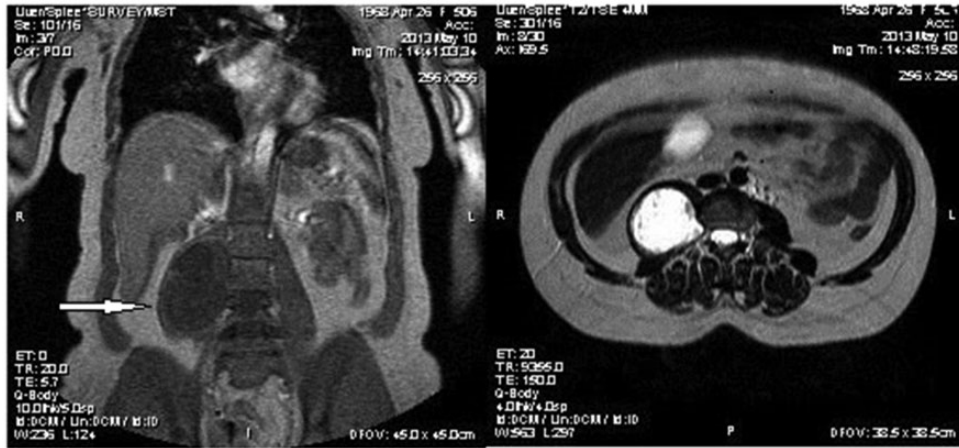
Schwannomas rarely occur in the retroperitoneum, and these comprise 1% to 10% of all primary retroperitoneal tumours.<sup>4</sup> Among schwannomas, only approximately 3% (0.7% are benign and 1.7% are malignant tumours) are reported to be located in the retroperitoneum.<sup>4,5</sup> Retroperitoneal schwannomas (RSs) are rare and may grow to a large size, displacing and rarely invading adjacent structures. Therefore, RSs are most often clinically "silent." RSs are incidentally found during imaging, until they enlarge to a size capable of compressing surrounding organs (mass effect) when presenting with vague, non-specific symptoms. Imaging modalities for RSs include abdominal ultrasound, computed tomography (CT), and magnetic resonance imaging (MRI). RSs present as well-defined, encapsulated masses, possibly with cystic degeneration and central necrosis. Percutaneous biopsy and a histopathological examination may be useful. However, these procedures are not always feasible and conclusive because of the risk of misinterpretation of cellular pleomorphism in degenerated areas, harvesting insufficient amount of tissue, and bleeding from vessels involved.<sup>2,6</sup> Additionally, there is a radiological resemblance between schwannoma and some tumours in which preoperative tissue sampling is discouraged, such as gastrointestinal stromal tumours.<sup>7</sup> Considering that

resection is the primary treatment option, obtaining preoperative histopathological diagnosis is not mandatory for an RS.

We report here a patient with a large, benign schwannoma in a rare location who presented with chronic back pain. We also discuss diagnosis and treatment strategies for this uncommon condition with a review of the literature.

## Case report

A 45-year-old woman presented to our centre with a 2-year history of continuous progressive right-sided lower back and dull lateral abdominal pain radiating to her posterolateral thigh. The pain was independent of activities and poorly responded to nonsteroidal anti-inflammatory drugs and physiotherapy. A physical examination showed a painless palpable mass in the right paraumbilical and lower abdominal region without peripheral neurovascular deficits. Laboratory tests were normal, except for an unspecific, mildly elevated erythrocyte sedimentation rate. Abdominal ultrasound showed a large, encapsulated, oval soft-tissue mass, which was 21 × 16 cm in diameter in the right retroperitoneum. Subsequent abdominal MRI confirmed a homogenous soft-tissue tumour with a thick, smooth, capsular lining. This tumour lay in the right retroperitoneum paravertebrally and displaced the iliopsoas muscle distally and the right kidney cranio-laterally, without any signs of ipsilateral hydronephrosis (Figure 1). Although the tumour was large, it was considered benign because it was not heterogenous, had no irregular margins, and there were no signs of adjacent organ involvement. Management options were discussed at a staff meeting. We decided not to perform a percutaneous biopsy because of the difficult position of the tumour. Symptom-relieving surgery was decided as the best choice of treatment.



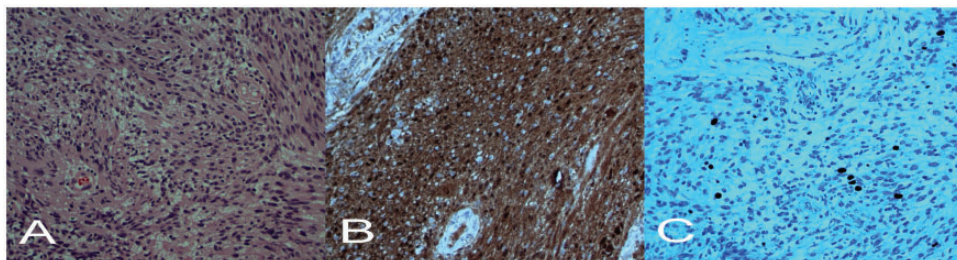
**Figure 1.** Magnetic resonance images showing a large, homogenous, soft-tissue tumour with a thick, smooth capsule in the right paravertebral retroperitoneum. This tumour displaces the iliopsoas muscle distally and the right kidney cranio-laterally, without any signs of ipsilateral hydronephrosis and adjacent organ involvement.

Following preparation, the patient underwent surgery. The encapsulated mass was removed in toto together with its capsular lining, but without definable and clear peripheral nerve encasement or invasion. Identification of the nerve of origin is common, despite complete tumour excision. The procedure was performed without complications and there was no need for blood product substitution. However, extensive bleeding may be encountered from the adjacent vessels.

A histological examination and immunohistochemical staining confirmed the diagnosis of benign encapsulated cellular schwannoma. The tumour had no infiltrative margin and there was a well-defined collagenous capsule and hyalinized vessels. The diagnosis was based on highly ordered, dense, cellular components, typical, well-organized spindle cells with a palisading arrangement (arranged in short bundles of interlacing fascicles), and with Verocay bodies formed by two parallel lines of nuclei with a nuclear space between them indicating an Antoni A pattern (Figure 2A). Reactive hyperplasia of

adjacent lymph nodes was also found. Immunohistochemistry showed S100 protein-positive (Figure 2B) and vimentin-positive, and epithelial membrane antigen-negative and CD34-negative tumour cells. The lack of epithelial membrane antigen immunoreactivity indicated the absence of perineurial cell differentiation. Therefore, the results of the immunohistochemical examination suggested benign schwannoma. The issue of whether the lesion was benign or malignant was resolved with the finding of a low proliferative index ( $Ki-67 = 1.6\%$ ), which highly suggested its benign nature (Figure 2C).

The patient's postoperative course was uneventful except for the transient mild posterolateral thigh pain, paraesthesia, and muscle weakness. She was administered pain relievers, vitamin B complex supplements, and exercise rehabilitation for 3 weeks. After medication and physiotherapy, there was no pain, her muscle power was completely restored, and paraesthesia greatly improved. Considering the benign nature of the tumour and an expected good prognosis, no adjuvant treatment



**Figure 2.** Histopathology of benign schwannoma. Spindle cells show a palisading arrangement with Verocay bodies formed by two parallel lines of nuclei and blood vessels. (A) Haematoxylin and eosin staining,  $\times 10$ . (B) S100-positive cells,  $\times 10$ . (C) The Ki-67 index was 1.6%,  $\times 20$ .

was administered. The follow-up period was 13 months. Abdominal ultrasound was performed every 3 months and MRI was performed at the end of follow-up. During the follow-up, the patient was well, asymptomatic, did not require any therapy and imaging, and did not show disease recurrence, which would require additional surgery. The patient provided verbal informed consent for this report.

## Discussion

Only a small number of schwannomas are found in the retroperitoneum and comprise up to 6% of all primary retroperitoneal tumours. When schwannomas are located in the retroperitoneum, they are usually located in the paravertebral or presacral space.<sup>8</sup> This uncommon site allows slow and asymptomatic expansion of these tumours. Additionally, when these tumours are enlarged they present with unspecific, vague, and bizarre symptoms, which cannot be considered diagnostic. The most common symptoms are pressure symptoms, such as poorly localized pain and discomfort, and non-specific digestive disturbances. Urinary or faecal incontinence and leg neuropathy are rare.<sup>9</sup> Therefore, chronic lower back and dull lateral abdominal pain radiating into the posterolateral thigh, occurring independently of activities and poorly responding to nonsteroidal

anti-inflammatory drugs and physiotherapy as in our patient, could be explained by a retroperitoneal peripheral nerve sheath tumour. However, considering these unspecific and inconclusive symptoms and normal laboratory results also typical for RSs, the diagnosis is based on exclusion, and is often delayed. Nevertheless, reported clinical presentations of RSs are numerous and extremely variable, and include colitis, adrenal mass, psoas abscess, and acute urine retention.<sup>1,10,11,12</sup> Although referred pain and neurological deficits in the lower extremities caused by RSs have also been described, this presentation is rare.<sup>13</sup> Additionally, a clinician rarely suspects RS because the occurrence of unspecific lower back pain in middle-aged patients is almost always regarded as an epidemic symptom affecting the majority of the population once or more during life.<sup>14</sup>

Considering the presentation of our patient, investigation of patients with lower back pain and neurological disturbances around the saddle area and thigh must include a thorough abdominal examination. Abdominal ultrasound is a rapid, inexpensive, and effective tool for initial diagnostic orientation regarding abdominal/retroperitoneal lesions, such as RS. Currently, MRI is accepted as the imaging method of choice for diagnosis of most soft-tissue tumours because of its higher diagnostic predictability compared with

ultrasound or CT. The features of neurogenic tumours, such as bundles (fascicular sign) and a hypointense centre with hyperintense periphery (target sign), are well presented on MRI scans. Additionally, schwannomas are characterized by low-signal intensity on T1-weighted imaging (similar to muscle) and high-signal intensity on T2-weighted imaging (similar to fat). Schwannomas may require acquisition of a fat suppression sequence for delineation from purely lipomatous lesions. However, although MRI may lead to a more accurate preliminary diagnosis, MRI may not reliably distinguish between benign and malignant lesions. Irregular contours, heterogeneity, large size (>5 cm) peripheral enhancement, degenerated areas, intratumoural lobulation, peritumoural oedema, non-cystic appearance of the tumour, and invasion of adjacent structures displayed on MRI scans have been reported as signs of malignancy.<sup>15</sup> These signs should alert surgeons to treat the schwannoma as potentially malignant, especially in patients with Von Recklinghausen's disease.

Definitive diagnosis and determination of the nature of schwannoma are ultimately based on a histological examination. This examination should be supported by immunohistochemical staining using immunoperoxidase techniques for S100 protein and CD34, which are highly positive and negative, respectively, in benign lesions. A strong diffuse expression of S100 protein is a distinct immunohistochemical feature of schwannoma.<sup>16</sup> Additionally, although positive cytoplasmic reactivity for vimentin may identify all mesenchymal cells, it is more consistently expressed in Schwann cells.<sup>17</sup> Furthermore, unlike neurofibromas, most schwannomas do not demonstrate major CD34 immunoreactivity.<sup>18</sup> However, preoperative CT- or ultrasound-guided biopsy may not always yield diagnosis because of harvesting a small tissue sample, and may be associated with the risk of

bleeding, infection, and tumour seeding. Therefore, a definitive diagnosis of schwannoma should be made on postoperative histopathology.<sup>16</sup>

Considering the lack of sensitivity to radiation and chemotherapy, surgery is the primary treatment choice for these tumours.<sup>8,19</sup> The malignancy of schwannoma cannot be totally excluded preoperatively. Therefore, a wide surgical resection of the tumour, even with excision of adjacent structures suspicious of involvement, is advocated by some authors as the treatment of choice.<sup>20</sup> Removal of an RS may be challenging regarding a potentially increased risk of uncontrollable haemorrhage due to tumour adherence to retroperitoneal vessels and its hypervascularity. The most frequent complication after radical surgery is postoperative neurological deficits, which occur in up to 80% of patients, regardless of the atraumatic technique used.<sup>21</sup> This was also the case in our patient who developed transient, mild, posterolateral thigh pain, paraesthesia, and muscle weakness, which responded well to administered therapy. The local recurrence of malignant schwannomas after marginal excision is expected in up to 72% of cases and in only 11.7% of patients after resection with a wide margin.<sup>22</sup> Therefore, in patients with postoperatively proven malignancy, a re-resection with an adequate surgical margin should be considered. Because malignant transformation of benign schwannomas almost never occurs, complete local tumour excision, or even just simple enucleation, may be regarded as the treatment of choice.<sup>23</sup> However, although the prognosis of benign RSs is good and local resection is generally sufficient, the incidence of tumour recurrence because of incomplete excision is 5% to 10%, and metastatic cases after surgery have been reported.<sup>12,24</sup> Therefore, these patients require long-term follow-up. Additionally, special care must be taken in patients with underlying Von

Recklinghausen's disease in whom malignant transformation (neurofibrosarcoma), although rare, is usually observed.<sup>25</sup>

## Conclusion

RSs are rare. Complete local tumour excision should be regarded as the treatment of choice for these tumours. Prognosis of benign RSs is good. Long-term follow-up of these patients is required for postoperative neurological deficits and local recurrence of the tumour. Because of numerous diagnostic and therapeutic problems that may be encountered, successful treatment of RSs require thorough preoperative planning and a multidisciplinary approach.

## Declaration of conflicting interest

The authors declare that there is no conflict of interest.

## Funding

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

## References

1. Fass G, Hossey D, Nyst M, et al. Benign retroperitoneal schwannoma presenting as colitis: a case report. *World J Gastroenterol* 2007; 13: 5521–5524.
2. Al Skaini MS, Haroon H, Sardar A, et al. Giant retroperitoneal ancient schwannoma: Is preoperative biopsy always mandatory? *Int J Surg Case Rep* 2015; 6: 233–236.
3. Sengar (Hajari) AR, Tilve AG, Kulkarni JN, et al. Malignant peripheral nerve sheath tumor of the uterine corpus presenting as a huge abdominal neoplasm. *J Can Res Ther* 2015; 11: 1023.
4. Kalayci M, Akyüz Ü, Demirağ A, et al. Retroperitoneal schwannoma: a rare case. *Case Rep Gastrointest Med* [Published online 2011 July 14] 2011; 2011, Article ID 465062, 3 pages, 2011. doi: 10.1155/2011/465062 Available from: <http://dx.doi.org/10.1155/2011/465062>
5. Gubbay AD, Moshilla G, Gray BN, et al. Retroperitoneal schwannoma: a case series and review. *Aust NZ J Surg* 1995; 65: 197–200.
6. Daneshmand S, Youssefzadeh D, Chamie K, et al. Benign retroperitoneal schwannoma: a case series and review of the literature. *Urology* 2003; 62: 993–997.
7. Choi JW, Choi D, Kim KM, et al. Small submucosal tumors of the stomach: differentiation of gastric schwannoma from gastrointestinal stromal tumor with CT. *Korean J Radiol* 2012; 13: 425–433.
8. Kapan M, Önder A, Gümüş M, et al. Retroperitoneal shwannoma. *J Surg Case Rep* 2011; 10: 1.
9. Song JY, Kim SY, Park EG, et al. Schwannoma in the retroperitoneum. *J Obstet Gynaecol Res* 2007; 33: 371–375.
10. Dadkhah F, Salimi M and Kaviani A. Benign retroperitoneal schwannoma mimicking adrenal mass. *Urol J* 2005; 2: 49–51.
11. Liu YW, Chiu HH, Huang CC, et al. Retroperitoneal schwannoma mimicking a psoas abscess. *Clin Gastroenterol Hepatol* 2007; 5: A32.
12. Wong CS, Chu TY and Tam KF. Retroperitoneal schwannoma: a common tumour in an uncommon site. *Hong Kong Med J* 2010; 16: 66–68.
13. Schindler OS, Dixon JH and Case P. Retroperitoneal giant schwannomas: report on two cases and review of the literature. *J Orthop Surg* 2002; 10: 77–84.
14. Micali S, Virgili G, Vespasiani G, et al. Benign schwannoma surrounding and obstructing the ureteropelvic junction: a case report. *Eur Urol* 1997; 32: 121–123.
15. Yu Y, Wu J, Ye J, et al. Radiological findings of malignant peripheral nerve sheath tumor: reports of six cases and review of literature. *World J Surg Oncol* 2016; 14: 142.
16. Chen W, Dang C, Zhu K, et al. Preoperative management of giant retroperitoneal schwannoma: A case report and review of the literature. *Oncol Lett* 2016; 11: 4030–4034.
17. Jaiswal S. Role of immunohistochemistry in the diagnosis of central nervous system tumors. *Neurol India* 2016; 64: 502–512.

18. Park JY, Park H, Park NJ, et al. Use of calretinin, CD56, and CD34 for differential diagnosis of schwannoma and neurofibroma. *The Korean Journal of Pathology* 2011; 45: 30–35.
19. Mastoraki A, Toska F, Tsiverdis I, et al. Retroperitoneal schwannomas: Dilemmas in diagnostic approach and therapeutic management. *J Gastrointest Cancer* 2013; 44: 371–374.
20. Okuyama T, Tagaya N, Saito K, et al. Laparoscopic resection of a retroperitoneal pelvic schwannoma. *J Surg Case Rep* 2014; 1: rjt122.
21. Park MJ, Seo KN and Kang HJ. Neurological deficit after surgical enucleation of schwannomas of the upper limb. *J Bone Joint Surg Am – Series B* 2009; 91: 1482–1486.
22. Das Gupta TK, Brasfield RD, Strong EW, et al. Benign solitary schwannoma. *Cancer* 1993; 72: 513–514.
23. Hide IG, Baudouin CJ, Murray SA, et al. Giant ancient schwannoma of the pelvis. *Skeletal Radiol* 2000; 29: 538–542.
24. Kim SM, Seo SW, Lee JY, et al. Surgical outcome of schwannomas arising from major peripheral nerves in the lower limb. *International Orthopaedics* 2012; 36: 1721–1755.
25. Karadurmus N, Basaran Y, Buyukturan G, et al. Neurofibromatosis type 1 complicated with malignant transformation and diffuse pulmonary disease. *J Oncol Sci* 2016; 2: 84–86.