Asian Journal of Case Reports in Surgery

8(1): 1-5, 2021; Article no.AJCRS.66096

A Rare Case of Large Retroperitoneal Schwannoma

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Authors' contributions

This work was carried out in collaboration among all authors. Author EP conducted the pathologoanatomical research. Authors GS and GF provided us with all the necessary elements of the case, as the responsible physicians of the patient. All authors read and approved the final manuscript.

Article Information

Editor(s):
(1) Dr. José Francisco de Sales Chagas, Sao Leopoldo Mandic Medical School, Brazil.
Reviewers:
(1) Eduardo Esteban-Zubero, Hospital San Pedro, Spain.
(2) Oshan Basnayake, University of Colombo, Sri Lanka.
Complete Peer review History: http://www.sdiarticle4.com/review-history/66096

Case Report

Received 28 December 2020
Accepted 02 March 2021
Published 19 March 2021

ABSTRACT

Aims: The aim of this manuscript is to present a rare case of a schwannoma in the retroperitoneal space.
Case: We present the case of a 61-year-old female with a retroperitoneal tumor which proved to be a schwannoma. After incomplete surgical excision, a second surgical exploration, due to tumor relapse, was conducted a year later.
Discussion: Schwannomas or Neurilemmomas are mesenchymal tumors arising from Schwann cells of peripheral nerve sheaths. The first tumors of peripheral nerves were described in 1910 by Jose Verocay, but it was not until 1935 that Arthur Purdy Stout used the term Neurilemmoma. Retroperitoneal space is an unusual location of schwannomas and their presence may only be expressed by insidious onset of nonspecific symptoms like vague abdominal pain. Definite diagnosis of schwannomas is only made post-operatively, by histopathological examination of the specimen.
Conclusion: Schwannomas, even if they are infrequent, should be included in the differential diagnosis of any retroperitoneal abdominal mass.

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Keywords: Retroperitoneal tumor; schwannoma; neurilemmoma.

1. INTRODUCTION

Primary tumors of the retroperitoneal space are quiet rare. Retroperitoneal schwannomas comprise only 1-10% of all retroperitoneal tumors, and approximately 3% of all schwannomas [1]. Schwannomas are most commonly present in head and neck region (44,8%), upper limbs (19,1%) and lower limbs (13,5%). They may, rarely, be described in the posterior mediastinum and the retroperitoneum, usually in the paravertebral space or the presacral region [2]. Among all schwannomas, only 0.7% of benign ones and 1.7% of malignant ones are retroperitoneal [3]. The age of presentation of these tumors is between the second and fifth decade of life, while their distribution according to sex is controversial, with some studies reporting a female predominance, whilst others note that there is no predisposition for sex [4].

Schwannomas are usually benign, but infrequently may undergo malignant transformation. The size of the tumor is not related to its malignant tendency. According to recent studies regarding schwannomas, 90% of cases are sporadic, while 3% are associated with Von Recklinhausen disease. It has been reported that when they are spotted in the retroperitoneal region, 5-18% of schwannomas are related to Von Recklinhausen disease [5]. In these cases, the location of schwannomas is usually atypical and malignant. Preoperative diagnosis of retroperitoneal schwannomas is very difficult because of their site and the lack of specific features on the imaging modalities while the definitive diagnosis is based on pathological, histological and immunohistochemical examinations postoperatively [6,7].

2. CASE PRESENTATION

A 61-year-old female was admitted to our department of surgery because of diffuse abdominal pain for several months. Past medical history included total abdominal hysterectomy due to multiple fibroids of the uterus. Clinical examination, including neurology examination, was completely normal. Laboratory tests were, also, normal. Computed tomography (CT) was performed, which showed the presence of a 10x9.8 cm solid mass in the retroperitoneal space, on the right side of the vertebral column, expanding in front of the sacrum. Open surgical exploration was performed and an encapsulated large solid mass, closely adhered to adjacent tissue, was found shown in Fig. 1. The mass was separated from adjacent tissues and excised from retroperitoneum. There was important blood loss intraoperatively, which was coped with crystalloids and blood transfusion. The rest of the post-operative course was uneventful. No neurological deficits or any other complications were observed.

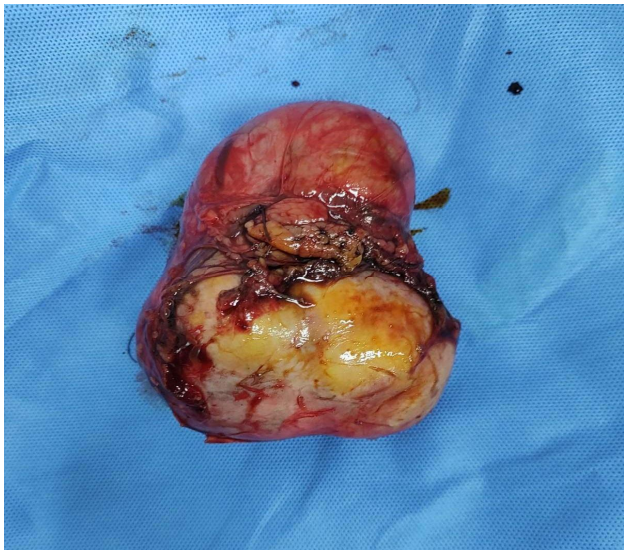

Fig. 1. Encapsulated, solid retroperitoneal mass

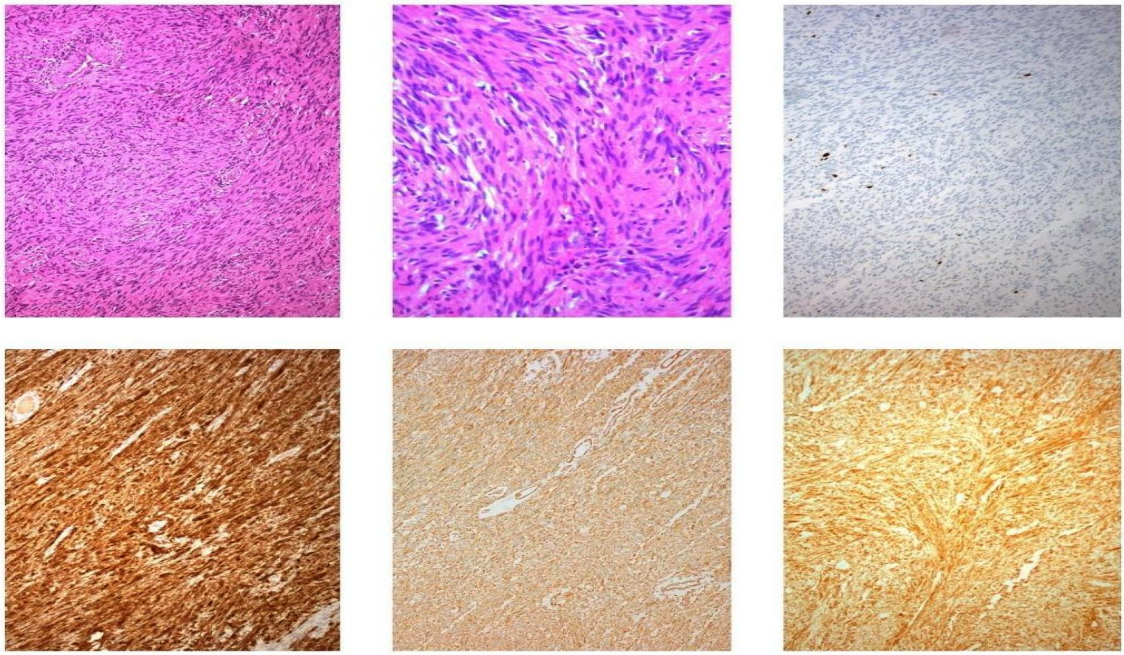

Fig. 2. Microscopic appearance of the case a) Splinde shaped cells in a typical palisading pattern/ Antoni A b) Myxoid and degenerative tissue with fewer cells/antoni B c) Ki67 expression d) S100 expression e) Vimentin expression f) GFAP

3. HISTOLOGICAL FINDINGS

Histopathological examination of specimen revealed a bipolar growth pattern, with areas of spindle-shaped cells in a typical palisading pattern (Antoni A) shown in Fig. 2a and areas of myxoid and degenerative tissue with fewer cells (Antoni B) shown in Fig. 2b. Mitotic rate was very low (less than 1/20 high power fields) and Ki67 shown in Fig. 2c was expressed in less than 5% of tumor cells. Immunohistochemical examination revealed cellular positivity for S-100, Vimentin and GFAP shown in Fig. 2d, 2e, 2f respectively.

The final diagnosis was benign cellular schwannoma. Positive surgical margins were discovered and a tumor relapse was faced with a second surgical excision, a year later. Since then, no evidence of recurrence has been observed.

4. DISCUSSION

Retroperitoneal schwannomas are encapsulated, solitary, well-circumscribed tumors, usually large in size. If they undergo malignant transformation, the tumors exhibit irregular contours and distinct invasiveness into the adjacent tissue [5]. Their main symptoms include abdominal distension, abdominal pain and nonspecific symptoms like backache and digestive disturbances. Atypical presentations are rare and include hematuria, headache, secondary hypertension and recurrent renal colic pain [8]. Our patient presented abdominal pain and distension, without any other symptoms.

Correct preoperative diagnosis of retroperitoneal schwannomas is difficult. The differential diagnosis with schwannomas include fibrosarcoma, liposarcoma, ganglioneuroma, histiocytoma, lymphoma, hepatic tumors, pancreatic cystic tumors and psoas abscesses [1,5]. Ultrasound, Computed Tomography (CT), and Magnetic Resonance Imaging (MRI) have been used, but the lack of specific imaging signs makes the diagnosis of retroperitoneal schwannomas a hard task for doctors. On contrast-enhanced CT (CECT) abdomen, they are visualized as well circumscribed rounded mass with heterogeneous contrast enhancement due to cystic and haemorrhagic changes and calcification.In MRI, schwannomas are described as masses of low signal intensity on T1-weighted images and high signal intensity on T2-weighted images [6]. A few well-known typical imaging characteristics for schwannomas, target sign and

fascicular sign, are not frequently seen in retroperitoneal schwannomas. The “fascicular sign” is the appearance of bundles, which is a general property of neurogenic tumors and the “target sign” is the presence of hypointense centre and hyperintense periphery on T2 weighted MRI [9]. US or CT guided core biopsy has been used to help the diagnosis but it has been found unreliable as it presents risks such as tumor dissemination, hemorrhage and infection [2]. Hence there is no gold standard preoperative diagnostic modality for retroperitoneal schwannoma. Microscopically, two patterns are recognized, clusters of compact elongated bipolar spindle cells arranged in a palisading pattern (Antoni A) and loosely arranged cells in a myxoid background (Antoni B areas) [5]. The immunohistochemistry panel is typically positive for S-100 protein, and sometimes for vimentin, Neuro-specific Enolase (NSE) and CD56. SOX10 has also been, recently, found to be expressed in schwannomas, to a lesser extent [1]. In our case, the mass was discovered in a CT, but definitive diagnosis was, only, made post-operatively by histological examination of the tumor.

The optimal treatment for retroperitoneal schwannomas is complete surgical excision, as these tumors demonstrate a lack of sensitivity to chemotherapy or radiotherapy [10]. However, a considerable controversy exists over negative soft tissue margins. Some argue for complete surgical excision that may include, if necessary, the sacrifice of adjacent tissue and viscera. It is argued that the local recurrence rate ranges from 16% to 54% after conservative intralesional enucleation [11,12]. Additionally, because malignancy cannot be excluded accurately preoperatively or even intraoperatively with frozen section analysis, complete surgical resection so as to attain negative margins is recommended. 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. The argument here is that the morbidity associated with resection of adjacent tissue would not be justified in the treatment of a benign lesion. The prognosis of retroperitoneal schwannoma is extremely good. Recurrence, as in our case, when reported (5-10%) is due to incomplete excision and surgical resection of the lesions is recommended if it occurs [1,6]. Periodic follow-up of patients with control imaging studies is suggested, with the minimum postoperative follow up time being 3 to 5 years after initial diagnosis [13,14].

5. CONCLUSION

Retroperitoneal schwannoma is a rare, benign tumor. Its diagnosis is based on histopathology and immunochemistry, and surgical excision remains the treatment of choice. Local recurrence is closely correlated with negative resection margins and pathology types. A postoperative follow up of 3 to 5 years should be applied to exclude tumor recurrence. Schwannomas should be included in the differential diagnosis of any retroperitoneal abdominal mass.

CONSENT

Written informed consent has been obtained by the patient.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the authors. The research was conducted ethically in accordance with the World Medical Association Declaration of Helsinki.

DATA AVAILABILITY

The data that support the findings of this case report are available from the corresponding author, I. Galanis, upon reasonable request.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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PMID: 29931989.

Peer-review history:
The peer review history for this paper can be accessed here:
http://www.sdiarticle4.com/review-history/66096