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Case report

# Giant pelvic schwannoma presenting with increased urinary frequency - Case report

Omejc Andrej [a], Vegan Lucija [a], Omejc Mirko [b,*]

[a] Medical Faculty, University of Ljubljana, Slovenia
[b] Department of Abdominal Surgery, University Medical Centre Ljubljana, Zaloska c.7, 1000 Ljubljana, Slovenia

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ABSTRACT

*Introduction and importance:* Schwannoma is a relatively slow-growing and usually benign tumor that consists of myelin-producing Schwann cells. It is most often associated with 8th cranial nerve (vestibular schwanomma) and only rarely presents outside the head and neck. Extremely rarely it occurs as a retroperitoneal mass that can remain asymptomatic until it reaches a size so big that it compresses the adjacent organs and therefore requires surgical intervention.

*Case presentation:* We present a case of 58 years old male with a benign pelvic schwannoma, adherent to S1-S2 intervertebral foramen, presenting with increased urinary frequency and treated with surgical *en block* excision.

*Clinical discussion:* Diagnostic procedure and surgical treatment of giant pelvic schwanomma could be both challenging. Radiologic investigation is essential for preoperative planning and helpful in diagnosis but does not offer a definitive diagnosis of pelvic schwannoma. Definitive diagnosis is therefore based on histological and immunohistochemical analysis of surgically removed specimens. Possible surgical interventions include complete or partial resection, both bearing its risks and advantages in terms of postoperative morbidity.

*Conclusion:* Although sometimes challenging, complete surgical resection remains the mainstay of treatment. If possible, it should be performed by *en block* resection since it provides the best short and long term prognosis for the patient.

## 1. Introduction

Schwannomas are a relatively slow- growing and usually benign tumors that arise from myelin- producing Schwann cells. Malignant transformation is rare and is usually associated with von Recklinghausen's syndrome (neurofibromatosis 1). It accounts for 5–18% of cases. The usual locations are head and neck (most commonly schwannoma of the 8th cranial nerve- vestibular schwannoma), occasionally they also occur in the mediastinum and extremities. Pelvic schwannoma is an extremely rare entity that accounts for less than 0,5% of reported cases [1,2].

Pelvic schwannomas often present as so called “ancient schwannomas”. This term is used for tumors with occurrence of degenerative changes, such as calcification, cyst formation and hemorrhage-features that are the result of tumor's extensive growth [3]. They are well encapsulated. However, because they don't possess any pathognomonic features on radiologic evaluation and preoperative biopsy is also not recommended, surgery is necessary both for definitive diagnosis and treatment.

Schwannomas in the retroperitoneal area are in general at first asymptomatic and usually occur as incidental findings on diagnostic imaging tests. Signs and symptoms often develop only once the tumor has reached a size so big that it compresses the surrounding tissues or nerve roots. They can be vague and nonspecific and can include abdominal heaviness and distention, backache, increased urinary frequency or constipation. They often mimic other pelvic lesions (fibrosarcoma, liposarcoma, ganglioneuroma, hydatid cyst, haematoma, connective tissue diseases…) and can therefore quickly lead to misdiagnosis. Preoperative radiographic imaging is therefore essential in order to determine tumor's size, location and relationship with surrounding tissue.

In our case report we present a patient with a large presacral schwannoma, adherent to S1 and S2 intervertebral foramen that was treated with surgical excision. This case report has been reported in line with the SCARE 2020 criteria [4].

* Corresponding author.
*E-mail address:* mirko.omejc@kclj.si (O. Mirko).

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## 2. Case report

A 58-year old otherwise healthy male patient complained of increased urinary frequency. History of medical conditions revealed left inguinal mesh hernioplasty and high blood pressure. He did not report any familial genetic conditions or allergies. Laboratory findings showed no abnormalities, CEA, CA 19-9 and PSA levels were all normal. Clinical examination revealed a palpable mass extending above the pubic bone to the area bellow umbilicus. He was referred to ultrasound investigation. Ultrasound investigation revealed a large mass in extraprostatic space. He was further investigated by MRI scan that revealed 12 × 13 × 10 cm-sized circumscribed pelvic tumor, which was not showing any signs of invasive growth. It has extended to S1-S2 intervertebral foramen which was dilated. The spinal canal was intact. The mass was displacing the rectosigmoid colon to the right side and the urinary bladder upwards. The compression of the bladder explained the patient's increased urinary frequency. The radiologic characteristics of the mass suggested pelvic schwanomma as the most possible diagnosis (Figs. 1, 2).

### 2.1. Surgery

Although surgical excision is curative in cases of benign schwannoma, malignant schwannomas carry a poor prognosis as they are commonly resistant to chemotherapy and radiotherapy [1,2,12]. Even excision of benign intrapelvic and retroperitoneal schwannomas can be associated with massive bleeding if the tumor capsule is adherent to the presacral venous plexus. It is therefore of utmost importance to avoid injury to the adjacent visceral, vascular, and nervous structures, which may result in complications like hemorrhage and neurological deficits.

In our case the only option for the removal of the mass was surgical resection. Due to the large size of the tumor and its location in a very narrow, obese male pelvis, anterior open approach was chosen. The patient was placed in supine position and lower midline incision from umbilicus to symphysis pubis was performed. After opening the abdomen, a large encapsulated retroperitoneal mass of about 20 cm in diameter was detected in retroperitoneum occupying the entire pelvis, displacing the urinary bladder upwards and rectosigmoid colon to the right side. The lesion was immobile, without evidence of local invasion. The excision of the mass was challenging. Due to its size, the peritoneum was opened over the mass. The sigmoid colon was mobilized from the presacral space to expose the tumor mass. The ureters were identified bilaterally and protected. A plane was then established between the tumor mass and the presacral alar tissue. Small feeding vessels were individually coagulated and transected to further mobilize the tumor by using bipolar cautery and ligasure device. Tumor was firmly attached by fibrous tissue to presacral fascia and left iliac vein. In circumferential fashion the tumor was completely mobilized from the sigmoid colon and upper rectum, the left iliac vein and other surrounding anatomical structures with great care of exact hemostasis. Finally, the remaining attachment of the tumor tissue to the presacral nerve root(s) was identified and carefully dissected free with ligasure device. The tumor was then completely mobilized and delivered en bloc from the surgical field (Figs. 3, 4).

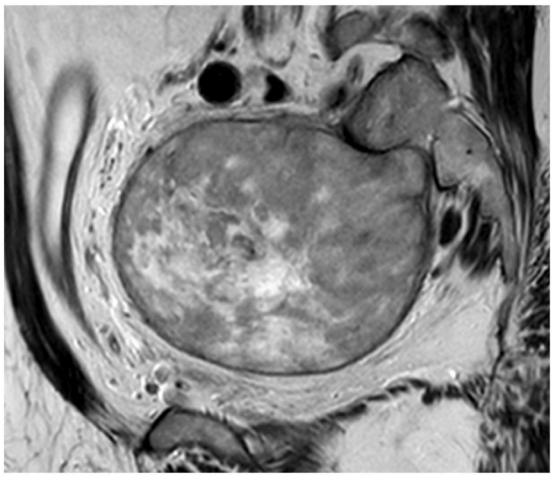
The wound was then closed in standard fashion. The duration of the surgery was 240 min. Intraoperative blood loss was less than 500 ml. The patient had an uncomplicated recovery and was discharged home on POD 6. Following the surgery, he did not show any complications such as urinary and stool incontinency or impotency at 4 month after surgery.

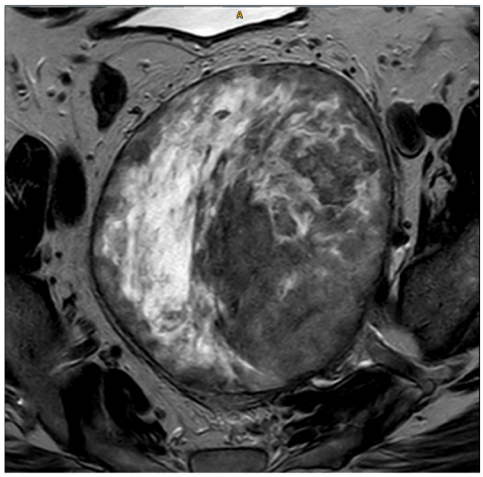
### 2.2. Pathology

Postoperative histopathologic investigation of the mass revealed its exact size (16 × 14 × 8 cm) and its significant weight (755 g). The tumor was oval and encapsulated. The cut surface was light yellow with areas of cystic degeneration. Histologically it was composed of spindle-shaped cells with variable cellularity. The stroma showed mixoid degeneration with focal presentation of small calcifications. Between tumor cells many foamy macrophages were observed. There was no visible mitosis or necrosis. At the periphery there were many lymphoid infiltrates. Immunohistochemically the tumor cells expressed S-100 protein. CD117, DOG1 and Desmin testing were negative (Figs. 5, 6).

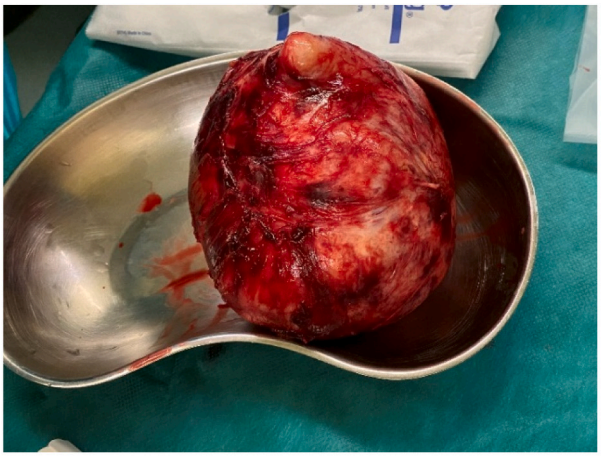
## 3. Discussion

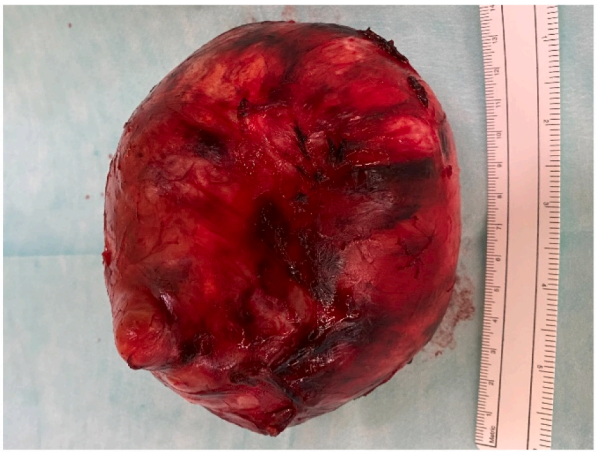
Pelvic schwannomas are rare tumors that account for 0.3–3.2% of all schwannomas and 0.4–15% of all retrorectal masses [1,5]. Classification based on the lesion's location have been described by Klimo and divided into three groups: purely sacral/intracanalicular (type I), purely presacral (type III), or mixed intracanalicular/presacral (type II) [9]. Type III tumors, according to this classification, limited to the presacral space, are among the rarest and the present case is one of the largest (16 × 14 × 8 cm) described in a male [10]. Pelvic schwannomas are slow-growing tumors that often remain asymptomatic until reaching a considerable size. Usually they present with non-specific symptoms such as low back pain, incontinence, urinary urgency and constipation. Slow growth and asymptomatic course often result in tumor's firm adhesions to the abdominal nerves and viscera. Therefore surgical excision of the mass is complicated and associated with significant postoperative morbidity and neurological deficits in 21% of cases [6,7]. The diagnostic procedure can be challenging since the mass often resembles other pelvic lesions. Differential diagnosis includes fibrosarcoma, liposarcoma,



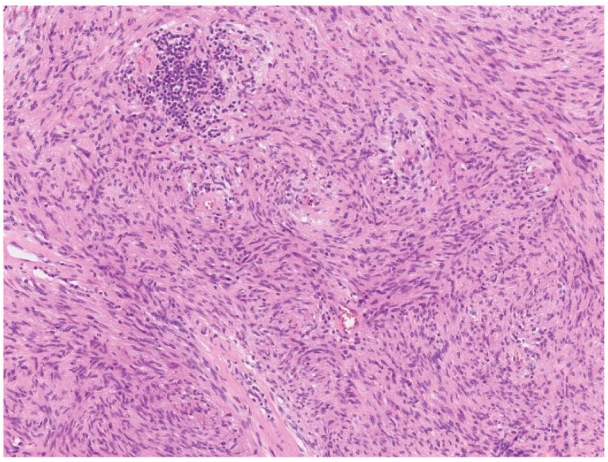


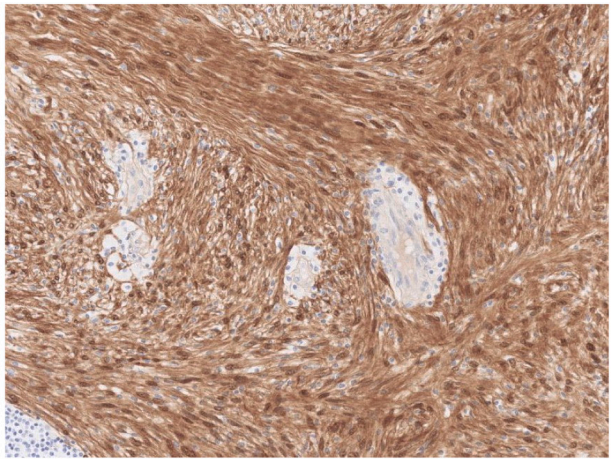
**Figs. 1, 2.** MRI of the pelvis showing the mass that displaces the rectum and the bladder with extension to S1-S2 intervertebral foramen.





**Figs. 3, 4.** Resected pelvic schwannoma (measured in centimeters).





**Figs. 5, 6.** Microscopic image shows spindle-shaped cells with variable cellularity, immunohistochemically, tumor cells are positive for S-100 protein.

ganglioneuroma, hydatid cyst, haematoma and connective tissue diseases [8]. There are no specific clinical or radiological findings and therefore misdiagnosis is a common occurrence. Still radiologic imaging plays a pivotal role in the preoperative diagnostic procedure. The method of choice is MRI that shows hypointensity on T1- weighted and hyperintensity on T2-weighted images but unfortunately this is not always the case [2]. Often suggested is also ultrasound guided biopsy, though it is often inaccurate and misleading. Due to the lack of effective radiologic procedures the definitive diagnosis of pelvic schwannomas is by histopathologic investigation of surgically resected specimens.

Possible surgical interventions include complete or partial excision. Gross total resection is preferred method, with several authors suggesting that risk of recurrence is decreased by en bloc resection [10,12]. However, a R0 “en bloc” resection is often very challenging, may cause unacceptable morbidity depending upon tumor morphology, surgeon's skill, and patient anatomy [10,11]. Extremely large, type III lesions may not be amenable to en bloc resection due to anatomic constraints. In such cases, safe dissection requires internal debulking (e.g., with an ultrasonic aspirator), followed by capsular dissection [3,12]. On the other hand, partial excision is associated with a higher incidence of relapse. The possible recurrence of the tumor results in higher chance of malignant transformation and higher postoperative mortality.

## 4. Conclusion

Giant pelvic schwannoma provides a challenge for the physician both in terms of diagnosis and treatment. Symptoms can often mimic other pelvic lesions and radiologic investigation isn't able to provide conclusive answers. However, radiologic findings play an essential part in preoperative planning. Definitive diagnosis is based on histological and immunohistochemical analysis of surgically removed specimens. If possible, this should be performed by complete *en block* resection since it provides the best short- and long- term prognosis for the patient.

## Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

## Provenance and peer review

Not commissioned, externally peer-reviewed.

## Ethical approval

The study is exempt from ethical approval.

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Not applicable.

## Credit authorship contribution statement

Mirko Omejc operated and conceived the case report.
Andrej Omejc, Lucija Vegan collected the data, contributed to the writing.

All authors discussed the results and contributed to the final manuscript.

## Declaration of competing interest

None.

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