A 68-Year-Old Woman Presenting with Recurrent Abdominal Pain and a Diagnosis of a Presacral Retroperitoneal Benign Schwannoma that Mimicked an Ovarian Tumor on Pelvic Magnetic Resonance Imagining

ABEF
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Patient: Female, 68-year-old
Final Diagnosis: Presacral retroperitoneal benign schwannoma
Symptoms: Recurrent abdominal pain
Medication: —
Clinical Procedure: Excision of the mass • laparotomy
Specialty: Surgery

Objective: Rare disease
Background: Schwannomas are benign tumors and their appearance in the pelvic region is rare and poses a major diagnostic problem. They can be sporadic or associated with genetic syndromes. They have a slow growth rate and may be asymptomatic for many years. Symptoms are usually nonspecific and due to compression of adjacent structures. Abdominal imaging modalities may not be able to differentiate a benign schwannoma from a malignant retroperitoneal tumor. This report is of a case of a 68-year-old woman presenting with recurrent abdominal pain and a diagnosis of a presacral retroperitoneal benign schwannoma that mimicked an ovarian tumor on pelvic magnetic resonance imaging.

Case Report: The patient had a history of a femoral hernia repair and recurrent lower abdominal pain. Pelvic imaging raised the suspicion of a primary ovarian tumor. The mass appeared to have clear cleavage planes with the surrounding structures, so the patient was proposed for an exploratory laparotomy. Prior to the surgery, an additional pelvic computed tomography (CT) was performed (10 months after the first one), which did not show progression of the disease. The histological examination result was compatible with a benign retroperitoneal schwannoma and not an ovarian tumor.

Conclusions: This report highlights that the diagnosis of retroperitoneal and pelvic masses can be challenging. In women, a primary ovarian tumor should be excluded on imaging and the diagnosis of a benign tumor, such as schwannoma, must be confirmed by histopathology, either preoperatively or following tumor resection.

Keywords: Diagnosis, Differential • Histology • Laparotomy • Neurilemmoma • Pelvic Neoplasms • Retroperitoneal Neoplasms

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Background

Schwannomas, also called neurilemmomas, are rare benign tumors arising from the Schwann cells of the nerve sheaths [1]. Although usually found in the head and neck, they can appear in other sites, like the pelvis, and mimic other entities, which makes the diagnosis challenging [2-4]. In fact, pelvic/presacral schwannomas are quite rare and represent 0.3% to 3.2% of all these tumors [3]. Most schwannomas occur in a sporadic setting. However, they can be associated with genetic syndromes like neurofibromatosis, schwannomatosis, or Carney’s complex [5]. Abdominal organs can be externally compressed when the mass reaches a considerable size. It is usually at this point that the patient becomes symptomatic [1-3], which can take up to several years considering they are slow-growing tumors [1,3,4,6]. However, most patients’ symptoms are nonspecific, like pelvic discomfort, constipation, or back pain [2,3,6] and there are cases of unusual presentations of presacral schwannomas described in the literature [1,3,4].

Imaging has an important role in the differential diagnosis and characterization of pelvic masses. Both computed tomography (CT) scans and magnetic resonance imaging (MRI) are useful [3,6]. While CT allows the identification of the mass as solid or cystic and evaluates the involvement of adjacent structures, MRI has a higher resolution in the evaluation of soft tissues, which can help the surgeon to better define the planes of resection and thus perform a safer surgery [6].

In some cases, even MRI is unable to differentiate a schwannoma from other tumors, and the correct diagnosis is only possible by histological confirmation. The classical characteristics are spindle-shaped cells with an eosinophilic cytoplasm, arranged in either a dense (Antoni A) or loose (Antoni B) pattern, with the presence of Verocay bodies [7].

Surgical resection is the first-line treatment for most presacral tumors, including schwannomas [1,3]. Giving its proximity to nerve roots, the surgical procedure must be conducted with extreme care to remove the entire lesion without damaging these structures [3,6]. The extent of the surgery depends on the size of the lesion and invasion of adjacent tissues. Both transabdominal and perineal approaches can be used [6].

This report is of a case of a 68-year-old-woman presenting with recurrent abdominal pain and a diagnosis of a presacral retroperitoneal benign schwannoma that mimicked an ovarian tumor on pelvic magnetic resonance.

Case Report

A 68-year-old woman, with a history of arterial hypertension and a femoral hernia repair 3 years earlier, was referred to our general outpatient clinic for assessment of frequent lower abdominal pain. On the initial physical examination, a recurrence of the hernia was suspected, so the patient underwent a pelvic ultrasonography. No hernia was detected on the ultrasound, but an apparent adnexal mass approximately 6 cm in diameter was found. For further characterization, an abdominopelvic CT was ordered, which revealed a well-circumscribed pelvic mass in the presacral region with a suspected relationship with the ovary, and some hypointense areas compatible with necrosis. The initial differential diagnosis was between subserosal pedunculated fibroma and ovarian fibrothecoma. The pelvic MRI showed “in the presacral region, slightly on the right, the presence of a neoplasm, round in shape, measuring 69×54×67 mm with well-defined borders, apparently encapsulated with a hypointense signal in T1 and T2 sequences and intense contrast enhancement”. No hemorrhage was found, but there were some cystic degeneration areas and calcifications inside the mass (Figure 1). It presented clear cleavage planes with the right ovary, the uterus, and the sacral bone. The first diagnostic hypothesis was a retroperitoneal tumor like a sarcoma or histiocytoma. Endoscopic examinations (upper and lower gastrointestinal endoscopy) were conducted and showed no involvement of the gastrointestinal tract.

The patient was therefore proposed for open surgical resection of the lesion. Prior to the surgery, another CT was performed (10 months after the initial examination) to better define the surgical strategy. This new image showed that the mass had barely changed in size (which made the malignant nature of the lesion unlikely) and was extrinsically compressing the right iliac vein and the bowel loops, without invading them. No secondary lesions were found.

Figure 1. A pelvic magnetic resonance imaging scan (T2 sequence) showing a hypointense well-circumscribed mass in the presacral region with cystic degeneration and calcification areas inside (shown by the arrow).
An exploratory laparotomy with a median infraumbilical incision was then performed. The sigmoid colon was mobilized from the presacral space, allowing the identification of a round-shaped mass with a hard consistency, below the bifurcation of the iliac vessels. The peritoneum was incised over the lesion, which was then carefully dissected from the surrounding tissues, including the right ureter and right iliac vessels. None of these structures was invaded but there were some small collaterals arising from the right iliac vein, which supplied the mass and had to be ligated. After that, it was possible to separate the whole mass from the sacral bone with bipolar cautery without damaging any nerve roots. The final surgical piece (Figure 2) was excised en bloc. A careful hemostasis of the presacral area was performed and the abdominal wall was closed.

The patient had an uneventful recovery and was discharged home on the third hospital day. At 1 and 5 months after surgery she was feeling well, with no residual pain or radiculopathy. One year after the surgery she remained asymptomatic and no complications of the wound healing occurred, like wound dehiscence or incisional hernia.

A histological examination showed an encapsulated nodular lesion with some areas of high cellularity and others composed of loose stroma, with elongated nuclei and no atypia (Figure 3). It was also possible to identify areas of nuclear alignment or palisading, in the periphery, as well as some cystic degeneration zones, with thick hyalinized blood vessels (Figure 4). The immunohistochemical (IHC) panel staining was negative for CD34, negative for epithelial markers CK7 and CK20, and negative for alpha-smooth muscle actin and desmin. On the other hand, it was strongly positive for S-100 protein. The Ki67 IHC staining was less than 5%. All these characteristics were compatible with a retroperitoneal benign schwannoma. The excision was complete and the margins were tumor-free.

**Discussion**

The diagnosis of presacral schwannomas is challenging, as shown by our case report [3]. The differential diagnosis of presacral tumors is wide and varies from benign congenital lesions, such as development cysts or benign neurogenic tumors, like schwannomas and neurofibromas, to malignant congenital tumors, like teratomas and soft-tissue sarcomas [3,5,7].

Most patients only become symptomatic when they have large tumors and the symptoms are usually vague and nonspecific. The physical examination may be normal [1,7]. In our case, the patient was initially referred for a recurrent abdominal pain and a suspected femoral hernia recurrence. As described previously in the literature, presacral schwannomas may present...
with abdominal pain or pelvic heaviness [1,4] but they can also be perfectly asymptomatic and diagnosed incidentally [3]. Both CT and MRI can aid in the diagnosis [3,6] – schwannomas usually appear as isoto-hypointense on T1-weighted images, hyperintense in T2, and show postcontrast enhancement in MRI [3,8]. However, they can be heterogeneous and sometimes it is difficult to differentiate a schwannoma from other soft-tissue tumors, including malignant ones [8], particularly when they appear in unusual locations, as in our case, where an ovarian tumor was initially suspected, but turned out to be a presacral benign schwannoma of the retroperitoneum, mimicking a primary tumor of the ovary. Imaging has also an important role in surgical planning. Endoscopic procedures are useful to assess gastrointestinal tract involvement [6].

The definite diagnosis is made through histological examination, by identification of Schwann cells with a spindle morphology (Antoni A pattern) alternating with less cellular areas with loose stroma (Antoni B) [3,5,7]. There can also be cystic degeneration areas or calcifications, as in our case. The typical immunostaining pattern is a strong positivity for S-100 protein in the cytoplasm and a negative CD34 [2,5].

According to the most recent literature, a preoperative biopsy, via transperineal or presacral, may be conducted in selected cases when the diagnosis of a malignant mass cannot be excluded, especially in patients who might benefit from neoadjuvant treatments. Other approaches, like transperitoneal, transrectal, or transvaginal are not recommended due to the high risk of complications, like infection, hemorrhage, and tumor seeding [6].

Regarding treatment, the surgical approach to a pelvic schwannoma can be tricky, considering the complex anatomy of that region [1,6]. The aim is to remove the lesion with negative margins and minimum morbidity for the patient. Invasion of other structures or organs will determine the extension of resection. If there is intrasacral involvement, for example, the surgery should be performed with the assistance of a neurosurgeon [7]. In our case, however, the mass was extrinsically compressing the iliac vessels and bowel, but there was no evidence of organ or bone invasion, so a complete resection was possible. We chose an anterior approach (transabdominal) to reach the mass, although the literature also describes a posterior (perineal) approach for small lesions below S3 level. For higher lesions, both techniques can be used [6]. The main problem with the anterior approach may be the control of the bleeding [7], considering schwannomas are usually well-vascularized, with multiple venous vessels that should be carefully ligated. On the other hand, a posterior approach usually makes the dissection of high lesions more difficult due to the lack of good exposure, which increases the risk of iatrogenic injuries. There are also cases of laparoscopic resection of pelvic schwannomas described in the literature [9,10]. Schwannomas are usually benign and they rarely undergo malignant transformation [1], so the prognosis is favorable after R0 resection.

Conclusions

Pelvic schwannomas are rare tumors and their accurate diagnosis can present a challenge due to the unusual location for this type of neoplasm. This case report highlights that the diagnosis of retroperitoneal and pelvic masses can be challenging. In women, a primary ovarian tumor should be excluded on imaging and the diagnosis of a benign tumor [4], such as schwannoma, must be confirmed by histopathology, either preoperatively or following tumor resection.

The first-line treatment for a benign schwannoma is complete resection of the lesion, which confers a good prognosis.

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