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Case Report

Schwannoma mimicking ovarian malignancy ☆

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ABSTRACT

In this article, we present a case of retroperitoneal schwannoma localized in the pelvic cavity mimicking ovarian carcinoma. A 60-year-old woman presented with a feeling of pelvic heaviness and dyspareunia for 3 months. On physical examination, a hardened mass is palpated on the cul-de-sac of Douglas, measuring approximately 10 cm. The sonographic study showed a retro-uterine solid mass, containing cystic areas, measuring 14 cm. Magnetic resonance imaging showed a solid left tumor in the small pelvis, posterior to the uterus, suspicious of an ovarian malign tumor. Surgery revealed a retroperitoneal pelvic tumor and uterus and adnexa without macroscopic changes. Pathology examination of the pelvic mass confirmed the diagnosis of schwannoma. In the present case, it is emphasized that it is easy to misdiagnose a pelvic mass as an ovarian tumor. While prompt recognition of ovarian cancer remains essential, awareness of processes that mimic ovarian tumors can avoid potential misdiagnosis. The pelvis has a complex anatomy and there are some imaging signs that help assessing the origin of a mass, especially in cases of masses abutting the ovary.

Introduction

The complex anatomy of the female pelvis and adnexa on imaging poses a diagnostic challenge in certain cases because it may not be clear to identify the origin of a lesion. Ovarian and extra-ovarian processes occupying the adnexa may appear as ovarian carcinoma, the second most common gynecologic malignancy and the fifth leading cause of cancer death in women [1]. Among the extra-ovarian etiologies, we can find involvement of gastrointestinal and genitourinary organs, and, less commonly, connective tissues, nerves, and lymphovascu-

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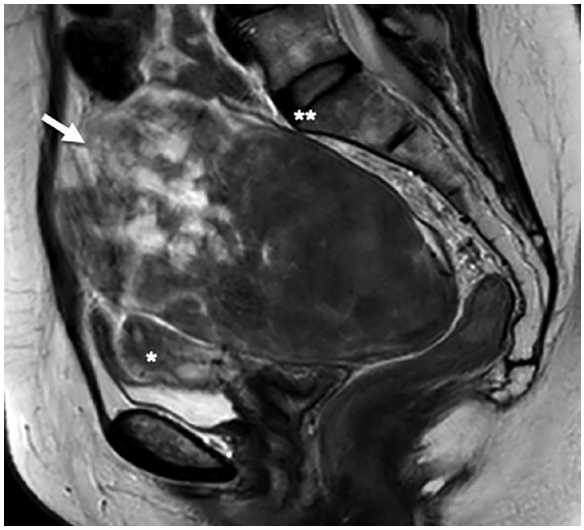

Fig. 1 – Sagittal T2-weighted image shows a well-defined solid mass (white arrow) posterior to the uterus (*), with posterior compression by the sacral promontory (**).

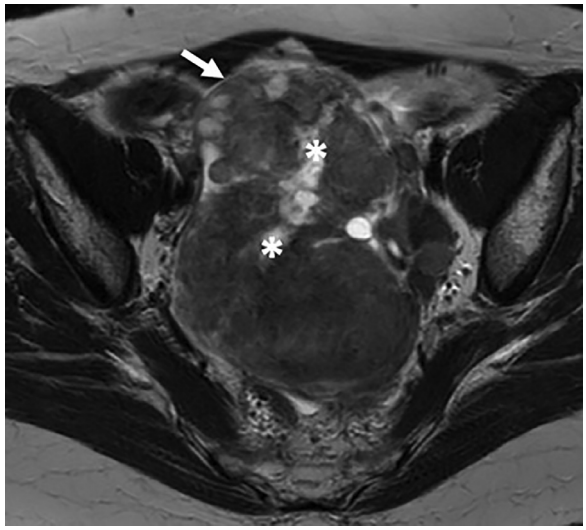

Fig. 2 – Axial T2-weighted image shows a well-defined solid mass (white arrow) located in the pelvis, with intermediate signal intensity and areas of hyperintensity consistent with cystic areas (*).

lar structures [2]. Although rare, schwannomas are one of the possible diagnoses of extra-ovarian masses [3].

Thus, while the immediate recognition of ovarian cancer is essential, it is also important to know how to recognize the processes that mimic ovarian tumors and avoid misdiagnosis. Imaging plays an important role as pelvic masses usually present with nonspecific symptoms and some relevant imaging features may help in the differential diagnosis of an adnexal lesion and which may indicate the origin of the tumor.

Here, we present a case report of a retroperitoneal pelvic schwannoma mimicking ovarian cancer.

Case report

A 60-year-old woman presented with a feeling of pelvic heaviness and dyspareunia for 3 months. On physical examination, a hardened mass is palpated on the cul-de-sac of Douglas. Ultrasound (US) imaging showed a retro-uterine solid mass, containing cystic areas, measuring 14 cm. Uterus had normal size and no other abnormalities were detected. Magnetic resonance imaging (MRI) revealed a solid, ovoid, left tumor, posterior to the uterus, with posterior compression by the sacral promontory (Figs. 1-3), measuring 13.8 × 8.7 × 9.7 cm of greatest diameters. The lesion displayed signs suspicious of malignancy, showing heterogeneous enhancement on contrast-enhanced T1-weighted images (T1WI) (Fig. 4) and restriction on diffusion-weighted images (DWI) and apparent diffusion coefficient (ADC) map (Fig. 5). The right ovary had no appreciable alterations and there was no ascites. A staging computed tomography (CT) reported no distant metastases (Fig. 6). CA125, CA19-9, and CEA were within the normal range.

Considering the diagnosis of a left ovarian malign tumor, the patient was proposed for surgery. A mass was identified in the left pelvis, retroperitoneal, multiloculated, with a predominance of solid components, measuring 20 × 10 cm. The mass was adherent to the bladder anteriorly and to the right fallopian tube laterally. There was no evidence of pelvic organ abnormality, with a normal appreciation of the uterus and ovaries. No pelvic or para-aortic lymphadenopathy was identified, and there was no ascites. Since gynecologic malignancy could not be excluded, the patient was submitted to total hysterectomy and bilateral salpingo-oophorectomy and to excision of the pelvic tumor.

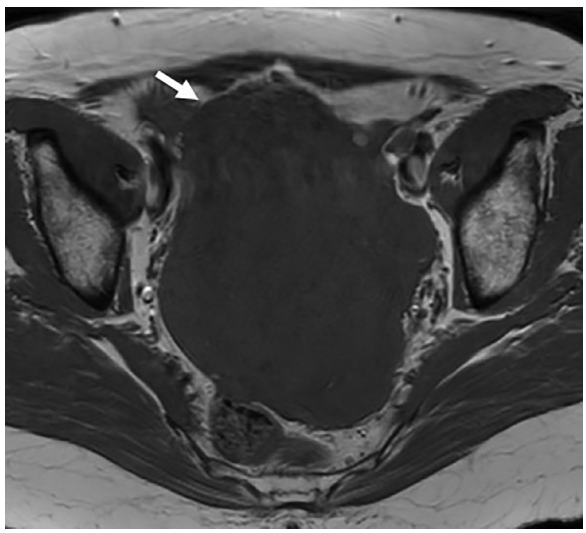

Fig. 3 – Axial T1-weighted image shows a well-defined isointense mass (white arrow) located in the pelvis.

The patient tolerated the procedure well and was discharged on postoperative day 3.

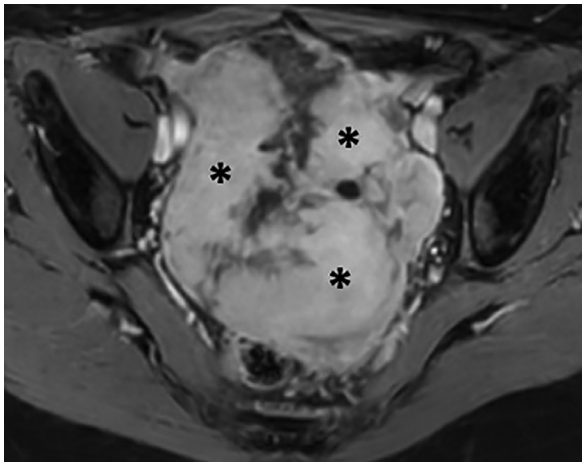

Fig. 4 – Axial dynamic contrast-enhanced T1-weighted image showing enhancement of the solid components (*).

The gross examination revealed a 17 cm yellow-white encapsulated multinodular tumor. The tumor was mostly solid with cystic degeneration in less than 10% of the tumor area (Fig. 7). Histologically, the tumor showed mixed hyper and hypocellular areas (Fig. 8A). The hypercellular areas contained fascicles of spindle cells with rows of nuclear palisading and whorling (Fig. 8B), whereas hypocellular areas presented a loose edematous matrix with cystic change. The spindle cells had no nuclear atypia and surrounded vessels with thickened hyalinized walls (Fig. 8C). Immunohistochemical analysis showed a strong and diffuse S100 expression (Fig. 8D). All these findings were concordant with a schwannoma.

The evaluation 4 weeks after surgery revealed no neurologic deficits and resolution of her pelvic symptoms.

Discussion

Diagnosis of masses in the pelvic region is challenging due to the complex anatomy of the female pelvis and adnexa. Ovaries have variable positions between patients and at different times in the same patient, and, after menopause, the ovaries may be more difficult to identify because of their small size and lack of follicles. In this context, imaging is essential for assessing mass size and location, evaluating the relationship to adjacent pelvic structures, and narrowing the differential diagnoses [1,2].

One of the first steps in diagnosing a pelvic mass is to determine whether the mass is of ovarian or extra-ovarian origin. Some signs suggestive of ovarian origin include the "beak" sign, where an acute angle ("beak") is identified between the ovary and the mass; the "ovarian vascular pedicle" sign, which is the identification of the gonadal vein junction next to the mass; the "ghost organ sign," which describes the absence of an ovary in a patient with no previous surgical history; and the "embedded organ sign," in which the ovary is identified involved by the mass. On the other hand, we can refer to some signs that indicate the extra-ovarian origin, including the absence of the "claw sign," which refers to the sharp angles on either side of the mass, formed by the normal parenchyma when the mass has arisen from the parenchyma; the "bridged vessel" sign on the Doppler study, that indicates the uterine origin of a mass by identifying a vascular point. Fallopian tube disease is identified usually by the tubular nature of the structure.

Another clue is the direction of displacement of organs surrounding the pelvic mass. Intraperitoneal masses tend to displace the uterus, rectosigmoid colon, and iliac vessels laterally or posteriorly; masses in the cul-de-sac of Douglas displace the uterus anteriorly and the rectum posteriorly; and extraperitoneal masses tend to displace anteriorly or centrally the uterus, rectosigmoid colon, and iliac vessels; and erase the pelvic sidewall muscles. It is possible to exclude the ovarian origin of a mass if a normal ipsilateral ovary is identified and/or the precise location of the pelvic mass as an extraperitoneal lesion is possible.

However, the diagnosis on imaging is not always clear, and ovarian and extra-ovarian processes occupying the adnexa can appear as an ovarian malignancy, as in the case reported. The pelvic extraperitoneal region is divided into the interconnected prevesical and perivesical spaces anteriorly and perirectal and retrorectal spaces posteriorly [2]. These spaces are most often involved by direct extension of gastrointestinal

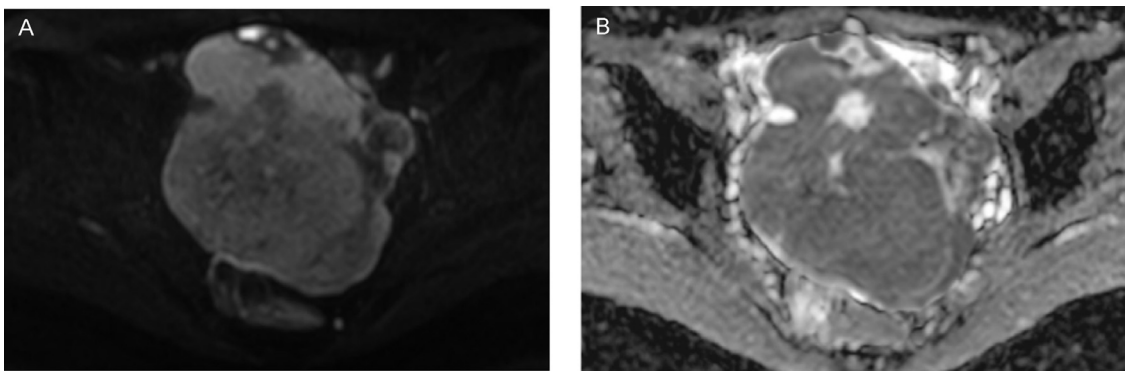

Fig. 5 – Axial diffusion-weighted image b-1000 (A) showing the mass with high signal intensity and apparent diffusion coefficient (ADC) map (B) with low signal intensity, compatible with restriction.

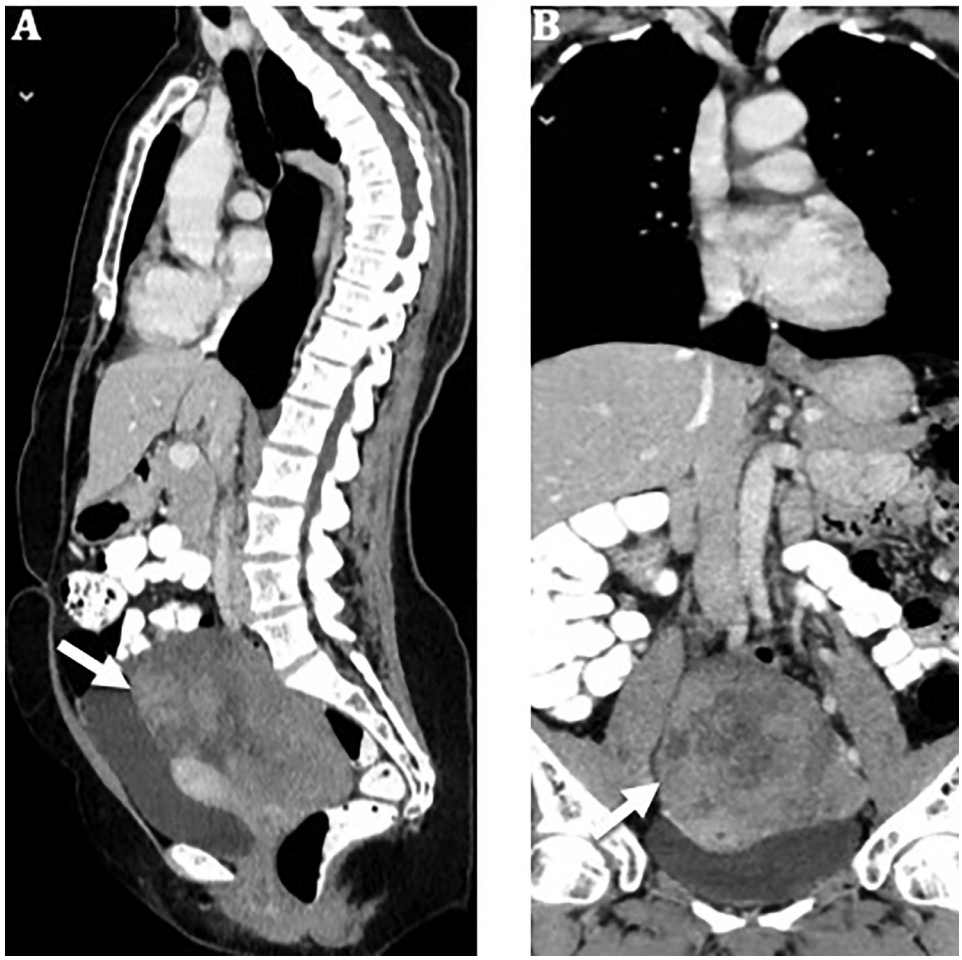

Fig. 6 – Sagittal (A) and coronal (B) contrast-enhanced CT shows a well-defined pelvic mass (arrows). There were no other signs of disease.

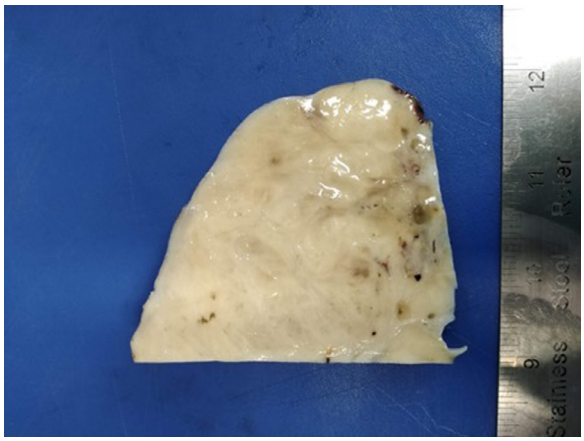

Fig. 7 – Grossly the tumor was mostly solid with scarce cystic degeneration.

or genitourinary tumors. However, primary tumors can also occur, such as neurogenic tumors [2].

Schwannomas are generally benign tumors originating from the Schwann cells that form the myelin sheath around the peripheral nerve [3]. Schwannomas in the retroperitoneal space are rare and mainly located in the pelvis, usually in the paravertebral and presacral region [4]. They are prevalent between 40 and 60 years of age and in female patients [4].

As they are slow-growing tumors, they usually present as solitary and asymptomatic masses [3,4]. When symptomatic, they usually present with compressive symptoms due to the size of the tumor. When located in the pelvis, the symptoms are vague and nonspecific, as in the presented case, and can include abdominal discomfort as well as tightness in the pelvic region and lower back. Neurological symptoms, including pain and paresthesia, have also been reported. Urinary and gastrointestinal symptoms caused by the compression of the urinary bladder and intestine may also occur [3,4]. Calcification is observed in 23% of cases and cystic degeneration in almost 60% of cases [3,4].

Preoperative diagnosis of pelvic schwannomas is extremely challenging due to their non-specific imaging characteristics [4]. It is an encapsulated well-defined nerve tumor. On CT, this tumor may contain calcifications. On MRI, the tumor presents with a low signal on T1WI, a high signal

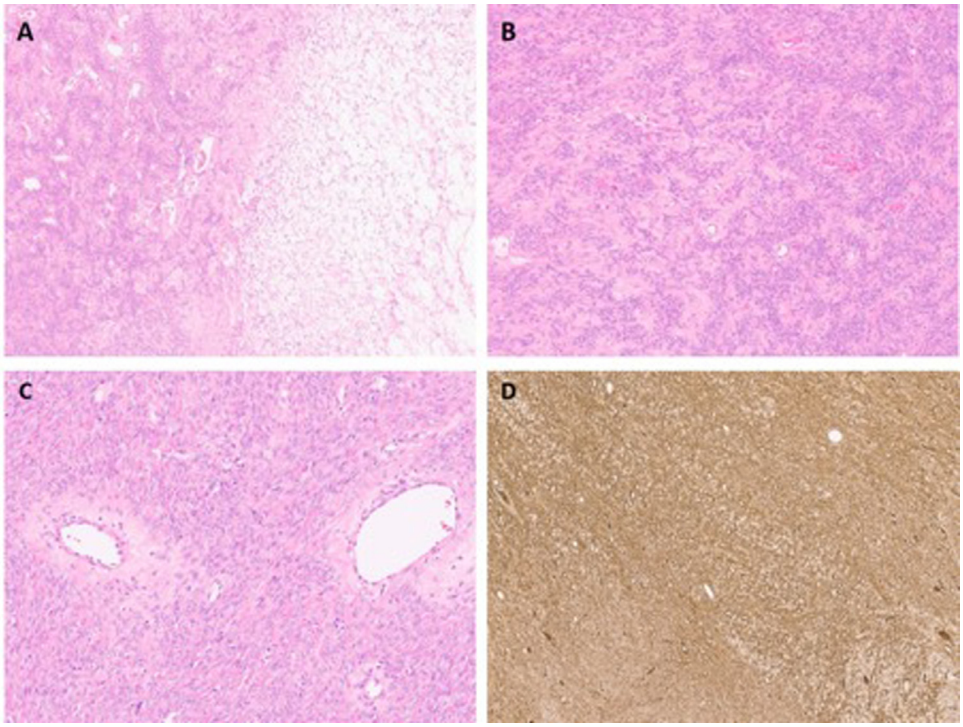

Fig. 8 – (A) Hypercellular areas and hypocellular areas with cystic change (H&E, ×40); (B) Fascicles of spindle cells with nuclear palisading and whorling (H&E, ×100); (C) Hyalinized vessel walls (H&E, ×200); (D) Strong and diffuse S100 expression (×40).

on T2-weighted images (T2WI), and heterogeneous contrast enhancement on postcontrast T1WI. When large, schwannoma can contain regions of cystic necrosis or hemorrhage [5].

The US is often the first imaging modality performed to characterize the adnexa, although it has significant limitations [2]. MRI is the preferred imaging modality for pelvic masses, as it provides multiplanar evaluation and better soft-tissue contrast [2]. CT also has limited value in the pelvis owing to its inferior soft-tissue contrast [2].

As such, MRI provides a better tissue analysis than CT and might be the superior imaging modality for the preoperative evaluation of schwannomas [1,2]. For the presented case, since the MRI suggested a possible ovarian malignancy, a CT scan was obtained later for staging. Although MRI has better visualization, in our case report the differentiation of a schwannoma from other ovarian mass was not possible, which might be due to the difficult identification of the normal left ovary in this postmenopausal patient and the absence of certain characteristic radiological signs of schwannoma in extraperitoneal location, such as anterior or lateral displacement of the rectosigmoid colon and iliac vessels. In situations where the diagnosis of a pelvic mass is uncertain, a combination of multiple imaging modalities with a needle biopsy might be necessary [3]. Since the pre-surgical diagnosis of a Schwannoma is difficult due to its uncommonness and the absence of distinguishing features by different imaging methods, these tumors tend to masquerade as another lesion, with definitive diagnosis rendered only after histopathologic examination, as in the case described.

Conclusion

The complex anatomy and similarity of imaging features of various pathologies in the pelvis can make accurate radiology interpretation difficult. While prompt recognition of ovarian cancer remains essential, awareness of processes that mimic ovarian tumors avoids potential misdiagnosis and unnecessary surgery.

This rare case of schwannoma highlights a few points to consider when evaluating and managing a pelvic lesion. It is vital to have appropriate knowledge of the pelvic spaces and to assess the most likely site of origin of the mass considering the diagnostic imaging clues for an ovarian or extra-ovarian tumor and its location within the peritoneal cavity versus the extraperitoneal spaces. Contrast-enhanced MRI is the best modality for the characterization of common tumors or tumor mimics in the female pelvis, providing information essential to narrow the differential diagnosis or to suggest the correct diagnosis.

Patient consent

The patient's informed consent for the publication of this case was granted. There are no ethical issues for the publication of this case report according to the standard of our institution.

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