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Retroperitoneal schwannoma: A rare cause of pelvic mass

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ABSTRACT

Introduction: Schwannomas are rare, usually benign, tumors that arise from the myelin sheath of nerves. Only 1% of schwannomas are localized in the retroperitoneal space. Preoperative diagnosis can be difficult as symptoms are non-specific.

Case Report: We present the case of a 32-year-old woman who presented with suspected appendicitis. A lateral uterine mass was discovered on imagery, and was surgically resected. Schwanomma was diagnosed on histopathological and immunohistochemical analysis with intense staining of the S100 protein. The resected mass had healthy margins. The patient reported neurological disorders three weeks after surgery which improved with physiotherapy.

Conclusion: This case illustrates the difficulties of diagnosing schwannoma and supports the relevance of a surgical approach. Complete surgical resection can prevent recurrence or worsening, and is recommended in despite the risk of morbidity.

Keywords: Laparoscopy, Lateral-uterine mass, Neurilemmoma, Retroperitoneal space, Schwannoma

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INTRODUCTION

Schwannomas, or neurilemmomas, are rare tumors originating from the neural sheath of peripheral nerves [1]. They usually present in the head and neck or in the upper extremities. Retroperitoneal pelvic origin is uncommon; only 1–3% of schwannomas are found in the retroperitoneal space [2–6]. They represent about 1% of all retroperitoneal tumors [4]. More than 90% of schwannomas are benign [2, 6]. Very rarely, malignant degeneration may occur [7], and is usually associated with the absence or alteration of the neurofibromin 2 (NF2) gene [1], found in 5–18% of these cases [5]. Preoperative diagnosis is often difficult because the symptoms are non-specific and depend on the location and size of the lesion [8].

Here we report a case of a young woman who was misdiagnosed with a lateral uterine mass. She underwent laparoscopic resection of a retroperitoneal schwannoma and the final diagnosis was established by histopathological examination. We present its radiographic and histological features.

CASE REPORT

A 32-year-old woman presented with pain in the right iliac fossa suggestive of appendicitis. A computed tomography (CT) scan revealed an adnexal mass. Initially, the CT scan described a round and well-demarcated 37×44 mm lesion, presenting an inhomogeneous structure with a hyperdense central zone and low-density peripheral zone located in the right para-ovarian fossa that suggested a hemorrhagic cyst. She underwent exploratory laparoscopy because of persistent pain. During surgery, the right ovary was described as “polycystic of banal appearance.” There was no description of the retroperitoneal space in the operative report.

Ten months later, she reported persistent pain in the right iliac fossa, especially during sexual intercourse. Clinically, the abdomen was supple and painless. The vaginal examination was normal, and nothing was apparent on speculum examination. On ultrasound, a septum cyst was visualized and a pelvic magnetic resonance imaging (MRI) exam was consequently performed. A macronodular formation measuring 41×56 mm with mixed tissue classified O-RADS 4 was found in the right ovary. A central fluid compartment was measured at 38 mm with T1 hypointensity, T2 hyperintensity, and contrast enhancement after injection. Magnetic resonance imaging also showed the absence of dilation of the excretory urinary tract, of retroperitoneal adenomegaly, of pelvic lymphadenopathy, and of intraperitoneal effusion (Figure 1).

The patient was referred to the University Hospital of Caen for an examination by a specialist. A second ultrasound was performed and found normal ovaries and a right lateral uterine mass of 40×40×50 mm suggestive of a pedicled sub-serous myoma. A second opinion was sought to interpret the MRI. They described the image as a non-suspicious, right juxta ovarian tissue formation, independent of the uterus, and suggested a right Fallopian tube lesion or ovarian fibroma.

After discussion, the patient underwent a second surgery by laparoscopy to excise the mass. The mass was located in the right parametrium between the ureter and the external iliac vessels. The uterus and the ovaries were normal. The resected mass presented an inferior and superior pedicle which resembled an adenomegaly. The operative time was 120 min and the bleeding volume was less than 100 cc. The patient was discharged the day following the operation. No perioperative complications were reported.

The surgical mass weighed 25 g, measured 50×35×20 mm, and was reshaped by a lesion of 40 mm with a long axis, well limited with an enucleated appearance. When cut, the lesion was white with a very discreet hemorrhagic center. On histological analysis, it was found to be formed by cell proliferation arranged in a short, crisscrossed bundle type pattern. Proliferating cells were fibrillar in appearance with a rather elongated, spindle-shaped nucleus with fine chromatin, and without visible nucleolus. More irregular-looking nuclei were numerous and were discreetly pleomorphic but without major cytonuclear atypia. The mitotic index was low with less than one mitosis per 10 fields at high magnification (×400). The cell proliferation was traversed by vessels of variable size, with a hyalinized wall. No true Verocay nodule was observed, but cell proliferation sometimes of a discreet palisade aspect. No necrosis was observed on the macroscopic sample taken. Expression of tumor receptors was positive for PS100 and negative for caldesmon, CKAE1/AE3, desmin, estrogen, and progesterone receptors. Finally, it was classified as a schwannoma of 5 cm with complete excision (Figure 2).

During the postoperative examination three weeks later, the patient presented neurological disorders including inability to maintain a standing position on the right leg, motor deficit of extension of the right foot and elevation of the knee, and pain and dysesthesia on the inner side of the right thigh. These disorders can be explained by involvement of a branch of the obturator nerve during excision, even though the obturator nerve was seen in full and spared during surgery. Physiotherapy was prescribed and an improvement was observed during the follow-up consultation. A CT scan will be performed at four months to rule out recurrence.

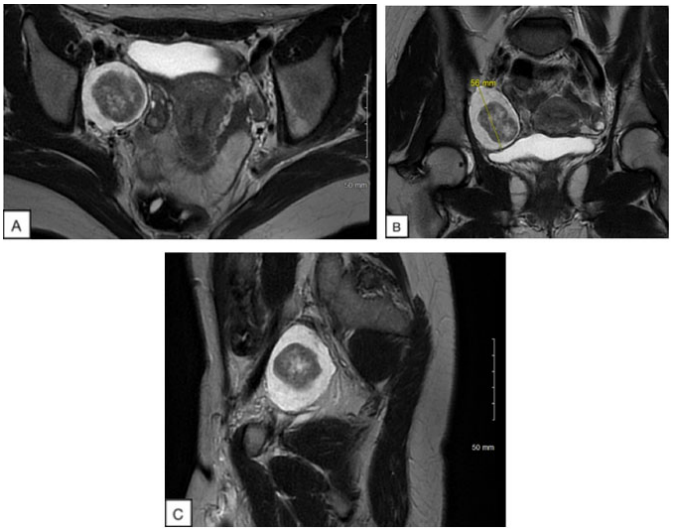

Figure 1: MRI: T2. (A) Axial. (B) Coronal. (C) Sagittal. Presence of a central compartment measured at 38 mm with T2 hyperintensity and contrast enhancement after injection.

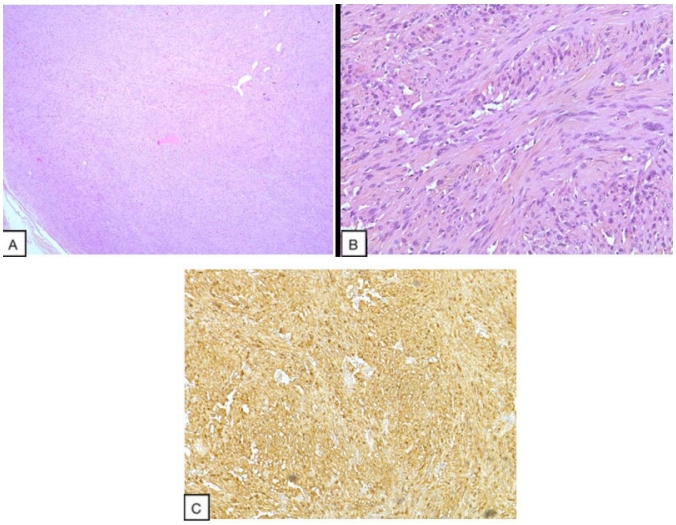

Figure 2: Histology: (A) HES coloration ×20, (B) HES coloration ×200, (C) Immunohistochemistry for S100 ×100.

DISCUSSION

Schwannomas are mesenchymal tumors that arise from Schwann cells of peripheral nerve sheaths. They

most commonly arise from peripheral nerves in the head, neck, and the upper extremities, but also from spinal and cranial nerves, especially the vestibular nerve [7]. Schwannomas are unusual in the digestive tract and even less common in the mesentery or retroperitoneum (1–3% of total), with few published cases [9], and even rarer in the pelvis [8, 10–13].

The disease occurs in individuals of all ages, with most cases diagnosed between the ages of 20 and 60 [6, 13–15]. It was thought to affect men and women equally [16], but a slightly higher incidence had been reported in women [6, 7]: Hughes et al. found a male/female ratio of 0.625:1 [17]. However, this is in contrast to the findings of Kransdorf in a large population-based analysis of 895 schwannomas including tumors in all locations [18]. Other authors have described a female predominance for benign retroperitoneal schwannoma [4, 5, 10].

Preoperative diagnosis is challenging, and a definitive diagnosis can only be made postoperatively by histopathological and immunohistochemical analysis of surgical specimens. Verocay first described peripheral nerve tumors histologically in 1908 [19]. Schwannomas show intense immunohistochemical staining for the S100 protein, confirming the neuroectodermal origin of the tumor cells [20]. In 1920, Antoni et al. classified them into two distinct histological patterns: compact cellular areas called Antoni A areas; and paucicellular areas called Antoni B areas [21]. Schwannomas are thus basically encapsulated biphasic nerve sheath tumors derived from Schwann cells with highly ordered cellular component (Antoni A) in a palisading pattern (Verocay bodies), plus a myxoid component (Antoni B). Small tumors may be completely Antoni A. Immunohistochemistry (IHC) is a useful technique for detecting tumors originating from Schwann cells because they stain positive for S100 antigen, collagen IV and laminin, and show an absence of reactivity for keratin, desmin, and vimentin [22].

They can either be isolated as sporadic lesions, or associated with schwannomatosis or NF2 (1). The NF2 tumor suppressor gene encodes for a protein called merlin or schwannomin [23]. Somatic inactivation of both alleles of this gene occurs in sporadic schwannomas (>90%) [23].

Schwannomas are usually slow-growing tumors [1]. The retroperitoneum, being non-restrictive, allows benign tumors, such as schwannomas, to grow to a large size before causing symptoms, which are usually non-specific [2, 24]. Benign schwannomas are not infiltrative but can cause compression of the adjacent structures and produce symptoms such as lumbar pain and neurological symptoms in the lower extremities [1], and renal colic pain, with or without hematuria if the urogenital tract is involved [25]. Abdominal complaints can also occur but are mainly vague and poorly localized, with some digestive disorders [1]. Tumor size does not appear to be associated with the degree of aggressiveness or malignant potential [14].

Preoperative diagnosis of retroperitoneal schwannoma is difficult because of the lack of specific symptoms and difficulties in detecting characteristic features on imaging [3, 10]. As in our patient, they may be incidentally discovered [5, 18, 20].

The characteristic, but non-specific, features of schwannomas on CT and MRI findings include a well-demarcated, homogeneous, spherical, solitary mass [2]. On MRI, benign schwannomas have smooth margins and are isointense with muscle on T1-weighted images and markedly hyperintense on T2-weighted images [15, 24]. We found the same characteristics in our case. However, these findings have been reported in only 57% of the cases in previous studies [26] and are what would be expected from any mass with extensive areas of necrosis [3]. Imaging, usually in the form of ultrasonography and CT, is helpful to estimate the size, location, presence of invasion, and involvement of surrounding organs. Some authors report difficulties in localizing the mass in the retroperitoneum, even confusing a large cystic retroperitoneal schwannoma as a pseudocyst [27]. As in our case, misdiagnosis is frequent on MRI [22]. Overall, schwannomas should be included in the differential diagnosis of any retroperitoneal abdominal mass: they account for approximately 4% of primary retroperitoneal tumors (paraganglioma, ganglioneuroma, neurofibroma, fibrous histiocytoma, liposarcoma, and lymphoma) [3].

Misdiagnosis of schwannomas is common. In a series of 82 cases, Li et al. reported that a correct preoperative diagnosis was made in only 13 (15.9%) of the patients [6], and a review paper found that preoperative diagnosis was incorrect in all the cases included [28]. The diagnosis of retroperitoneal schwannomas is often delayed and misdiagnosed as an adnexal tumor, as in our patient, especially due to the location in the pelvic cavity [13]. Other case reports have described rare cases of schwannoma presenting as an adnexal mass [11], a mass in the paracolpium [29], or mimicking uterine myoma [12].

Computed tomography-guided core biopsy and fine-needle aspiration have been found to be unreliable for the diagnosis of retroperitoneal schwannoma [5, 24, 25]. Ultrasound-guided fine-needle aspiration was used in only one case of Li et al.'s series of 82 patients to confirm diagnosis [6].

Our patient's lesion was well-limited. On gross appearance, schwannomas are well-demarcated, firm, solid tumors with a smooth surface and have an ovoid or spherical shape [5, 24]. Secondary degenerative changes such as hemorrhage, cysts, calcification, and hyalinization can sometimes be present [1, 30]. The largest review of retroperitoneal schwannomas (133 cases of retroperitoneal schwannomas, both benign and malignant) reported that cystic appearance may be one of the cross-sectional features of retroperitoneal schwannomas, because 63% of benign and 75% of malignant schwannomas demonstrated cystic changes,

whereas other types of retroperitoneal tumors do not usually show such changes [30].

Surgical resection with negative margins is recommended to completely remove the tumor and thus prevent recurrence [7] or possible malignant degeneration, and to establish a definitive histological diagnosis [20]. Although complete resection may be associated with a higher morbidity (for example, neurological disorders [24] as in our case) than partial resection, one must consider the possibility of recurrence [20].

Prognosis is usually good and recurrence is very rare. Incomplete resection may result in local recurrence because of the strong connection with surrounding tissue. Previous reports have demonstrated that incomplete excision leads to local recurrence in approximately 10–20% of patients [30]. For Li et al., only one out of 82 cases had a local recurrence three years after surgery in the group of patients who had undergone sub-total resection [6]. No recurrence occurred in the patients receiving complete resection and the overall low recurrence rate was similar to that of 1–3% reported by others [6]. No cases of metastasis have been reported [12].

Other treatments such as chemotherapy or irradiation appear to be ineffective [11, 30].

CONCLUSION

Despite the rare occurrence of schwannomas, pelvic retroperitoneal nerve sheath tumors should be included in the differential diagnosis of an adnexal mass. Other diagnoses such as paraganglioma, ganglioneuroma, neurofibroma, and retroperitoneal malignancies, such as malignant fibrous histiocytoma, liposarcoma, and lymphoma, should also be considered. Complete surgical resection of the tumor is recommended to prevent recurrence or possible malignant degeneration. The patient should be warned of the risk of neurological deficits in relation to the area affected by the schwannoma.

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Lisa Godet – Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Anne-Cécile Pizzoferrato – Design of the work, Analysis of data, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Dylan Owen – Analysis of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Stanislas Mulot – Design of the work, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Raffaèle Fauvet – Conception of the work, Acquisition of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.