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Case Report

Retroperitoneal pelvic schwannoma: A rare case report and review of the literature ☆,☆☆

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ABSTRACT

Schwannoma, typically a non-cancerous tumor originating from Schwann cells, seldom occurs in the retroperitoneal area. Its clinical manifestation varies, often remaining asymptomatic for an extended period until it enlarges, exerting pressure on neighboring organs. This article presents a unique instance of retroperitoneal pelvic schwannoma in a 75-year-old woman, initially presenting with unusual lower back discomfort. Imaging was used to characterize the tumor, and anatomopathological examination established the preoperative diagnosis of pelvic schwannoma and its benign nature. The clinical, imaging, and anatomopathological aspects of this pelvic schwannoma case are elucidated herein.

Introduction

Schwannomas, typically benign tumors originating from Schwann sheath cells, primarily affect cranial and upper extremity nerves [1]. Their occurrence in the retroperitoneal area is exceedingly rare [2], constituting approximately 0.3% to 3% of all schwannomas [3]. Pelvic localization is even more uncommon. Pelvic schwannomas remain asymptomatic for a long time, revealing only non-specific signs such as abdominal pain, constipation, or venous thrombosis. While schwannomas are generally benign, malignant variants are more commonly associated with Von Recklinghausen syndrome (present in 4% of cases) or other neurofibromatoses [4]. We report a case of a pelvic retroperitoneal Schwannoma with review of the literature.

☆ Author contribution: All the authors contributed to study concept, data analysis, and writing the paper.

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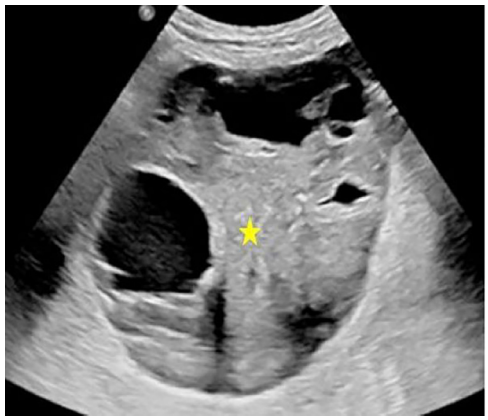

Fig. 1 – Pelvic ultrasound reveals a large, well-defined and heterogeneous mass (★) with a cystic and solid component.

Clinical case

A 75-year-old women, presented with an eight-month history of continuous lower back pain independent of activities with no particular irradiation or neurovascular deficits.

Physical examination demonstrated a lower abdominal mass.

Abdomino-pelvic ultrasound (Fig. 1) showed a retro-uterine pelvic mass with regular borders. The mass exhibited a dual nature, comprising anechoic cystic areas and isoechoic tissue. Notably, it was observed to be separate from both the uterus and rectum.

Pelvic CT and MRI (Figs. 2 and 3) showed a large retro-uterine pelvic mass, predominantly located on the left side. The mass appeared oval-shaped with clearly defined borders and was characterized by the presence of calcifications and extensive fluid-filled areas. It exhibited heterogeneous enhancement upon imaging and measured approximately 132 × 126 × 103 mm. This mass was observed to be in close proximity to the uterus, exerting pressure on nearby digestive

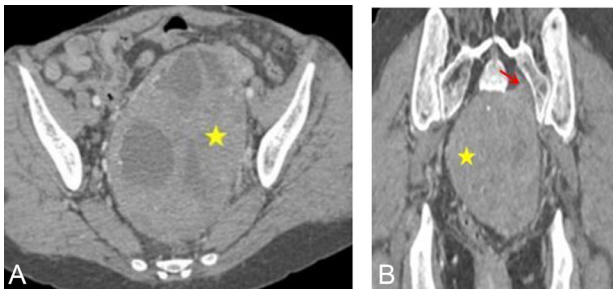

Fig. 2 – Contrast-enhanced computed tomography in axial (A) and coronal (B) sections showing a large solid-cystic mass (★) arising from sacrococcygeal region which appears to impinge on the left first sacral foramen (red arrow).

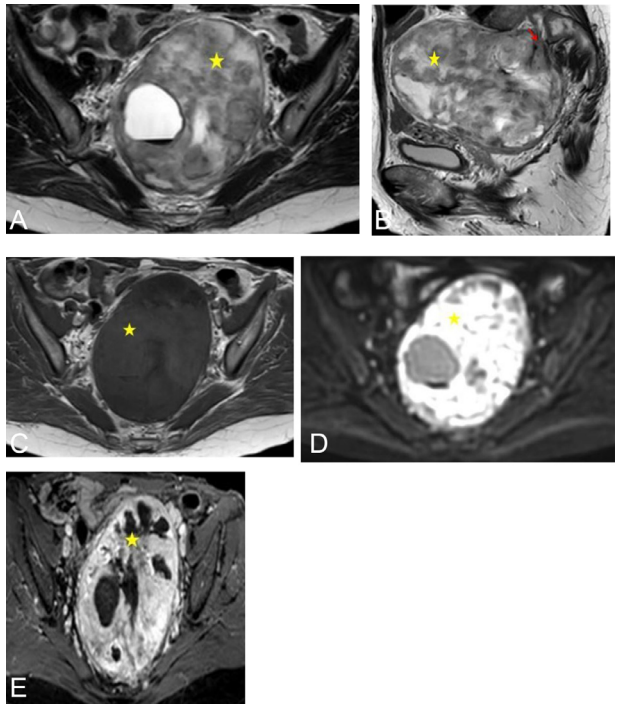

Fig. 3 – Injected pelvic MRI showing a voluminous retroperitoneal mass (★), well encapsulated, occupying the sacral concavity, in T1 iso signal (A), in T2 heterogeneous signal (B and C), with a fluid portion in hypersignal and a tissue portion in intermediate signal which appears to impinge on the left first sacral foramen (red arrow), restrictive to diffusion (D) and heterogeneously enhanced after injection of Gado (E).

structures without signs of invasion. Notably, it appeared to extend into the left first sacral foramen, causing lateral displacement of the iliac vessels, and posterior displacement of the rectum.

Biological tests revealed normal levels of tumor markers, including CA19-9 antigen (<2U/mL for a normal of <33) and CA 125 antigen (5.20 U/mL for a normal of <36) (Table 1).

During explorative laparotomy, a retroperitoneal well-defined mass was identified, characterized by a dual-component structure. The mass enveloped the right internal iliac artery and right ureter. Subsequently, a surgical biopsy was carried out for further evaluation.

Histological examination (Fig. 4) revealed spindle-cell proliferation, exhibiting both hypo- and hyper-cellular, organized in diffuse sheets. The tumor cells displayed a spindle-shaped morphology, with elongated, slightly anisokaryotic nuclei and fine chromatin. The fibrous tumor stroma contains numer-

Table 1 – Results of biological tests.

Biological Tests	Results	Standard Values
CA19-9 antigen	<2U/mL	<33 U/mL
CA 125 antigen	5.20 U/mL	<36 U/mL

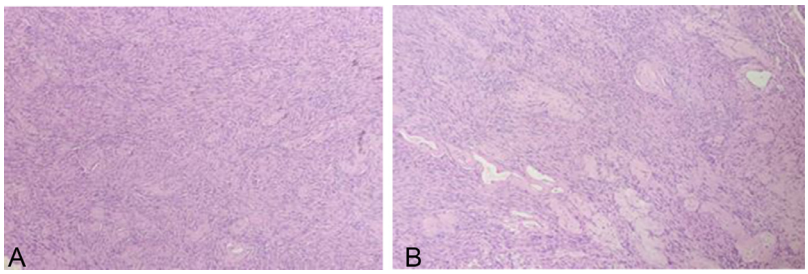

Fig. 4 – Histopathological morphology showing on standard coloration a spindle-shaped proliferation of cells without cyto-nuclear atypia, with numerous hyalinized vessels.

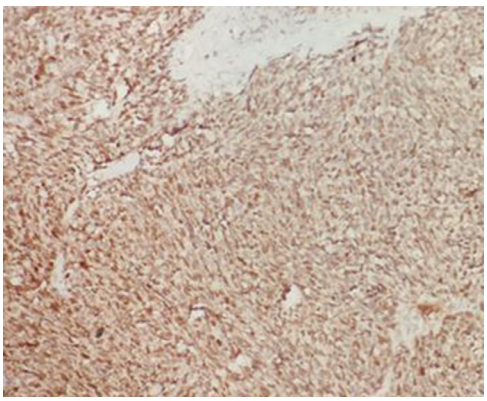

Fig. 5 – Immunohistopathology showing a positive staining of tumor cells by anti-pS100 antibody.

ous hyalinized vessels. Immunohistopathology as depicted in (Fig. 5) demonstrated positive staining of tumor cells by anti-pS100 antibody suggesting a benign schwannoma.

Discussion

Schwannoma is a nerve tumor arising from Schwann sheath cells [5,6], seen mostly in women (male/female ratio 2:3) between the 3rd and 5th decade. Often benign in 97%-98% [7,8], presenting as a single or sporadic form. The rarer malignant form is often associated with neurofibromatoses (Von Recklinghausen disease) [4], as well as the multiple form.

Schwannomas have the potential to develop in any part of the body, with most frequently the limbs in 53.1% of cases, the trunk : 13%, and the head and neck : 13.9% [9].

Retroperitoneal schwannoma is indeed rare [10,11], constituting only 3% of all schwannomas and 4% of primary retroperitoneal tumors [10,12]. Specifically, this retroperitoneal localization accounts for merely 0.7% of benign schwannomas and 1.7% of malignant schwannomas [10,11]. Additionally, pelvic schwannomas are even rarer, with an estimated prevalence ranging from 0.3% to 3.2% across all localized occurrences [13]. It can involve the organs of the pelvis, including the external genitalia [14]. Pelvic schwannomas develop mainly posteriorly in the presacral space, originating in the sacral root or hypogastric plexus. When the tumor starts laterally, it originates from the sciatic, femoral, or obturator nerve [14,15].

The discovery of pelvic schwannoma is most often fortuitous and late, given the latency of the tumor's evolution. It usually manifests as non-specific symptoms associated with compression of neighboring organs: abdominal distension, vague abdominal pain [16,17], constipation, but also deep vein thrombosis [18] or ureteral compression [19]. The absence of specific signs makes the diagnosis of retroperitoneal schwannoma a diagnosis of exclusion.

Imaging methods for retroperitoneal schwannomas include ultrasound, CT, and MRI. Abdominal ultrasonography is the first-line investigation due to its promptness, cost-effectiveness, and reproducibility. IT is useful for determining the solid or cystic nature of the mass [10].

Abdominal CT usually shows an encapsulated, solid mass when small, although hemorrhagic and necrotic remodeling is frequently found when larger [19]. Intratumoral cysts are observed in 63% of benign schwannomas and in 75% of malignant schwannomas; this is a major argument, as retroperitoneal tumors rarely present with intratumoral cysts [2]. Calcifications are also possible, an argument in favor of degeneration [20].

MRI is an important diagnostic method for this type of tumor, as it can better demonstrate cystic degeneration of the tumor, define margins, identify the neuronal point of origin and determine the invasive nature [21]. Schwannomas are characterized by the association of a T1 hyposignal identical to adjacent skeletal muscle, and a T2 hypersignal similar to fat [22,23]. These findings are characteristic but not specific for schwannomas, and are present in only 57% of cases. MRI also helps to clarify the relationship with neighboring pelvic organs (sacrum, rectum, and bladder), which is an important consideration for the surgical procedure. MRI is unable to formally distinguish benign from malignant schwannomas [7]. Certain signs are associated with malignancy, such as irregular contours, a heterogeneous appearance and a long axis >5 cm [24]. In our case, the tumor was well limited, with regular contours, respecting the surrounding structures, and showing cystic degeneration, but the histology confirmed its benignity.

The differential diagnosis of Schwannoma includes neurofibroma, paraganglioma, pheochromocytoma, liposarcoma, malignant fibrous histiocytoma, and hematoma [25].

The diagnosis of Schwannoma is suggested preoperatively in 1 case out of 3 [15]. Diagnosis of certainty is based on histology.

Percutaneous biopsy using scannography or ultrasound is not recommended by most authors, given the difficulties of interpretation, the risk of neoplastic dissemination in the case of malignant tumors and peri-tumoral hyper-vascularization [5,6], or the risk of bleeding and infection. Nevertheless, surgical excision is recommended in view of the histological heterogeneity of retroperitoneal tumor [26].

Pathologically, there are 2 histological types: Antoni A is characterized by organized structures with compact, elongated cells, while type B is characterized by disorganized structures with sparse cells [27]. The expression of S100 protein in immunohistochemical studies is indicative of neurectodermal differentiation, and its positivity therefore points to a schwannoma [28,29].

Surgical excision is the gold standard of treatment, and is sometimes difficult because of its intimate relationship with the large vessels and noble organs. Recurrence is rare if excision is incomplete, and malignant transformation of a benign Schwannoma is exceptional; only one case has been described at a distance from excision of a benign Schwannoma [30], and this risk of recurrence and degeneration, even if low, makes annual CT surveillance necessary.

Although schwannomas are essentially benign, there are some malignant forms whose histological nature is controversial due to their frequent association with Von Recklinghausen syndrome (4% of cases) and other neurofibromatoses [4]. Some authors consider them to be neurofibrosarcomas [20].

Malignant schwannomas are treated surgically, as they respond poorly to radiotherapy or chemotherapy [30].

Metastases are mainly to the liver, lungs, bones, and subcutaneous tissues [31]. Lymphatic dissemination is very rare. The association of malignant schwannoma with neurofibromatosis is a pejorative factor, since the 5-year survival rate drops from 47% to 23%.

Conclusion

Retroperitoneal schwannoma is a rare tumor with a good prognosis in its benign form. Its clinical diagnosis is often delayed due to a borrowed symptomatology. Paraclinical examinations clarify its retroperitoneal origin and evaluate the possibilities of excision. Radical surgery is the treatment of choice. However, because of the risk of recurrence, or even malignant transformation, subsequent surveillance is necessary.

Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

REFERENCES

[1] Choudry HA, Nikfarjam M, Liang JJ, Kimchi ET, Conter R, Gusani NJ, et al. Diagnosis and management of retroperitoneal ancient schwannomas. World J Surg Oncol 2009;7(12):1–5.

[2] Takatera H, Takiuchi H, Namiki M, Takaha M, Ohnishi S, Sonoda T. Retroperitoneal schwannoma. Urology 1986;28(6):529–31.

[3] Strauss DC, Qureshi YA, Hayes AJ, Thomas JM. Management of benign retroperitoneal schwannomas: a single-center experience. Am J Surg 2011;202(August (2)):194–8.

[4] Jow W, Satchidanand S, Spninazze E, Lillie D. Malignant juxtadrenal schwannoma. Urology 1991;4:383–6.

[5] Krid M, Elleuch A, Gouchem M, Hbacha M, Ben Sorba N, Tlili Graies K, et al. Schwannome rétropéritonéal, démarche diagnostique et thérapeutique. Acta Urol Belg 1997;65(1):69–72.

[6] Weber O, Zerbib M, Peyret C, Deslignères S, Debré B. Schwannome bénin rétropéritonéal. J Urol 1993;99(4): 189–191.

[7] Li Q, Gao C, Juzi JT, et al. Analysis of 82 cases of retroperitoneal schwannoma. ANZ J Surg 2007;77:237–40.

[8] Habek D, Habek JC, Sindik N. Pelvic schwannoma. Eur J Obstet Gynecol Reprod Biol 2004;116:248–9.

[9] Ben Moualli S, Hajri M, Ben Amna M, Kolsi K, Chebil M, Ben Jilani S, et al. Le schwannome rétropéritonéal. À propos d'un cas. Ann Urol 2001;35:270–2.

[10] Garcia G, Anfossi E, Prost JT, Ragni E, Richaud C, Rossi D. Schwannome rétropéritonéal bénin : à propos de trois cas. Progrès en Urologie 2002;12:450–3.

[11] Aubert J, Debiais F, Levillian P. Schwannoma of the obturator nerve with urologic manifestations: report of a case. Ann Urol 2000;34:58–65.

[12] Pennington Z, Westbroek EM, Ahmed AK, Cottrill E, Lubelski D, Goodwin ML. Surgical management of giant presacral schwannoma: systematic review of published cases and meta-analysis. J Neurosurg Spine 2019;5:1–12.

[13] Khatig RA, Khalil AM, Saba MI, Aswad NK, Mroueh AM. A pelvic retroperitoneal schwannoma presenting as an adnexal mass. Gyn Oncol 1994;53(2):242–4.

[14] Fauchery A, de Meeus JB, Turc I, Bascou V, Goujon JM, Magnin G. Le schwannome bénin pelvien. J Gynécol Obstet Biol Reprod 1994;23(3):279–82.

[15] Schindler OS, Dixon JH. Retroperitoneal giant schwannomas: report on two cases and review of the literature. J Orthopaed Surg 2002;10(1):77–84.

[16] Li ZQ, Wang HY, Li J, Teng L. Recurrent retroperitoneal Schwannomas displaying different differentiation from primary tumor: case report and literature review. World J Surg Oncol 2010;8:66.

[17] Harada TL, Nagao G, Aoyagi T, Kuroda I, Tokuyama N, Takahashi M, et al. Giant retroperitoneal schwannoma in a 52-year-old man. Radiol Case Rep 2018;13(4):810–14 août.

[18] Micali S, Virgili G, Vespasiani G, Silecchia A, D'Alessandro P, Micali F. Benign schwannoma surrounding and obstructing the ureteropelvic junction. Eur Urol 1997;32:121–3.

[19] Donnal JF, Baker ME, Mahony BS, Leight GE. Benign retroperitoneal schwannoma. Urology 1988;4:332–4.

[20] Goh BK, Tan YM, Chung YF, et al. Retroperitoneal schwannoma. Am J Surg 2006;192:14–18.

[21] Hughes MJ, Thomas JM, Fisher C, Moskovic EC. Imaging features of retroperitoneal and pelvic schwannomas. Clin Radiol 2005;60(8):886–93.

[22] Hayasaka K, Tanaka Y, Soeda S, Huppert P, Claussen CD. MR findings in primary retroperitoneal schwannoma. Acta Radiol 1999;40(1):78–82.

[23] Yu Y, Wu J, Ye J, Chen M. Radiological findings of malignant peripheral nerve sheath tumor: reports of six cases and review of literature. World J. Surg. Oncol. 2016;14(1):1–8.
[24] Harkin I. Pathology of nerve sheath tumors. Ann NY Acad Sci 1986;486:147–54.
[25] Rossi D, Delpero JR, Jacquemier J, Resbeut M. Tumeurs rétropéritonéales primitives. In: Néphrologue-urologue. Paris: EMC; 1993. p. 10. 18-083-A-10, Cancérologie, 60-18-250-A-10.
[26] Hide IG, Baudouin CJ, Murray SA, et al. Giant ancient schwannoma of the pelvis. Skelet Radiol 2000;29:538–42.
[27] Weiss SW, Langloss JM, Enziger FM. Value of S-100 Protein in the diagnosis of soft tissue tumors with particular reference to benign and malignant Schwann Cell tumors. Lab. Invest 1983;49:299–308.
[28] Yamamoto K, Miyagawa J, Katsura H. Retroperitoneal cellular schwannoma : report of a case diagnosed by the presence of S-100 protein. JPN J. Med. 1991;30:487–90.
[29] Das Gupta TK, Brasfield RD, STrong EW, Hajolu SZ. Benign solitary schwannomas. Cancer 1969;24:355–8.
[30] Zhang G, Chan K, Marrivel C, Fraley E. Sarcomas of the retroperitoneum and genitourinary tract. J. Urol 1989;141:1107–10.
[31] Saito R, Ishizuka E, Iwasaki A, Kobayashi K. Retroperitoneal malignant schwannoma in the pelvis: a case report. Hinyokika Kiyo 1997;43:25–8.