Pelvic retroperitoneal schwannoma presenting as a gynecologic mass: Case report

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Summary

Schwannomas of the sacral plexus are retroperitoneal tumors, usually benign, that result from proliferation of perineural cells. They are rare, pelvic, well defined tumors that present infrequently as gynecologic masses. Misdiagnosis is not uncommon in these cases, since these masses are not often encountered in clinical practice and preoperative imaging methods can be only suggestive of the diagnosis. Furthermore, these masses are not often encountered in clinical practice. We report a case of a 28-year-old woman who presented with a pelvic mass and obscure clinical signs. Imaging methods suggested that this tumor was most probably an ovarian mass of embryonic origin. After dissection by laparotomy, the microscopic examination showed typical futures of a neurilemoma (ancient schwannoma). The goal of the operating intervention is to excise the tumor avoiding major trauma, thus cooperation between surgeons is indispensable. The patient has had no motor or sensory disturbances after surgery.

Key words: Pelvic mass; Schwannoma; Diagnosis; Resection.

Introduction

Benign nerve sheath tumors are pseudoencapsulated tumors that arise from the neoplastic transformation of nerve sheath cells [1]. They are classified into two major categories, neurilemomas and neurofibromas. These neoplasms, associated with von Recklinghausen's disease (VRH) may originate at any anatomic site, but usually arise in the cranial nerves and occasionally in the nerves of the extremities [2]. Tumors of the sacral plexus are exceedingly rare [3]. We report a case of a pelvic mass that turned out to be a retroperitoneal schwannoma (ancient schwannoma), not associated with VRH.

Case Report

A 28-year-old woman, gravida 1, para 1, presented at our clinic with lower pelvic pain, irregular menses and frequent urination. The medical history was insignificant with no past or family history of VRH. The woman denied any changes in bowel habits. Gynecological examination revealed an immobile, solid, large mass occupying the pelvis and expanding to the right side. The rest of the clinical and laboratory examination was normal. Serum levels of cancer indexes were also within normal range. Ultrasound sonography disclosed a mass approximately 15 x 15 cm with cystic and solid elements (Figure 1). Pelvic radiography showed hydronephrosis of the right kidney and displacement of the right ureter. Computed tomography (CT) confirmed the existence of a pelvic tumor with a size of 14 x 14 cm, suggesting that it was probably an ovarian mass of embryonic origin (Figure 2).

After insertion of ureteral stents, the patient underwent an exploratory laparotomy that revealed normal uterus and adnexes and a retroperitoneal pelvic mass. Abdominal washing showed benign cytology. The histological report of the excised tumor (Figure 3) described lesions that correspond to a degenerated schwannoma (ancient schwannoma) with spindle cells ordered in bundles and rare mitoses. Immunohistochemically the tumor cells were strongly positive to vimentine (DACO 1:40) and S-100 protein (DACO 1:1000) (Figure 4). The postoperative course was uneventful without neurologic or other deficits observed.

Discussion

Neurilemomas are usually found incidentally and can occur with equal frequency in males and females, mostly between ages 20-50 [2]. Peripheral nerve sheath tumors arising in the pelvic retroperitoneum present infrequently as gynecologic masses. In particular, retroperitoneal schwannomas originating from the sacral nerves, like in our case, are exceedingly rare [4]. Commonly, these masses remain asymptomatic attaining a considerable size before being discovered. Even with large tumors they still remain a diagnostic dilemma because of the non-specific symptoms and findings.

Diagnosis can be established only after histological examination. Imaging methods can sometimes provide additional information. CT findings usually present masses with a heterogeneous pattern and areas of cystic degeneration and calcification. CT-guided biopsy may be helpful only if the sample contains enough Schwann cells to visualize microscopically, but is still unreliable misinterpretating degenerative cells as malignant [5]. Ogose *et al.* [6], reported a case where needle insertion caused pain along the sciatic nerve and supported that CT-guided needle biopsy is worthwhile for making preoperative diagnoses though there is still risk of hemorrhage or

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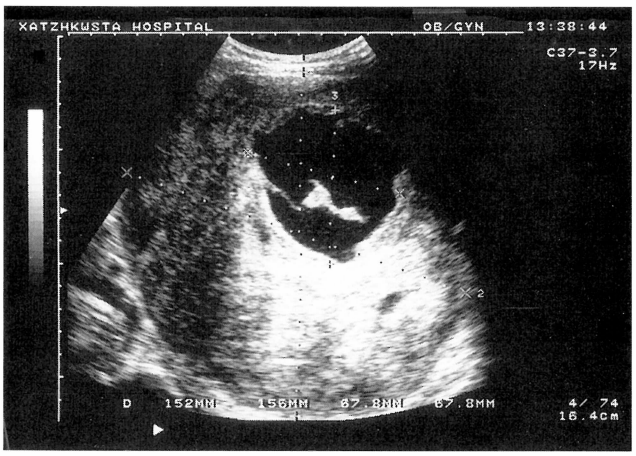
Fig. 1



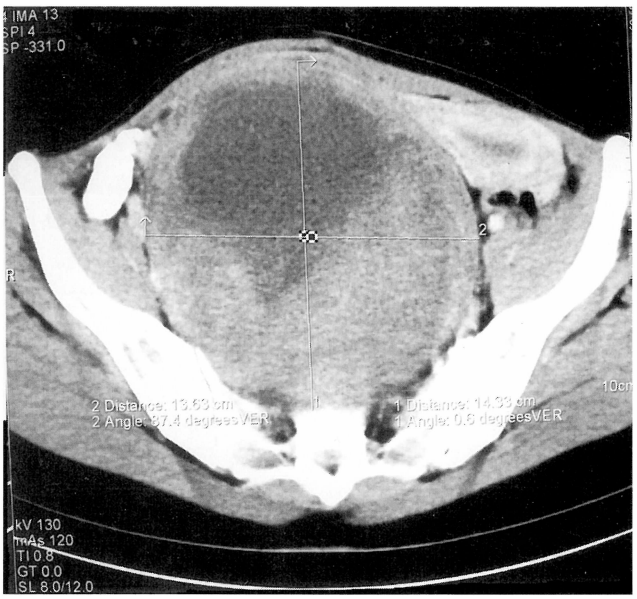
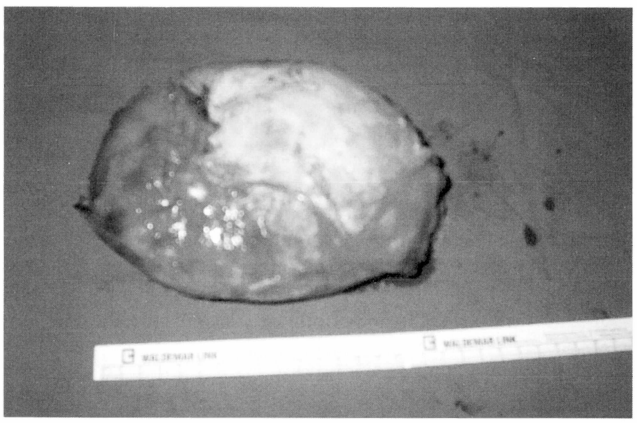


Fig. 3



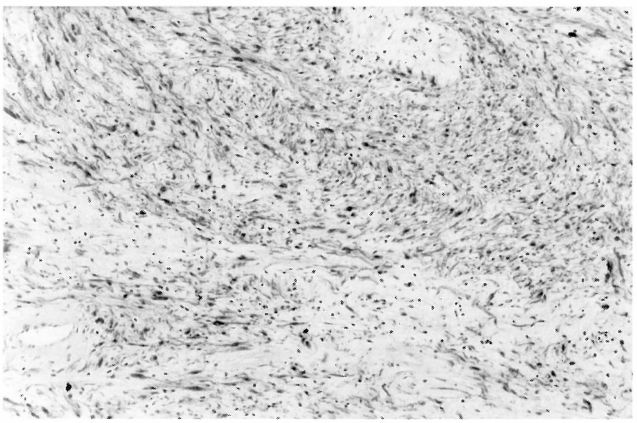


Figure 1. — Ultrasound examination showing the pelvic mass with cystic and solid elements.
Figure 2. — Computed tomography showing the relation of the mass with other tissues.
Figure 3. — Macroscopic appearance.
Figure 4. — Microscopic appearance (S-100 protein, x 200).

infection. Ueda *et al.*, mentioned that a cystic appearance is present in approximately 66% of the cases, helping in the differential diagnosis from other types of retroperitoneal tumors. [7]. However, neurilemomas with totally cystic form have also been reported [6].

These tumors are usually encapsulated and often appear with degenerative changes such as cyst formation, calcification and hyalinization. In particular, if they show degenerative changes and degenerative nuclear atypia in histological examination, they are called ancient schwannomas.

Retroperitoneal schwannomas are rare tumors difficult to diagnose [4, 6]. The initial thinking was that this tumor was an ovarian mass while clinical symptoms had no diagnostic value. There was no radiating pain along the sciatic nerve and the report of irregular menses was confusing. The same preoperative diagnosis has also been reported [4, 8]. Appropriate management depends on the age of the patient, the symptoms and the tumor's location. White *et al.* [9] in a study of 57 patients reported incomplete resection in about 10% of the cases. Dominquez *et al.* [10] mentioned a recurrence rate of 16% after a conservative approach, while Abernathy [1], recommended aggressive resection due to high recurrence rate. Persistence also seems to be associated with the involvement of pelvic bones, vessels, and major pelvic nerves. However, the aim of surgical therapy is to excise the tumor avoiding nerve injury. If extensive dissection is necessary clear understanding of the anatomy and cooperation between surgeons is essential.

Up to date, 20 cases of schwannomas of the pelvis have been reported [11]. Despite their rare occurrence they should be included in the differential diagnosis of gynecologic masses. Our patient has been followed for eight months after surgery and presents asymptomatic, with no motor or sensory disturbances and no evidence of recurrent tumor.

References

[1] Abernathy C., Onofrio B., Scheithauer B., Pairolero P.C., Shivis T.C.: "Surgical management of giant sacral schwannomas". *J. Neurosurg.*, 1986, *65,* 286.

[2] Hunter V.P., Burke T.W., Crooks L.: "Retroperitoneal nerve sheath tumors: An unusual case of back pain". *Obstet. Gynecol.,* 1988, *71,* 1050.

[3] Whitaker W.G., Droulias C.: "Benign encapsulated neurilemoma: a report of 76 cases". *Am. Surg.,* 1976, *42,* 675.

[4] Rajab K.E., Issa A.A., Skerman J.: "Unusual presentation of an abdominoinguinal schwannoma". *Obstet. Gynecol.,* 2003, *102,* 1158.

[5] Daneshmand S., Youssefzadeh D., Chamie K., Boswell W., Wu N., Stein J.P., et al.: "Benign retroperitoneal schwannoma: a case series and review of the literature". *Urology,* 2003, *62,* 993.

[6] Ogose A., Hotta T., Sato S., Takano R., Higuchi T.: "Presacral schwannoma with purely cystic form". *Spine,* 2001, *26,* 1817.

[7] Ueda M., Okamoto Y., Ueki M.: "A pelvic retroperitoneal schwannoma arising int the right paracolpium". *Gynecol. Oncol.*, 1996, *60,* 480.

[8] Kentopp K., Jones M.A., DeCain M., Tarraza H.M.: "Pelvic retroperitoneal schwannoma mimicking an ovarian neoplasm: report of a case and review of the literature". *Eur. J. Gynecol. Oncol.,* 1998, *19,* 57.

[9] White W., Shiu M.H., Rosenblum M.K., Erlandson R.A., Woodruff J.M.: "Cellular schwannoma. A clinicopathologic study of 57 patients and 58 tumors". *Cancer,* 1990, *66,* 1266.

[10] Dominguez J., Lobato R.D., Ramos A., Rivas J.J., Gomez P.A., Castro S.: "Giant intrasacral schwannomas: report of six cases". *Acta. Neurochir.,* 1997, *139,* 954.

[11] Duran B., Guvenal T., Yildiz E., Cetin M., Erden O., Demirkoprulu N.: "An unusual cause of adnexal mass: fallopian tube schwannoma". *Gynecol. Oncol.,* 2004, *92,* 343.

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