CASE REPORT

Benign Presacral Schwannoma Mimicking Pedunculated Leiomyoma

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Abstract

Schwannomas, also known as neurilemmomas, are rare tumors that arise from the peripheral nerve sheath. We present a young woman with a presacral schwannoma mimicking a pedunculated leiomyoma and producing sciatic pain. Gynecological examination revealed a hard, palpable, tender mass in the right adnexal region. Transvaginal ultrasound revealed a homogenous tumor mass measuring 4x5 cm next to the right ovary but not attached to it. Color Doppler ultrasound showed the vascularity of the tumor to match closely the vascularity of the uterine muscle. The suspected diagnosis was subserous uterine leiomyoma. The patient underwent surgery 4 weeks later. An encapsulated yellowish-white tumor was tightly attached to the sacrum, and to prevent rupture of the tumor, the laparoscopic procedure was converted into laparotomy. The histopathological diagnosis was a benign soft tissue tumor, a schwannoma. The patient's recovery was uneventful. She has now been asymptomatic for 9 months, and her lower back pain has disappeared. The present case demonstrates the difficulty to discriminate between a presacral schwannoma and a pedunculated leiomyoma.

Keywords: Schwannoma, presacral tumor, retroperitoneal tumor

Özet

Pedinküler Leiomiyoma Benzeyen Benign Presakral Schwannoma

Nörilemmoma olarak da bilinen Schwannomalar, periferal sinir kılıfından ortaya çıkan nadir tümörlerdir. Burada pedinküler leiomiyoma benzeyen ve siyatik ağrılarına yol açan genç bir kadındaki iyi huylu presakral Schwannoma sunulmaktadır. Jinekolojik muayenede sağ adneksiyal bölgede sert, hissedilir bir hassas kitle saptanmıştır. Transvajinal ultrasonografik muayenede sol overe yakın, ancak yapışık olmayan 4x5 cm'lik homojen bir tümöral doku görülmüştür. Renkli Doppler sonografide ise tümördeki vasküler yapının, uterin vasküler yapıyla belirgin derecede benzeştiği izlenmiştir. Subseröz uterin leiomiyom ön tanısı ile hasta 4 hafta sonra ameliyat edildi. Kapsüllü, sarımsı-beyaz bir tümörün sakruma sıkıca yapışık olduğu izlendiğinden tümörün rüptürünü engellemek amacıyla laparoskopik girişim laparotomiye çevrilmiştir. Histopatolojik tanı, iyi huylu bir yumuşak doku tümörü olan Schwannoma idi. Hastanın postop seyri olağan idi. Son 9 aydır asemptomatik olan hastanın alt sırt ağrıları da ortadan kalktı. Bu vaka sunumunda, presakral Schwannoma ile pedinküler leiomiyomu ayırt etmedeki güçlükler tartışılmaktadır.

Anahtar sözcükler: Schwannoma, presakral tümör, retroperitoneal tümör

Introduction

Schwannomas (neurilemmomas) are soft-tissue tumors arising from the peripheral nerve sheath, more specifically the Schwann cells. These tumors have a predilection for the head and neck, flexor surfaces of upper and lower extremities and posterior mediastinum (1). Only 0.3-3.2% of benign schwannomas are found in retroperitoneal locations, constituting approximately 4% of retroperitoneal tumors (2-5). They, most frequently occur in patients aged 20 to 50 years, more frequently in men than in women (6).

Schwannomas are almost invariably slow growing, non-aggressive neoplasms that are solitary in the vast majority of cases. In 5 to 18% of cases they are associated with neurofibromatosis Type I (7), in which case lesions may be multiple and often plexiform. Malignant transformation is exceedingly rare. However, malignant tumors have been reported (8).

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We present a young woman with a presacral schwannoma mimicking a pedunculated leiomyoma and producing sciatic pain.

Case

A 22-year-old woman, gravida 0, para 0, presented at the student health center for painful menstruation. She had been experiencing some degree of pelvic discomfort for two years. The pain was referred to the lower back and the right leg. Her menarche had been at the age of 12, and she had a regular menstrual cycle. Condom was used for contraception. The patient had no stigmas of neurofibromatosis. Gynecological examination revealed a hard, palpable, tender mass in the right adnexal region. Transvaginal ultrasound showed a homogenous mass measuring 4x5 cm that seemed to be attached to the right ovary.

The patient was referred to our hospital for further examinations. Transvaginal ultrasound revealed a normal uterus and adnexal structures. There was a tumor mass next to the right ovary but not attached to it (Figure 1). Color Doppler ultrasound showed the vascularity of the tumor, to match closely the vascularity of the uterine muscle. The suspected diagnosis was subserous uterine leiomyoma. To confirm the diagnosis and to extirpate the tumor, the patient underwent laparoscopic surgery 4 weeks later.

Laparoscopy showed a normal uterus. Both ovaries were normal in size. There were small spots of endometriosis on the ovarian surfaces. A small endometriotic spot was also seen behind the uterus. The fallopian tubes were normal. On the right side of the pelvis there was a solid retroperitoneal tumor that was located between the midline and the large pelvic vessels, partly under the right ureter. The right ureter was not substantially dilated. A peritoneal incision revealed an encapsulated yellowish-white tumor. The tumor was tightly attached to the sacrum, and to prevent rupture of the tumor, the laparoscopic procedure was converted into laparotomy. Open biopsy was performed, and the histopathological diagnosis of the frozen section was a benign soft tissue tumor, most likely a schwannoma. The tumor was completely removed. There was some bleeding from the presacral veins, but complete hemostasis was achieved. A total blood loss of 800 ml was estimated.

Macroscopically, the tumor was encapsulated, homogenous and firm measuring 5.5x4x3 cm (Figure 2). Histological examination of the tumor showed that it was composed of intersecting fascicles of spindle-shaped cells (Figure 3), which occasionally were arranged in loose palisade formations. As a minor component some hypocellular areas were present in the tumor and occasionally there were blood vessels showing hyaline thickening of their walls. The tumor cells showed no nuclear atypia. Mitotic counts ranged from 0 to 1 mitotic figures per 10 high-power fields and there was no tumor necrosis. Immunohistochemical analysis demonstrated a strong immunoreaction for S-100 and vimentin, whereas desmin, caldesmon, α-SMA and CD117 were negative in the tumor cells. These findings supported the diagnosis of a benign schwannoma.

The patient's recovery was uneventful. She has now been asymptomatic for 9 months, and her lower back pain has disappeared.

Discussion

Presacral schwannomas are situated deep in the pelvic floor. The retroperitoneum is an expandable space, and the tumor may grow quite large before the patient becomes symptomatic. The symptoms may arise when the tumor compresses a retroperitoneal organ or surrounds and compresses the nerve itself. The symptoms are more or less vague and non-specific, not always neurological, and may include voiding problems, renal colic, slow urinary stream and hesitancy, constipation, lumbar or back pain radiating down the leg, inguinal swelling, and hydronephrosis (9).

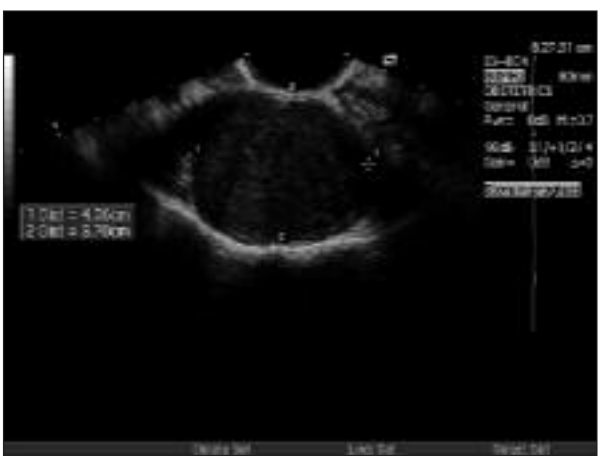

Figure 1. Ultrasound image of the pelvic tumor mimicking pedunculated leiomyoma.

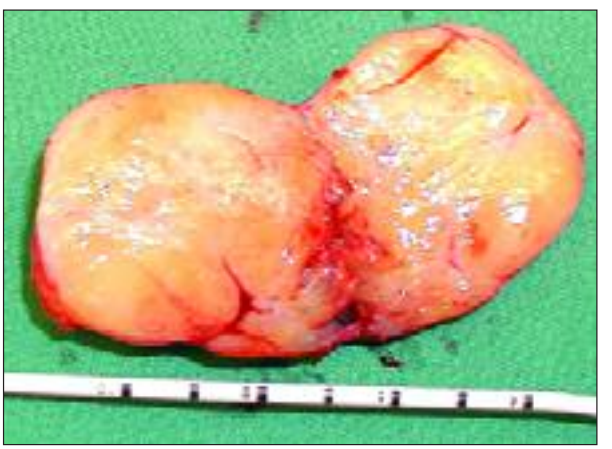

Figure 2. Cross-section of the presacral schwannoma, 5.5x4x3 cm in size.

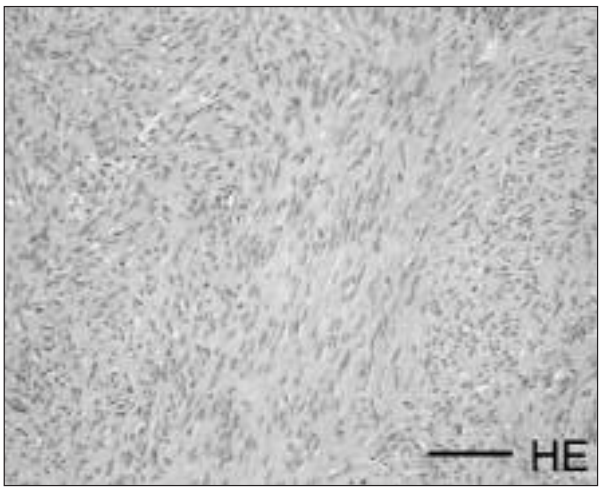

Figure 3. Histological section of the tumor stained with hematoxylin and eosin. Benign-apperaring spindle-shaped cells arranged in interwoven fascicles. Bar 0.01 mm. Original magnification x100.

Presacral schwannomas are extremely difficult or even impossible to diagnose preoperatively. They are often asymptomatic and found incidentally. Our patient had been suffering from pelvic discomfort for two years. These symptoms suggest pelvic endometriosis, a common chronic gynecologic disorder associated with pelvic pain. The pain was also referred to the lower back and to the right leg, suggesting that the presacral tumor was bearing down on the sacral nerves and the right sciatic nerve. Pelvic schwannoma is a rare cause of sciatic and low back pain (10).

Ultrasonography, computer tomography (CT), and magnetic resonance imaging (MRI) help to approximate the size and location of the tumor. CT and MRI also give information about the invasion of the presacral tumor and the organs involved. Especially large schwannomas including degenerative changes, such as a cyst, calcification, hyalinization, or hemorrhage, may be misleading upon imaging (9). In our case, transvaginal ultrasonography failed to differentiate presacral schwannoma from pedunculated or parasitic leiomyoma. CT-guided fine needle aspiration biopsy was not performed because it does not appear to provide an accurate preoperative diagnosis (9). Moreover, we preferred surgery to fine needle biopsy because the possibility of a malignant pelvic tumor had not been excluded.

Histologically, the tumor was well encapsulated and composed of benign appearing spindle shaped cells arranged in tight fascicles showing nuclear palisading. These cellular areas are called Antoni A areas in schwannomas (11). There was also some loosely textured hypocellular Antoni B areas and hyalinized blood vessels typical of schwannoma. Mitotic figures were very rare. Verocay bodies, which are often seen in schwannomas, were not present in this tumor. These are parallel arrays of tumor cell nuclei, separated by eosinophilic dense packed tumor cell processes, and basement membranes. The main differential diagnostic considerations (12) are leiomyoma, neurofibroma and gastrointestinal stromal tumor (GIST). Leiomyoma can histologically mimic schwannoma because of its composed spindle cell fascicles and nuclear palisading. However, this tumor showed no immunoreaction with the antibodies against smooth muscle and was strongly positive with the antibody S-100. Neurofibroma is also immunoreactive with S-100, but it lacks encapsulation, palisading of nuclei and blood vessels with hyaline thickening. The metastasis of gastrointestinal stromal tumor can also be present as a spindle cell tumor with nuclear palisading and low mitotic rate, but it is immunoreactive with CD117. We found that the histological picture and immunohistochemical findings are consistent with the diagnosis of schwannoma.

Surgery is the treatment of choice. A laparoscopic approach offers the advantage of thorough evaluation of the abdomen. Successful laparoscopic resection of a retroperitoneal schwannoma has been reported (13). Conversion from laparoscopy into laparotomy was obligatory in our case because the tumor was firmly attached to the sacrum. Surgery may be difficult and includes some risks. The adjacent structures, i.e. the ureter or bladder, may sustain injury. Neurological impairment is possible because the schwannoma arises from the supporting cells of a nerve. Presacral schwannoma is often attached to the presacral venous plexus, and complete resection may cause profuse bleeding (9). Yano et al. (2003) suggested that large retroperitoneal schwannomas should be removed by leaving the capsule intact, to prevent injury to the adjacent structures (14). A literature survey shows, however, that patients undergoing complete surgical resection do well without evidence of early recurrence (9). Our understanding is that the surgical approach should focus on complete excision of the encapsulated tumor and be performed by an experienced gynecological surgeon.

In conclusion, unspecific pelvic pain referring to the lower back or leg may be the presenting feature in presacral schwannoma. Transvaginal ultrasonography is widely used in gynecological practice. However, it is incapable of reliably distinguishing between presacral schwannoma and pedunculated leiomyoma.

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