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MORPHOLOGICAL STUDY

The anterior extraperitoneal approach to the rare presacral/ /retroperitoneal schwannoma

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Abstract: Presacral/retroperitoneal schwannomas are extremely unusual and their surgical approach is challenging. Various surgical approaches have been proposed. Here we describe our experience with the anterior extraperitoneal approach for the Type 3 presacral/retroperitoneal schwannoma. A 33-year-old woman presented with a history of infertility and a presacral/retroperitoneal mass. The patient underwent abdominal/pelvic CT and MRI that demonstrated presence of a solid, well circumscribed Type 3 presacral/retroperitoneal schwannoma. The anterior extraperitoneal approach was successfully used to remove the presacral/retroperitoneal schwannoma. The histopathological evaluation confirmed the preoperative diagnosis. The anterior extraperitoneal approach is helpful as an alternative approach in the surgical management of benign Type 3 presacral/retroperitoneal schwannomas *(Fig. 4, Ref. 15)*. Full Text in free PDF *www.bmj.sk.*
Key words: extraperitoneal approach, presacral/retroperitoneal, schwannoma.

Schwannomas are benign nerve sheath tumors that arise from the Schwann cells of the neural sheath. They may occur nearly anywhere in the body and are the cause of 6% of primary retroperitoneal neoplasms (1, 2). About 60 cases of retroperitoneal schwannomas have been reported, with fewer than 20 of them situated in the pelvis (3). Presacral/retroperitoneal schwannomas are unusual; they can be large and difficult to diagnose (4).

Based on their location with respect to the sacrum, Klimo et al (5) suggested that these tumors be classified into one of three types: Type I tumors are confined to the sacrum; Type II tumors originate within the sacrum but erode into either the anterior or posterior sacral wall and spread to adjacent spaces; and Type III tumors are located primarily in the presacral/retroperitoneal area (5). Removal of Type 3 presacral/retroperitoneal schwannomas usually requires a team approach involving neurosurgeons and general surgeons (5).

A total excision via the open abdominal or sacral approache has been traditionally advocated as the best treatment for presacral/retroperitoneal tumors. The anterior extraperitoneal approach is an attractive and effective alternative approach for the surgical management of benign Type 3 presacral/retroperitoneal schwannomas (3, 6, 7).

We present a case of Type 3 presacral/retroperitoneal schwannoma which was treated successfully with an anterior extraperitoneal approach without persistent neurological deficit.

Case

A 33-year old woman presented to the obstetrics-gynecology clinic for infertility. She had no abdominal pain, pelvic pain, urinary complaints, intestinal complaints, or lower extremity numbness or weakness. During her infertility work-up, a well-circumscribed, homogenous 4-cm mass adjacent to but separate from the left ovary was found on transvaginal ultrasound. Subsequent CT scan revealed a well-defined, 4 cm diameter solid mass emanating from the left S-2 anterior sacral foramina (Fig. 1). Sacral foraminal widening and concomitant erosion were not detected. Pelvic MR images revealed a Type 3 presacral/retroperitoneal schwannoma, which completely enhanced with contrast and originated from the S-2 nerve root (Fig. 1). The patient was referred to the neurosurgery department for further evaluation. The patient had no neurological, bladder, or bowel symptoms. On physical examination, her abdomen was soft and not tender with no palpable masses. No *café-au-lait* spots or dermal neurofibromas were present. Although appearance of the mass on imaging studies was consistent with a benign schwannoma, resection was discussed with the patient in light of her infertility and likely continued enlargement of the tumor over time. An extraperitoneal approach was preferred due to its decreased chance of interfering with intestinal and gynecologic functions. The patient decided to proceed with excision of the tumor.

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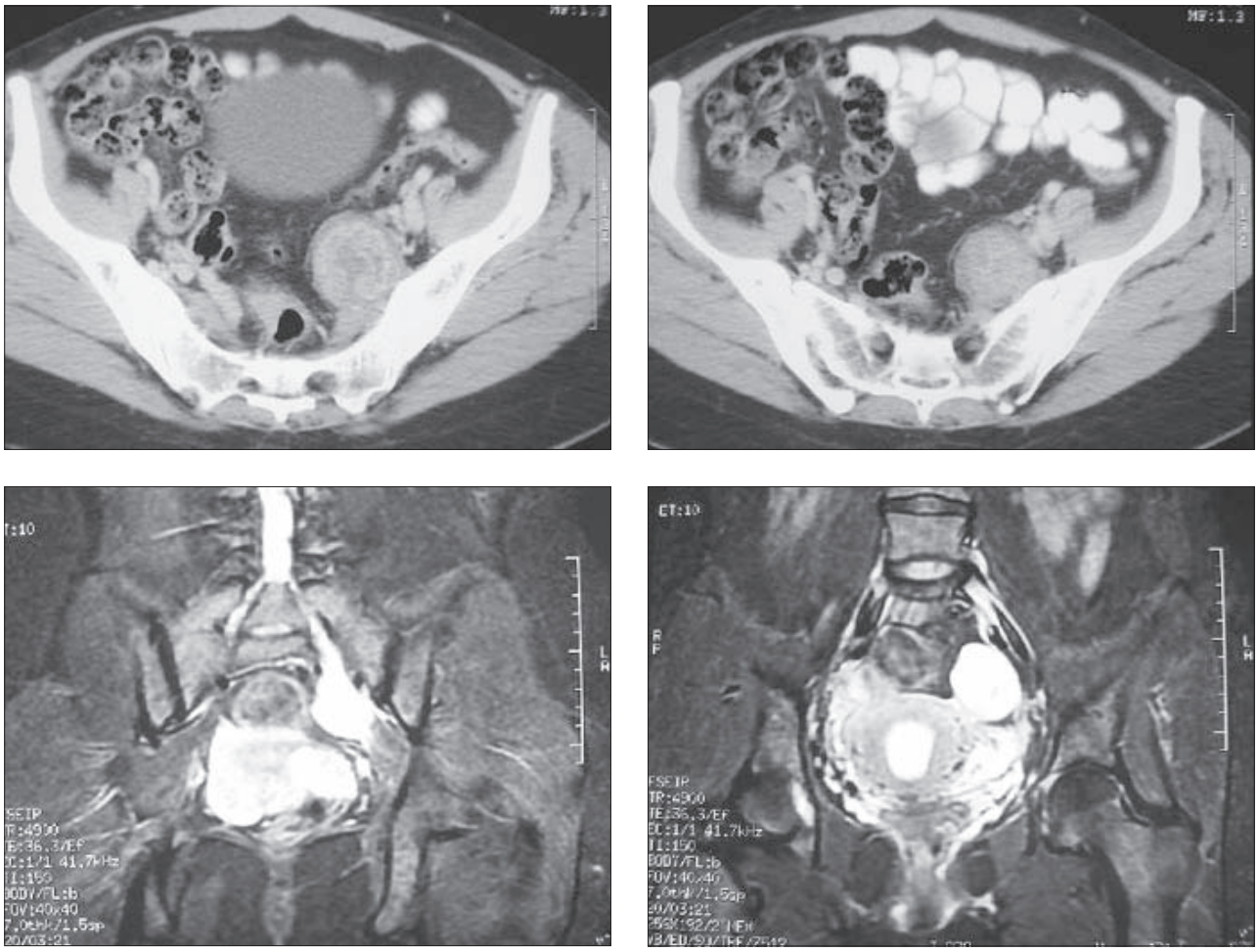

Fig. 1. Presacral benign schwannoma of the pelvis in a 33-year-old woman (a) CT scan following contrast injection demonstrates central enhancement in the well-defined solid tumor in the left pre-sacral pelvic area. (b) Fat-saturated T2-weighted coronal MRI image shows the hyperintense tumor and its continuity with the neural fibers.

Under the general endotracheal anesthesia, the patient was placed in supine position with an adjustable block underneath the lumbo–sacral junction. A left paramedian skin incision was made from the umbilicus to the symphysis. The superficial sheet of the rectus abdominis muscle was vertically cut and the muscle was moved to the right. The abdominal muscles were separated from the retroperitoneal fat to the medial side of the psoas muscle. The peritoneum was dissected from the left abdominal wall. The left iliac artery and vein were moved to the left (Fig. 2). The ureter was identified and protected during the dissection (Fig. 2). The tumor was then directly exposed on the left sacral plexus (Fig. 3). With this approach, an adequate exposure deep in the presacral space was obtained. The superior gluteal vessels were laid on the superior aspect of the tumor. The encapsulated tumor was dissected from the associated left sacral plexus, preserving neural continuity (Fig. 3). There were no intraoperative complications or need for blood transfusion.

In the immediate postoperative period, no neurological, bladder, or bowel symptoms were observed. The patient was discharged to home on the fourth postoperative day and recovered without complications.

The tumor specimen measured 4x4x4 cm in size (Fig. 3). The histopathologic examination revealed a benign schwannoma, with no evidence of malignancy. Mitotic figures were absent (Fig. 4). Immunohistochemically, the tumor cells were strongly and diffusely positive for S-100 protein. The schwannoma cells exhibited a mild pleomorphism, with varying degrees of cellularity. Both Antoni A ("dense") and B patterns ("loose") were present (Fig. 4). The hallmark histologic feature of a schwannoma, Verocay bodies, were also seen (Fig. 4).

Discussion

Presacral/retroperitoneal schwannomas may arise from the retroperitoneal space or by local extension from the sacrum (8, 9, 10, 11). The low incidence of this tumor and the lack of specific signs and symptoms make preoperative diagnosis very difficult. They are often asymptomatic and found incidentally by

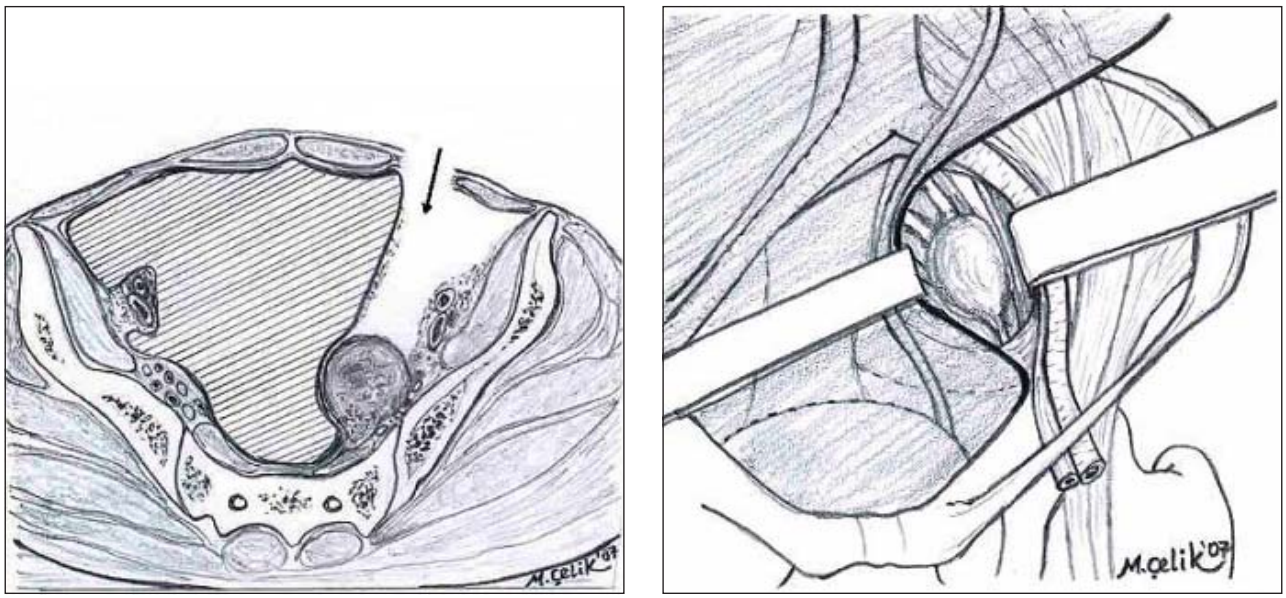

Fig. 2. (Left) Retroperitoneal area during intraoperative gross dissection. (Right) The photo of the excised tumor (4 x 4 x 4 cm) after bisection. The solid tumor was well-encapsulated without evidence of invasion of surrounding structures. The tumor was of white-yellow color with homogeneously gray and gelatinous cut surfaces.

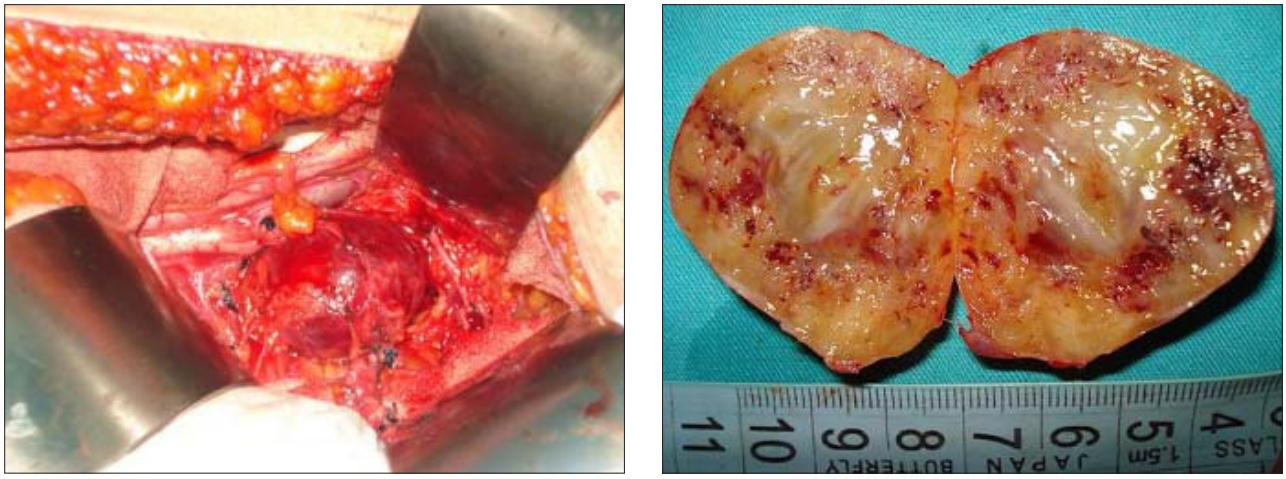

Fig. 3. (Left) Cross-sectional schematic of the anterior retroperitoneal approach of the pelvis. The avascular path is between the peritoneal cavity and the psoas muscle.
(Right) Schematic of the retraction of the retroperitoneal vascular and urinary elements for gaining access to the presacral schwannoma.

obstetricians and other clinicians, as was the case in our patient (3, 6, 7, 12). Delayed presentation is common because of their solitary nature, slow growth, permissive surrounding anatomical environment, and the lack of specific symptoms. They can often reach massive size before producing neurological symptoms (1, 5). The schwannoma in our patient was caught in an early, asymptomatic stage through the incidental exam of a gynecologist.

The preferred surgical approach (anterior vs. posterior) for the resection of presacral/retroperitoneal schwannomas varies according to tumor location (5, 8, 9, 10, 11, 12). Type I tumors may be resected via a posterior approach alone, Type III may require an anterior approach, and Type II tumors usually require a combined anterior–posterior surgical approach (5).

In general, a median laparotomy with a transabdominal/transperitoneal dissection can be performed to reach the Type 3 presacral/retroperitoneal schwannomas. However, operative blood loss can be profuse due to adhesion of the tumor capsule to the presacral venous plexus (10, 12, 13). Establishing a lasting haemostasis may be very difficult (14).

The anterior extraperitoneal approach for Type 3 presacral/retroperitoneal schwannomas has been previously described in one patient by Rousseau et al (15). Our procedure varied from Rousseau's in that we retracted the iliac artery and vein laterally in our patient.

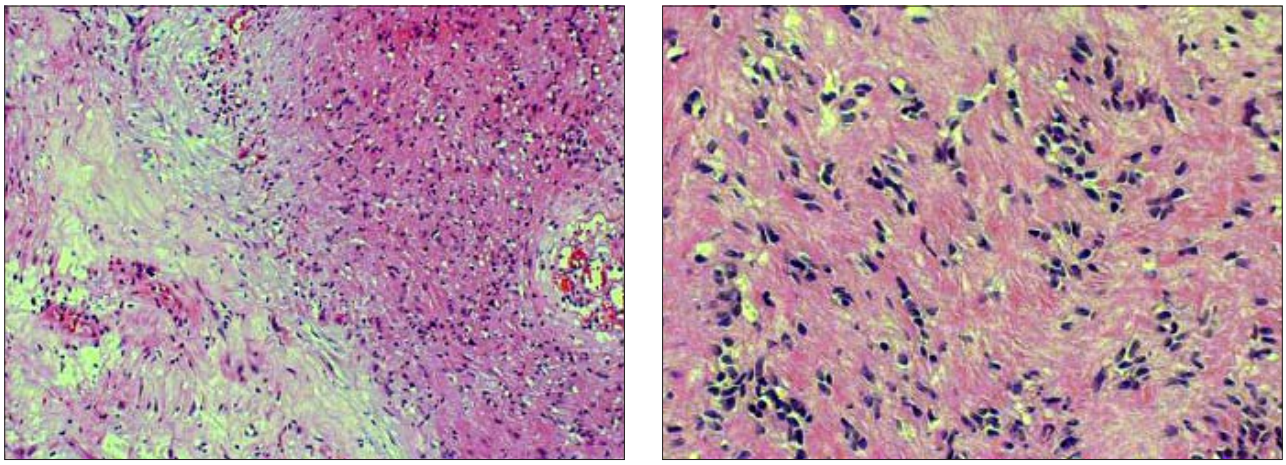

Fig. 4. (Left) The characteristic histological features of schwannoma include the presence of alternating Antoni A and Antoni B areas (HE x10). (Right) Classic peripheral schwannoma with wavy nuclei and Verocay bodies (HE x20).

Usually, the schwannoma is able to be dissected cleanly from its associated nerve, and the total resection is curative. Once totally excised, recurrence of benign schwannomas is not expected. Adjuvant therapy is therefore not needed (1). In our patient as well, neural continuity was preserved after the total resection was performed.

Our preference is for a general surgical and neurosurgical team to work together through the anterior extraperitoneal approach for the resection of Type 3 presacral/retroperitoneal schwannomas. This approach provides ample exposure for hemostasis and total excision without residual neurological deficit. Because the peritoneal cavity is not violated, few complications are expected during the postoperative course.

Conclusion

The anterior extraperitoneal approach is an effective option in approaching Type 3 presacral/retroperitoneal schwannomas. Use of the paramedian incision in the extraperitoneal approach to structures in the pelvis is recommended for its ease and excellent exposure to the area for haemostasis and tumor resection.

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