CASE REPORT

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Multidisciplinary Surgical Management of a Giant Presacral Schwannoma Involving the Internal Iliac Artery: A Case Report

Candost Hanedan[1], Neslihan Öztürk[1], Firdevs Şahin Duran[2], Berna Turhan[3], İnan Uzunoğlu[4]

[1]Clinic of Gynecologic Oncology, University of Health Sciences Türkiye, Ankara Etlik City Hospital, Ankara, Türkiye
[2]Clinic of Pathology, University of Health Sciences Türkiye, Ankara Etlik City Hospital, Ankara, Türkiye
[3]Clinic of Radiology, University of Health Sciences Türkiye, Ankara Etlik City Hospital, Ankara, Türkiye
[4]Clinic of Neurosurgery, University of Health Sciences Türkiye, Ankara Etlik City Hospital, Ankara, Türkiye

Abstract

Presacral (retrorectal) schwannomas are rare, benign tumors originating from the Schwann cells of the sacral nerve roots. Their deep pelvic location and proximity to major neurovascular structures make surgical management challenging. We report a case of a 57-year-old postmenopausal woman who presented with postmenopausal bleeding, an incidental finding that led to the discovery of the pelvic mass, and who was found to have a giant presacral mass involving the left internal iliac artery. Magnetic resonance imaging revealed a 12×8×12 cm lesion, consistent with a Klimo type III schwannoma arising from the presacral region. The tumor was completely excised through an anterior multidisciplinary surgical approach with intraoperative neuromonitoring, followed by total hysterectomy and bilateral salpingo-oophorectomy. The postoperative course was uneventful, and the patient remained disease-free at 39 months of follow-up. This case highlights the importance of meticulous preoperative imaging, multidisciplinary surgical planning, and intraoperative neuromonitoring for the safe management of large presacral schwannomas with major vascular involvement.

Keywords: Presacral schwannoma, klimo classification, pelvic tumor, internal iliac artery, multidisciplinary surgery

INTRODUCTION

Schwannomas, also referred to as neurilemmomas, are benign tumors originating from well-differentiated Schwann cells of the peripheral nerve sheath. They represent the most common benign tumors of the peripheral nervous system and typically exhibit slow growth and an indolent clinical course. Because of their gradual enlargement, many patients remain asymptomatic for long periods, and tumors are often detected incidentally during imaging performed for unrelated reasons. Malignant transformation is extremely rare and is most commonly associated with neurofibromatosis type 1 or prior radiation exposure.[1,2]

Schwannomas arising from the sacral nerve roots and extending into the presacral or retrorectal space are particularly rare.[3] Their deep pelvic location and close anatomical relationship with major neurovascular structures and pelvic organs often complicate both diagnosis and surgical management. The Klimo classification system categorizes sacral schwannomas into three types based on their anatomical location: type I (confined within the sacrum), type II (extending beyond the sacral bone margins), and type III (located entirely in the presacral space).[4] Because type III schwannomas are located within the presacral space, they are often in close proximity to major pelvic vessels, thereby increasing the risk of vascular involvement and significant intraoperative bleeding.

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ORCID IDs of the authors: C.H. 0000-0003-3435-8943; N.Ö. 0000-0002-5578-8718; F.Ş.D. 0000-0001-9244-9590; B.T. 0000-0003-2522-5719; İ.U. 0000-0001-5859-0443.

Corresponding author: Candost Hanedan
E-mail: cnhanedan@gmail.com
ORCID ID: orcid.org/0000-0003-3435-8943

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Preoperative imaging, particularly magnetic resonance imaging (MRI), plays a crucial role in defining tumor size, anatomical relationships, and potential involvement of surrounding structures, thereby facilitating appropriate surgical planning.[5,6]

We report a rare case of a giant presacral schwannoma involving the internal iliac artery, which was successfully managed through a multidisciplinary surgical approach at our tertiary referral center. This case highlights the importance of detailed preoperative imaging and coordinated surgical planning in the safe management of giant presacral tumors.

CASE REPORT

A 57-year-old postmenopausal woman was referred to our tertiary referral center in November 2022 with postmenopausal bleeding and a pelvic mass detected on abdominal computed tomography. Pelvic examination revealed a heterogeneous mass with both solid and cystic components extending into the abdomen; the patient had no neurological symptoms such as radicular pain. Transvaginal ultrasonography demonstrated a myomatous uterus, normal-appearing ovaries, and a solid, hyperechoic mass with cystic components extending into the upper abdomen. Tumor markers and other laboratory findings were within normal limits.

Pelvic MRI, performed for further evaluation, demonstrated a 12×8×12 cm lesion arising from the presacral region (Figure 1). The patient's medical and family histories were unremarkable. Based on radiological findings, the lesion was classified as a Klimo type III presacral schwannoma. Preoperative cervical cytology and endometrial sampling (pipelle biopsy) were normal. Because the patient had a myomatous uterus associated with abnormal bleeding , a concomitant hysterectomy was planned. The case was discussed at a multidisciplinary team meeting involving neurosurgeons and radiologists, and it was decided to proceed with a combined surgical procedure via an anterior approach.

At exploratory laparotomy, a well-circumscribed presacral mass measuring approximately 12×12 cm and arising from the sacral region was identified, occupying a large portion of the pelvic cavity. A myomatous uterus and normal adnexal structures were also observed. Bilateral retroperitoneal dissection was performed, and both ureters were identified and suspended for protection. The retrorectal space was entered, dissection planes were developed, and the rectosigmoid colon was mobilized laterally. To minimize the risk of hemorrhage, major pelvic arterial and venous structures were carefully identified and controlled (Figure 2). Tumor dissection resulted in approximately 1200 mL of intraoperative blood loss, attributable to tumor invasion of the left internal iliac artery and diffuse bleeding from the presacral venous plexus. Blood loss was estimated by the anesthesia team based on the volume collected in the suction reservoir. Three units of packed red blood cells were transfused intraoperatively. Intraoperative neuromonitoring was performed at the neurosurgical team's request because of the tumor's close proximity to the sacral nerve roots. Neural signals from the first and second sacral nerve roots nerve roots were detected during dissection, allowing complete tumor excision without neurological complications.

Following tumor resection, total hysterectomy with bilateral salpingo-oophorectomy was performed. The postoperative course was uneventful, and the patient was discharged on postoperative day 4 without neurological deficits. Histopathological examination revealed spindle cell proliferation with nuclear palisading and verocay body formation, consistent with schwannoma. Immunohistochemical staining demonstrated diffuse, strong S100 positivity in tumor cells. No evidence of atypia or necrosis was identified (Figure 3). At the 39-month follow-up, the patient remained asymptomatic, and a follow-up MRI demonstrated no evidence of residual or recurrent disease. Written informed consent for publication of this case report and the accompanying images was obtained from the patient.

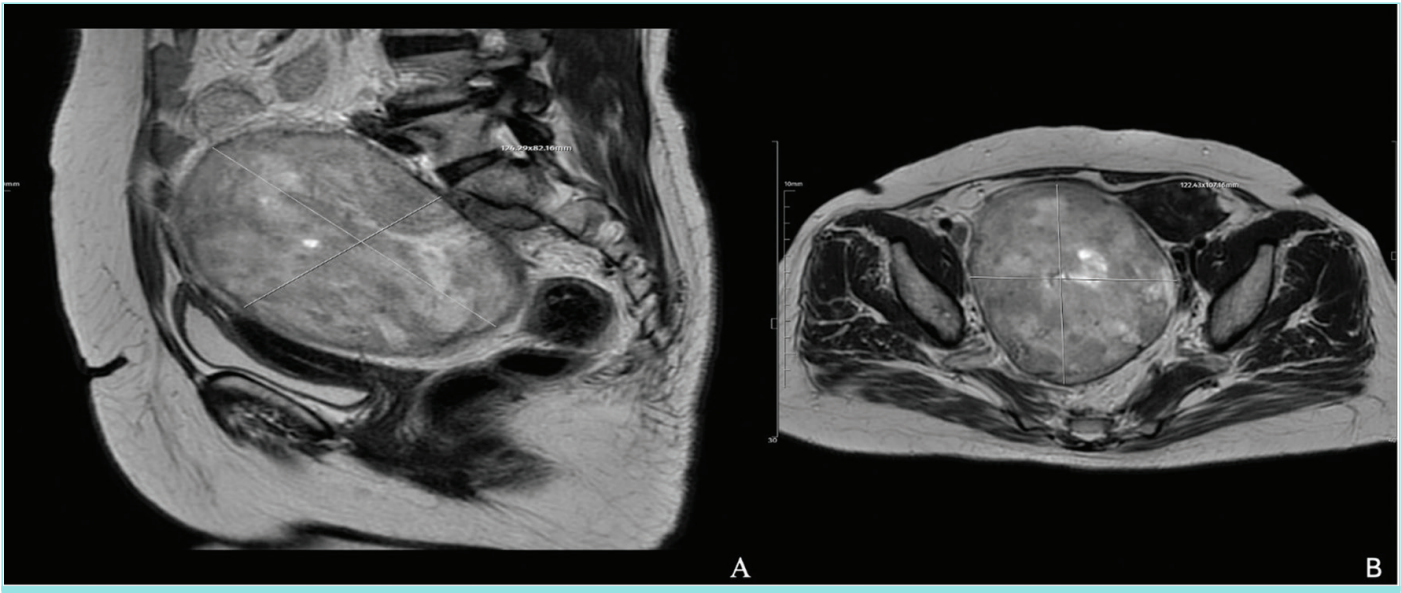

Figure 1. Pelvic magnetic resonance imaging. **A)** Sagittal view demonstrating a large presacral mass measuring approximately 128×82×122 mm. **B)** Axial view showing the lesion with a prominent solid component and cystic areas.

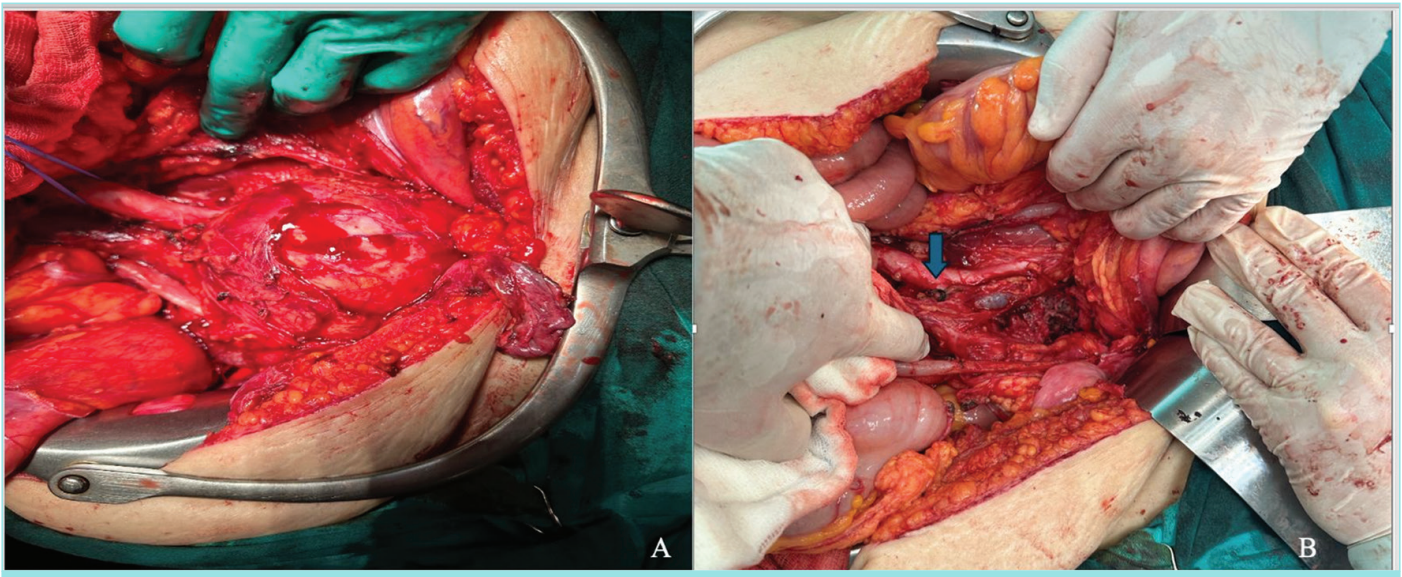

Figure 2. A) Intraoperative view of the giant presacral schwannoma. **B)** Postoperative view after tumor resection. The blue arrow indicates the ligated left internal iliac artery.

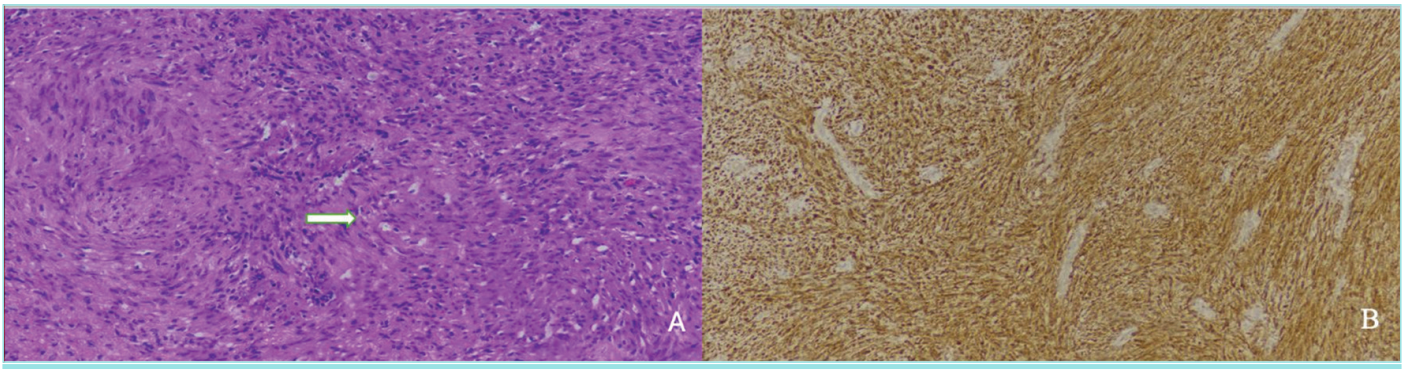

Figure 3. A) Histopathological examination showing spindle cell proliferation with nuclear palisading and Verocay bodies, consistent with schwannoma. **B)** Immunohistochemical staining demonstrated diffuse, strong S100 positivity.

DISCUSSION

Presacral schwannomas are rare benign tumors that arise from Schwann cells and represent a small proportion of all schwannomas. Their occurrence in the sacral or retrorectal region is particularly uncommon. Although schwannomas are typically benign, their deep pelvic location can make diagnosis and surgical management challenging because of their close relationship with major neurovascular structures and pelvic organs.[7] For this reason, careful preoperative imaging and multidisciplinary evaluation involving specialties such as neurosurgery and radiology are essential for optimal surgical planning. To our knowledge, a schwannoma with direct invasion of a major pelvic artery is exceedingly rare in the literature. While vascular involvement has been sporadically reported in iliac vessel schwannomas,[8] complete surgical resection of a Klimo type III presacral schwannoma with internal iliac artery invasion, managed through a multidisciplinary approach with intraoperative neuromonitoring and a 39-month disease-free follow-up, has not been previously described. This represents the primary novelty of the present case.

Complete surgical excision is considered the treatment of choice for pelvic schwannomas.[9] However, in cases of large presacral tumors, achieving complete resection may be technically demanding because these tumors are often located near major vessels and neural structures. A multidisciplinary surgical approach is therefore crucial, involving gynecologic oncologists familiar with retroperitoneal anatomy, radiologists who can accurately evaluate the tumor's relationship with major vascular structures, and neurosurgeons experienced in the management of nerve sheath tumors. Subtotal resection may be necessary to reduce the risk of major bleeding or irreversible neurological damage.[10] Although minimally invasive laparoscopic approaches have been increasingly reported for presacral schwannomas,[11] open anterior laparotomy was preferred in our case due to the tumor's large size (12 cm) and major vascular involvement of the left internal iliac artery, which carried a significant risk of intraoperative hemorrhage. The anterior approach was selected over posterior or combined approaches, as it provided direct access to major pelvic vessels, enabling rapid vascular control, a critical consideration given the internal iliac artery involvement.[3,10] Pennington et al.[3,10] and Sarhan et al.[7] reported that

complete resection is achievable in the majority of Klimo Type III schwannomas; however, subtotal resection may be considered when complete excision poses unacceptable neurological risk. In our case, complete resection was achieved without neurological deficit and was supported by intraoperative neuromonitoring; this outcome is consistent with the best results reported in the literature. Several reports have noted that the close anatomical relationship between presacral schwannomas and major pelvic vessels can increase the risk of bleeding during surgical dissection.[11,12] In our case, approximately 1200 mL of intraoperative blood loss occurred because of tumor involvement of the left internal iliac artery, necessitating arterial ligation to achieve hemostasis. This finding underscores the importance of careful preoperative vascular assessment and multidisciplinary surgical planning. Ligation of the left internal iliac artery was well-tolerated, and no postoperative complications, such as gluteal ischemia or impaired pelvic organ perfusion, were observed. Follow-up MRI performed during the 39-month postoperative period demonstrated no ischemic changes in the pelvic region, consistent with the well-established safety profile of unilateral internal iliac artery ligation due to the rich collateral pelvic circulation.

Preoperative imaging is essential in the evaluation of presacral schwannomas. MRI provides important information about tumor size, anatomical location, and its relationship to surrounding structures. MRI also helps differentiate schwannomas from other presacral or retrorectal tumors. In our patient, MRI demonstrated a large presacral mass measuring 12×8×12 cm and allowed classification of the lesion as a Klimo type III schwannoma.[4] The imaging findings also suggested involvement of adjacent vascular structures. Because of the tumor's large size and deep presacral location, a multidisciplinary surgical approach involving both gynecologic oncology and neurosurgical teams was adopted. The tumor was approached through an anterior laparotomy, which provided adequate exposure of the pelvic cavity and surrounding structures. In large presacral tumors, dense adhesions and bleeding from the presacral venous plexus are frequently encountered.

Therefore, critical anatomical structures, including the ureters and major pelvic vessels, should be carefully identified and protected before tumor resection. Despite significant intraoperative bleeding in our case, coordinated management by an experienced multidisciplinary team enabled effective hemostasis and complete tumor removal. The involvement of multiple specialties facilitated safe dissection around the sacral nerve roots and major vascular structures, thereby minimizing surgical complications. Intraoperative neuromonitoring played an additional role in preserving neural integrity during tumor dissection. Real-time monitoring of neural signals allowed the surgical team to identify and protect sacral nerve roots, thereby reducing the risk of postoperative neurological deficits. Previous studies have similarly demonstrated that neuromonitoring can be useful in complex nerve sheath tumor surgeries to preserve neural function.[13,14]

Although schwannomas are predominantly benign tumors, malignant transformation into malignant peripheral nerve sheath tumors is extremely rare and occurs in less than 1% of sporadic cases.[15] Certain histopathological features, including increased mitotic activity, nuclear pleomorphism, necrosis, loss of encapsulation, and infiltrative growth patterns, may suggest malignant potential. Therefore, careful histopathological evaluation remains essential following tumor excision. No histopathological features suggesting malignancy were identified. Complete surgical excision remains the primary treatment for schwannomas. Adjuvant therapies are generally not required for benign lesions but may be considered in cases of malignant transformation, positive margins, or recurrent disease.[16] Long-term follow-up is recommended even after complete resection because recurrence may occur, albeit rarely.

CONCLUSION

This case highlights the importance of multidisciplinary surgical management in giant presacral schwannomas with major vascular involvement. To our knowledge, this is one of the few reported cases of a Klimo type III presacral schwannoma with direct internal iliac artery invasion that were managed via an open anterior approach with intraoperative neuromonitoring, demonstrating that complete resection is achievable even with major vascular involvement. Long-term follow-up remains essential.

MAIN POINTS

- Presacral schwannomas are rare benign tumors that may present as large pelvic masses and mimic other retrorectal lesions.
- Preoperative imaging is essential to evaluate tumor size, anatomical relationships, and potential vascular involvement.
- Multidisciplinary surgical management with intraoperative neuromonitoring can facilitate safe resection of large presacral schwannomas.
- Careful surgical planning is crucial in cases of major vascular involvement to minimize intraoperative complications.

ETHICS

Informed Consent: Written informed consent was obtained from the patient for publication of this case report and the accompanying images.

Footnotes

Authorship Contributions

Surgical and Medical Practices: C.H., İ.U., Concept: C.H., F.Ş.D., Design: C.H., N.Ö., B.T., Data Collection and/or Processing: C.H., N.Ö., İ.U., Analysis and/or Interpretation: C.H., B.T., Literature Search: C.H., F.Ş.D., Writing: C.H.

DISCLOSURES

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