

Ganglioneuroma of the Sacrum

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Presacral ganglioneuromas are extremely rare benign tumors and fewer than 20 cases have been reported in the literature. Ganglioneuromas are difficult to be differentiated preoperatively from tumors such as schwannomas, meningiomas, and neurofibromas with imaging modalities. The retroperitoneal approach for resection of presacral ganglioneuroma was performed for gross total resection of the tumor. Recurrence and malignant transformation of these tumors is rare. Adjuvant chemotherapy or radiation therapy is not indicated because of their benign nature. We report a case of a 47-year-old woman with a presacral ganglioneuroma.

Key Words: Ganglioneuroma, Presacral, Anterior retroperitoneal approach

INTRODUCTION

Ganglioneuroma is an uncommon benign tumor of neural crest origin which is mainly localized in the posterior mediastinum, retroperitoneum, and adrenal gland^{1,6}. Presacral ganglioneuromas are extremely rare and known as predominantly occurring in women¹⁵. Patients are usually young and show neurological signs and symptoms when the tumor developing a significant size^{4,7}. As imaging modalities have become more widely performed, the number of ganglioneuromas found has increased. However, it is very difficult to diagnose ganglioneuroma early in the evaluation of the lesion using image modalities only¹⁴. We report our experience with a case of presacral ganglioneuroma.

CASE REPORT

A 47-year-old woman presented to hospital with a month's duration of intermittent flank and iliac fossa pain, abdominal discomfort, and tingling sensation on left thigh. She had a history of transvaginal myomectomy with symptoms of mild, temporary diarrhea and abdominal discomfort 3 years prior to admission. There was no specific family history. There were no palpable masses observed on abdominopelvic examination. Computed tomography (CT) scans of the abdomen and pelvis revealed a finely demarcated, oval shaped mass arising separately from the ovary on the right side of her pelvis (Fig. 1). Pelvic magnetic resonance (MR) images showed an elliptical tumor on the left side of the pelvis which were well enhanced by contrast (dimensions: 3.4 cm×2.4 cm×4.5 cm). The tumor was adjacent to the left sacrum and

displacing the left sacral nerve roots, without any evidence of bony invasion (Fig. 2).

We performed surgery via anterior retroperitoneal approach and meticulous adhesiolysis was necessary because of massive abdominal adhesion due to the previous gynecologic surgery. Especially, the left internal iliac vein and tumor capsule were adherent and were carefully dissected. We could discern the left S1 root using a nerve stimulator and the tumor was attached to the S1 nerve in the posterior side. Since the tumor was partly in the left S1 sacral foramen, the foramen was unroofed with drill and Kerison punch to expose and remove the mass completely. The encapsulated retroperitoneal tumor located on the left side of the sigmoid colon was excised by piecemeal pattern and complete resection was performed (Fig. 3). Postoperative histopathologic study confirmed the lesion as a ganglioneuroma (Fig. 4). After the surgery, the patient recovered and discharged without any neurological deterioration.

DISCUSSION

Ganglioneuromas are well-differentiated, rare benign tumors of neural crest origin. Especially, presacral location of ganglioneuroma is extremely rare^{10,13,17,18}. Ganglioneuromas often locate in the posterior mediastinum and retroperitoneum. The tumors usually show a slow-growing pattern with a predominance in women¹⁵. Most tumors are diagnosed at the progressed state of the tumor in patients between 10 and 30 years of age because symptoms only appear when the mass becomes large enough to exert a mass effect^{4,7}. Ganglioneuromas can occur anywhere in the autonomic nervous system and can cause a mass effect on the spinal

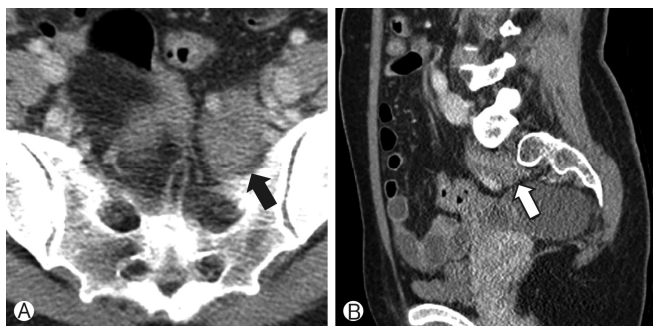


Fig. 1. A pelvic computed tomography scan shows a well-demarcated and heterogeneous mass arising from the left S1 sacral foramen (arrow). Axial (A) and sagittal (B) view.

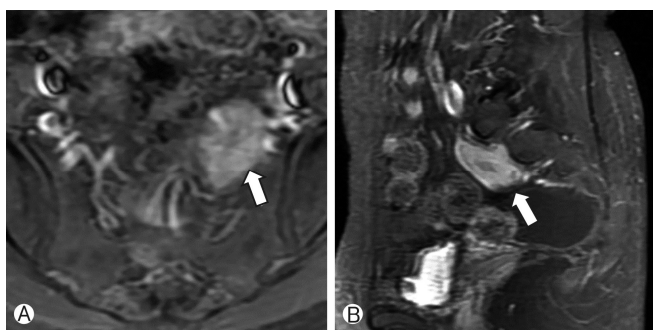


Fig. 2. Magnetic resonance images show a presacral 3.4×2.4×4.5-cm mass (arrow). Contrast enhanced T1-weighted magnetic resonance image with intermediate high signal intensity. Axial (A) and sagittal (B) view.

nerves and sympathetic nerves, leading to neurological abnormalities such as back pain, neurogenic bladder, urinary frequency, constipation, and bowel obstruction. Spontaneous or radiotherapy-induced malignant transformation of ganglioneuromas has been reported but this is extremely uncommon²⁾.

According to our literature search, fewer than 20 cases of presacral ganglioneuroma have been reported worldwide. Ganglioneuromas arise from sympathoblasts derived from the embryonic neural crest. Histologically, ganglioneuromas consist of Schwann cells with mature ganglion and are considered to be part of the neuroblastoma group, along with neuroblastomas and ganglioneuroblastomas^{3,9,10,13,14,19)}. Preoperative evaluation could be performed by using fine-needle aspiration biopsy (FNAB), but the approach is often technically difficult because of the anatomical location. Also, using a single FNAB sample can lead to inaccurate diagnoses and cells should be sampled at multiple sites within the tumor⁸⁾.

It is not easy to differentiate between ganglioneuroma and other tumors (such as schwannoma), only with clinical or imaging findings. There is no case report that patient was diagnosed with ganglioneuroma preoperatively with imaging modalities¹⁴⁾. Magnetic resonance imaging (MRI) is a tool that not only helps to assess the nature of the lesion but also helps in planning of

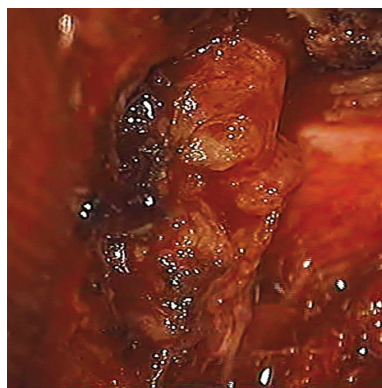


Fig. 3. Intraoperatively, a tumor is apparent after unroofing the left S1 sacral foramen.

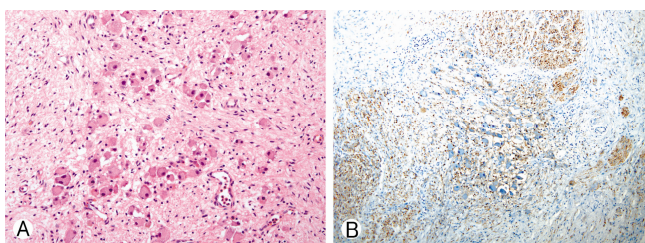


Fig. 4. Histopathologic findings of ganglioneuroma. (A) Hematoxylin and Eosin staining with ×200 magnification. The specimen was composed of fibrous and edematous matrix with nests of ganglion cells and Schwann cells in the matrix. (B) S-100 and neurofilament staining at ×100 magnification.

the surgery.

CT scans of ganglioneuromas may have dot-like calcifications in two-thirds of cases (this was not seen in the reported case), which can be a clue for differential diagnosis with neuroblastomas¹¹⁾. MRI helps in planning treatments by distinguishing lesions of spinal origin from lesions of pelvic origin^{14,16)}. On MR images, ganglioneuromas may show a high signal intensity during late enhancement and a heterogeneous mass on T2-weighted image. The structural and morphological characteristics of ganglioneuromas, such as the presence of mature sympathetic ganglion cells, can help distinguish them from other pelvic benign tumors such as schwannomas, meningiomas, or neurofibromas^{4,6,7,11)}. Appropriate therapy for ganglioneuromas is complete surgical excision^{2,5)}. The complete resection of presacral ganglioneuroma provides for both effective diagnosis and treatment. Adjuvant chemotherapy or radiation therapy is not indicated due to the nature of this benign disease¹²⁾.

In our case, an anterior retroperitoneal approach was used, which is a effective method for mass removal of presacral lesions because it provides a sufficient field of view to identify the origin of the lesion. This approach provided excellent access to the lesion and facilitated complete resection. Other groups have also reported that the posterior transsacral approach was proposed to remove a tumor completely involving the sacral nerve root. Modha et al.¹⁴⁾ prefer the anterior approach, but a staged operation of laminectomy and foraminotomy can be added to manage

any radicular symptoms after removal of the tumor.

Recurrence and malignant transformation of this tumor have been rarely reported²⁾. In that case, the only treatment available is surgical resection. The patients' prognosis is usually good and there is no consensus as to a follow-up plan.

CONCLUSION

Ganglioneuromas could be considered as a differential diagnosis in the treatment of presacral tumors. Anterior retroperitoneal approach is efficient method in complete removal of the tumor without neurological deficit.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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