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Case Report

Presacral Ganglioneuroma in a 30-Year-Old Female: Case Report and Literature Review

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Abstract

Background: Ganglioneuromas are rare, benign tumors derived from neural crest cells of the sympathetic nervous system. These tumors are typically found in the posterior mediastinum and retroperitoneum, and are characterized by well-differentiated ganglion and Schwann cells. Presacral ganglioneuromas are extremely uncommon, with few cases documented in the literature. Their rarity and nonspecific clinical presentation can pose diagnostic challenges. It is worth mentioning that there are few cases in the literature report ganglioneuroma in association with MEN type 2B. *Case Presentation:* We report the case of a 30-year-old female who presented with a three-month history of mild lower abdominal discomfort and non-radicular lower back pain. Her physical examination was unremarkable, and routine laboratory investigations, including tumor markers and catecholamine levels, were within normal limits. Initial ultrasonography was inconclusive. However, contrast-enhanced computed tomography (CT) revealed a well-circumscribed, 5 × 3 cm lesion in the presacral region, causing widening of the right middle sacral foramina. Magnetic resonance imaging (MRI) further characterized the mass as isointense on T1-weighted imaging and heterogeneously hyperintense on T2-weighted imaging, with peripheral enhancement after gadolinium administration. Given its location and imaging characteristics, the lesion was surgically excised via microscopic resection. The postoperative course was uneventful, and the patient was discharged in stable condition. Histopathologic analysis confirmed the diagnosis of ganglioneuroma, revealing mature ganglion cells embedded in a schwannian stroma with fibrous and edematous areas. *Conclusion:* This case underscores the importance of considering ganglioneuroma in the differential diagnosis of presacral masses. Timely imaging and surgical intervention are key to effective management and favorable outcomes. We present a rare case of a presacral ganglioneuroma in a 30-year-old female patient who experienced mild lower abdominal discomfort and non-radicular lower back pain over a three-month period, without significant neurological symptoms. Diagnostic imaging revealed a presacral lesion at the S2-3 level, leading to a surgical intervention that resulted in a subtotal excision of the mass. Histopathological analysis confirmed the diagnosis of ganglioneuroma. This case underscores the importance of considering presacral ganglioneuroma in the differential diagnosis of presacral lesions, given their subtle presentation and potential complications.

Keywords

Presacral Mass, Ganglioneuroma, Abdominal Pain, Back Pain, Surgery

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1. Introduction

Ganglioneuromas are rare, benign neuroblastic tumors that can arise in various regions of the body, with a predilection for the posterior mediastinum and retroperitoneum. [1, 9, 12, 14]

These tumors originate from sympathetic nerve cells, and their degree of cellular differentiation determines their nature: poorly differentiated cells give rise to neuroblastomas, while well-differentiated cells form ganglioneuromas. [2, 10, 11, 15]

In this report, we present a unique case of a presacral ganglioneuroma discovered in a 30-year-old female, further contributing to the understanding of this uncommon neoplasm.

2. Case Presentation

A 30-year-old female presented to our hospital with a three-month history of mild lower abdominal pain and non-radicular lower back pain. Her past medical and surgical history was unremarkable, and she reported no known allergies. The patient had no neurological symptoms, remained physically active, and experienced no changes in bowel habits. Routine blood tests, including tumor markers (α-fetoprotein, carcinoembryonic antigen, cancer antigen 15-3, cancer antigen 19-9, and cancer antigen 125), were within normal limits. Additionally, laboratory studies showed no evidence of catecholamine excess.

Abdominal ultrasonography (performed at an external facility, details unavailable) yielded unremarkable findings. Subsequent abdominal Computed Tomography (CT) revealed a well-circumscribed lesion in the presacral region, measuring 5 × 3 cm in size with a rim of peripheral enhancement after contrast administration, which mostly originated from the neural canal, causing widening of the right middle neural sacral foramina and remodeling of the adjacent structures with no calcifications. (Figure 1).

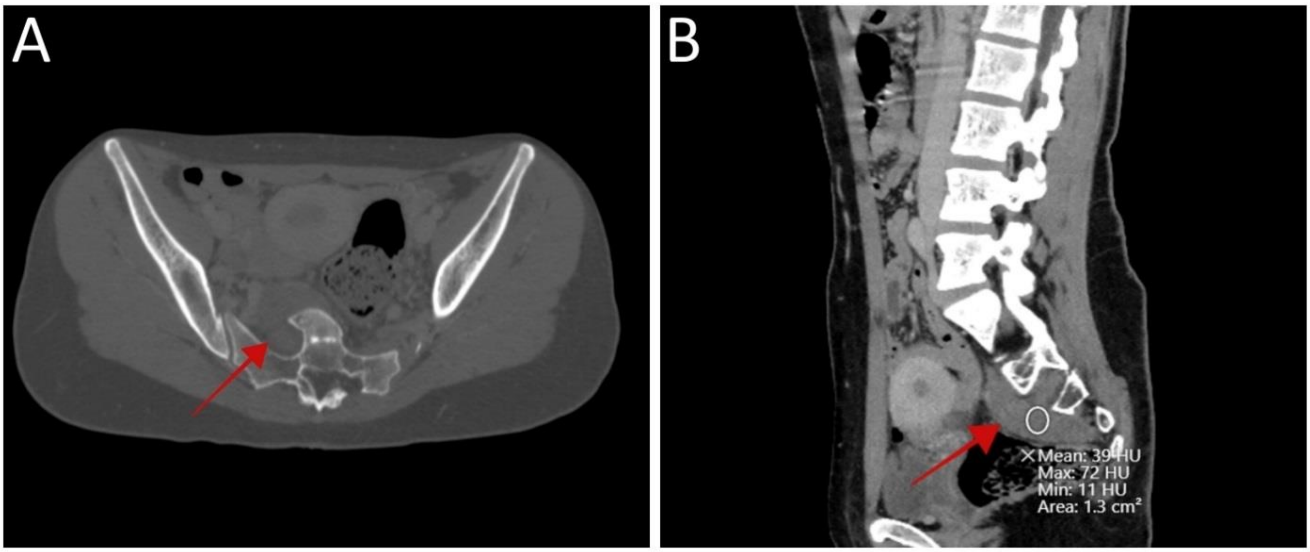

Figure 1. *Pre-operative CT scan with contrast displaying well-circumscribed lesion in the presacral area with a rim of peripheral enhancement causing widening of the right middle neural sacral foramina and remodeling of the adjacent structures with no calcifications (red arrow) and the mean Hounsfield unit is 39: (A) axial view and (B) sagittal view. [CT = computed tomography, HU = Hounsfield unit]*

Subsequent pelvic magnetic resonance imaging (MRI) showed a rounded lesion opposite to S2-S3 level foraminal and extraforaminal lesion extended to presacral region, isointense on T1-weighted image (T1WI) (Figure 2A), heterogeneous hypointense on T2-weighted images (T2WI) (Figure 2B) with peripheral enhancement after GAD administration (Figure 2C and 2D).

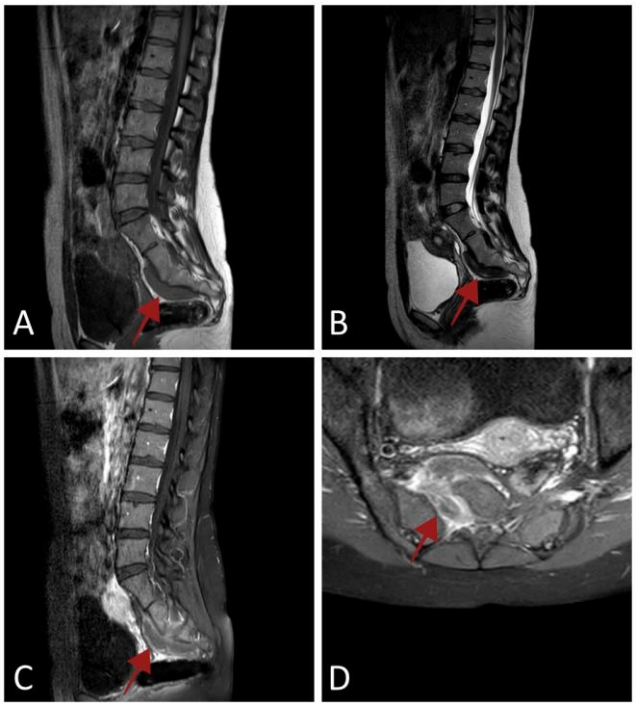

Figure 2. *Pre-operative pelvic MRI exhibiting: (A) an isointense presacral lesion opposite to S2-S3 level on sagittal T1WI, (B) a heterogeneous hyppintense on sagittal T2WI, and (C: sagittal and D: axial) a peripheral enhancement after GAD administration on T1WI.*

[MRI = magnetic resonance imaging, T1WI = T1-weighted images, T2WI = T2-weighted images, GAD = gadolinium]

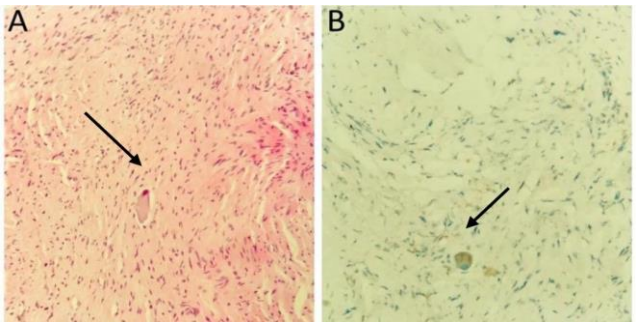

Figure 3. *Histopathological slide showing: (A) an intersecting bundles of schwann cells (arrow) and scattered nests and clusters of ganglion cells [H&E stain, original magnification ×20], and (B) synaptophysin immunostain highlights ganglion cell (arrow).*

Based on the MRI findings, the decision was made to proceed with a microscopic surgical resection, aiming for the maximum safe removal of the lesion. The surgery was successfully performed without any complications. Postoperatively, the patient was closely monitored and maintained in excellent general condition, with stable vital signs throughout. She was discharged on the fifth postoperative day, having recovered well.

Histopathologic examination (Figure 3) confirmed the presence of a ganglioneuroma, characterized by schwann cells mixed with ganglion cells. Fibrous and edematous matrix are also seen.

3. Discussion

Presacral ganglioneuroma is an exceedingly rare tumor arising from the sympathetic chain in the presacral region. Accurate recognition of its clinical presentation and a thorough understanding of its pathophysiology are essential for timely and effective intervention. This work seeks to underscore the clinical features of presacral ganglioneuroma, promoting quicker diagnosis and the delivery of high-quality care.

Clinical studies have shown that presacral ganglioneuroma can present with various symptoms, such as vague abdominal pain and bowel or bladder disturbances. Due to these nonspecific symptoms, the tumor can be easily mistaken for other conditions [4, 8, 12]. Therefore, it is essential for physicians to consider this rare entity in their differential diagnosis to ensure appropriate management. It is worth noting that several case reports in the literature have described an association between ganglioneuroma and MEN type 2B. Therefore, it is important to consider this rare but significant association when evaluating a patient diagnosed with ganglioneuroma [16].

Diagnostic modalities like CT and MRI are vital for identifying and evaluating presacral tumors. [5, 9] However, a definitive diagnosis requires a biopsy of the lesion. [6, 11, 15] One of the primary challenges in treating these tumors is achieving complete surgical excision, which can be complex and carries a significant risk of morbidity. Nevertheless, debulking with maximum safe resection remains a key treatment priority. [7, 10, 13] Multidisciplinary team discussions are highly recommended to provide optimal patient care.

While some studies suggest that chemotherapy or radiotherapy may aid in the management of presacral ganglioneuroma, especially when complete excision is not feasible, the evidence supporting their effectiveness is limited. [3, 9, 14] Further high-quality research is needed to determine the role of adjuvant therapy, particularly in cases of incomplete excision or recurrence.

In conclusion, this work underscores the clinical aspects of presacral ganglioneuroma, emphasizing the importance of early diagnosis and appropriate management. Given the tumor's rarity and nonspecific presentation, physicians should maintain a high index of suspicion. Timely diagnosis and intervention are crucial for successful patient outcomes.

4. Conclusion

Ganglioneuromas could be considered as a differential diagnosis in the diagnosis of presacral tumors.

Abbreviations

MEN	Multiple Endocrine Neoplasia
CT	Computed Tomography
HU	Hounsfield Unit
MRI	Magnetic Resonance Imaging
T1WI	T1-weighted Image
T2WI	T2-weighted Image
GAD	Gadolinium
H&E	Hematoxylin and Eosin

Patient Consent

Written informed consent was obtained from the patient.

Conflicts of Interest

The authors declare no conflicts of interest.

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