

## Ganglioneuroma of Lumbar Region

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**Abstract:** Ganglioneuromas (GNs) arising from neural crest sympathogonia are rare benign neurogenic tumors. The most commonly affected sites are the posterior mediastinum, the retroperitoneum and the adrenal gland. Here, we are presenting such a case in a 53-year old female patient with complaints of nonspecific, non-colic, intermittent pain, in the lumbar region predominantly on the left side. Clinically it was diagnosed as nerve sheath tumour or lipoma and surgery was done and confirmed by microscopic study.

**Keywords:** Ganglioneuroma, lumbar region, asymptomatic.

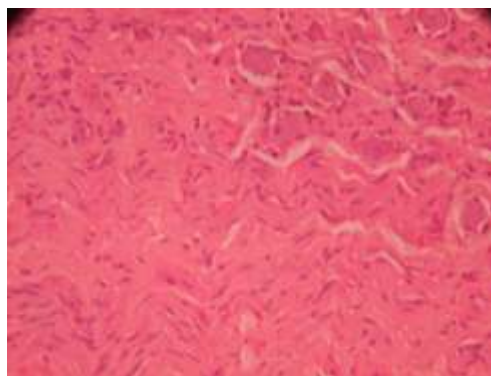
### INTRODUCTION

Ganglioneuromas (GNs) arising from neural crest sympathogonia are rare benign neurogenic tumors. The most commonly affected sites are the posterior mediastinum, the retroperitoneum and the adrenal gland. GNs often present as a solitary, painless and slow-growing mass. GNs are fully differentiated tumours that contain no immature elements. They are rare compared with other benign neural tumours, such as schwannoma and neurofibroma, but they outnumber neuroblastomas along the sympathetic axis by about 3 to 1. Surgical excision is the treatment of choice and the prognosis is excellent even in cases where complete excision cannot be achieved. Ganglioneuromas most commonly arise from sympathetic ganglia [1]. These neoplasms may be located wherever ganglion cells are normally found from the skull base to the pelvis, including the adrenal medulla. They are usually asymptomatic and hormonally silent. Majority of cases are detected incidentally during work-up for unrelated conditions. Currently, histopathologic examination is the only tool to diagnose ganglioneuroma and to differentiate it from other neural crest tumors. Here we present a case of GN in the lumbar region.

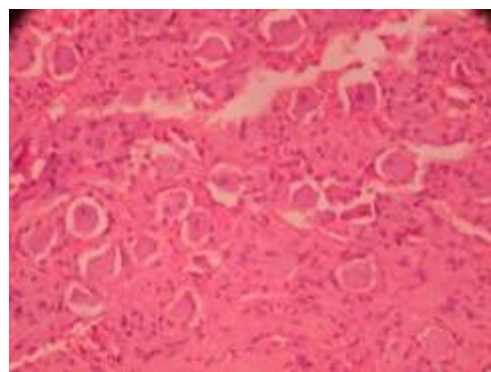
### CASE REPORT:

A 53-year old female patient was admitted with complaints of nonspecific, non-colic, intermittent pain, in the lumbar region predominantly on the left side. She had pain for two months. The patient's history did not contain any systemic diseases or previous operations. Physical examination revealed a swelling of size 2x2cm, firm with restricted mobility. Clinically it was diagnosed as nerve sheath tumour or lipoma. Surgery was done and we received a 2x1.5cm greywhite soft tissue mass, firm which was submitted fully. Microscopy showed clusters of ganglion cells in

a background of bundles of longitudinal and transversely originated Schwann cells that crisscross each other in an irregular fashion which was confirmed as GN [ Figure 1 & 2]



**Fig-1: Section showing Ganglion cells in a background of Schwannian stroma.**



**Fig-2: Section showing clusters of ganglion cells in a background of bundles of longitudinal and transversely originated Schwann cells that crisscross each other in an irregular fashion.**

## DISCUSSION

GN originates from cells of the neural crest that include the sympathetic ganglia and the adrenal glands. It is composed of ganglion cells, neurites, schwann cells, and fibrous tissues. This tumor affects children and young people preferentially with three-fifth developed before age 20. Females are more prone to be affected than males. Majority of GNs are thoracic and retroperitoneal in location while adrenal GNs are rare.

Clinical symptoms of GNs are non-specific, mostly hormonally silent and related to their size/locations. Despite their generally benign nature, GNs may come to attention due to compression of their neighboring structures [2]. Approximately up to 30% of patients were found to have elevated plasma and urinary catecholamine but they rarely develop symptoms of vasoactive amines excess. Immature element such as neuroblast is not part of mature GN explaining lower rate of metaiodobenzylguanidine (MIBG) uptake (57%) compared to neuroblastoma (92%) [3]. The most significant differential diagnosis of ganglioneuroma is neuroblastoma. Increased level of urinary noradrenaline, dopamine, HVA, and VMA are frequently encountered in neuroblastoma, while the level of urinary catecholamine, HVA, and VMA are usually normal in ganglioneuroma [4].

Peripheral neuroblastic tumors represent a spectrum of diseases from undifferentiated and malignant NB to well-differentiated and benign GN. According to the international neuroblastoma pathology classification (the Shimada System), neuroblastic tumors are classified into 4 groups: NB (schwannian stroma-poor), intermixed GNB (schwannian stroma-rich), nodular GNB (schwannian stroma-rich/stroma-dominant and stroma-poor) and GN (schwannian stroma-dominant), which is further divided into 2 subtypes (maturing and mature). Typical GN is composed of mature ganglion cells and schwannian stroma; however, complete maturation of ganglion cells is rare (approximately 7%) and slightly atypical ganglion cells could be detected in the present case. Immaturity of ganglion cells did not influence the diagnosis of GN.

Complete surgical excision is the treatment of choice, in which a transcervical or transoral approach may be employed depending on the location of the tumor. The risks are mainly related to the intraoperative sacrifice of the neural structures and the vasculature associated with the tumor. The prognosis of GN is usually good.

## REFERENCES

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