Giant Ganglioneuroma of Thoracic Spine: A Case Report

Das S¹, Alam K², Rashid MM³, Islam MM⁴, Mahbub H⁵, Khan MSI⁶

Abstract:
Ganglioneuromas most commonly arise from sympathetic ganglia. These neoplasms may be located wherever ganglion cells are normally found from the skull base to the pelvis, including the adrenal medulla. We describe a 15-year-old girl with giant Ganglioneuromas in the thoracic spine, who underwent successful resection (T4-5 level) of the tumor. Histopathological examination confirmed the diagnosis. Ganglioneuromas should be considered in the differential diagnosis of any paraspinal mass. A high index of suspicion and correlation of clinico-radiological findings is necessary in differentiating a large benign tumor from a malignant growth. Complete surgical excision is the treatment of choice; however, tumor size and location need to be considered for the surgical approach (one-step or multiple surgeries). Close follow-up after surgery is mandatory.


Introduction:
Ganglioneuromas are rare benign tumors that originate from a neural crest or sympathetic ganglion¹. They most commonly appear in the posterior mediastinum and abdomen². The patients exhibit no obvious symptoms upon nervous system examination. The ganglioneuromas are often found in females, while the male/female ratio is approximately 2/3³. The incidence of ganglioneuroma is not well documented, but it is estimated to characterize 0.1 to 0.5 % of total central nervous system (CNS) tumors⁴. Paravertebral ganglioneuroma and scoliosis is rarer and has only been sporadically reported. We report a giant paraspinal Ganglioneuroma extending into extradural space and thoracic cavity.

1. Dr. Sukriti Das, Associate Professor, Department of Neurosurgery, Dhaka Medical College & Hospital, Dhaka.
2. Dr. Kamrul Alam, Professor and Head of the Department of thoracic surgery, Dhaka Medical College & Hospital, Dhaka.
3. Dr. Md. Mamunur Rashid, Phase-B, Resident, Department of Neurosurgery, Dhaka Medical College & Hospital, Dhaka.
4. Dr. Md. Manirul Islam, Medical Officer, Department of Neurosurgery, Dhaka Medical College & Hospital, Dhaka.
5. Dr. Hasan Mahbub, Phase-B, Resident, Department of Neurosurgery, Dhaka Medical College & Hospital, Dhaka.
6. Dr. Mohammed. Shamsul Islam Khan, Medical Officer, Department of Neurosurgery, Dhaka Medical College & Hospital, Dhaka.

Address of Correspondence: Dr. Sukriti Das, Associate Professor, Department of Neurosurgery, Dhaka Medical College & Hospital, Dhaka, Cell: +8801711676848, e-mail: sukriti66@yahoo.com,
Discussion:
About 10% of Ganglioneuromas may involve the spinal canal. Paraspinal Ganglioneuromas can extend into the spinal canal, forming dumbbell shaped tumor. However, in rare cases intradural extension has been reported. Most Ganglioneuromas are incidentally detected, and the symptoms, if any, are usually due to the mass effect. Rarely, the tumor may secrete vasoactive intestinal polypeptide, resulting in diarrhea. As this slow growing tumor extends through the neural foramen into the spinal cord, some patients may present with neurological deficits or scoliosis. Thoracic intradural extramedullary Ganglioneuromas are very rare. In our patient, the characteristic feature was the remarkably large tumor size infiltrating into the thoracic cavity along with an intradural component. Microscopically, these tumors contain large ganglion cells and show areas with smaller lymphocyte-like cells within a matrix of fibrous stroma and schwann cells. The distinction from malignant tumor is based on the absence of necrosis or presence of any immature ganglion cells.

It is usually safe and feasible to perform complete excision of Ganglioneuromas. However, in case of multiple and/or large-sized tumors, multi-stage dissection should be considered. In the present case, there were dense adhesions of the tumor with the nerve roots at the foraminal portions, which were left undisturbed during dissection. Ganglioneuromas generally has a favorable prognosis given its low metastatic potential.

Conclusion:
This report describes a rare case of multiple Ganglioneuromas of the thoracic spine with intradural and thoracic extension. Ganglioneuromas should be considered in the differential diagnosis of a paraspinal mass. Although complete surgical resection is the best treatment option, stage-wise surgical resection should be considered in large-sized and/or multiple tumors, with close follow up.

References:


