Giant Ganglioneuroma of Thoracic Spine: A Case Report

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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.

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Introduction:

Ganglioneuromas are rare benign tumors that originate from a neural crest or sympathetic ganglion[1]. They most commonly appear in the posterior mediastinum and abdomen[2]. The patients exhibit no obvious symptoms upon nervous system examination. The ganglioneuromas are often found in females, while the male/female ratio is approximately 2/3[3]. 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) tumors[4]. 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.

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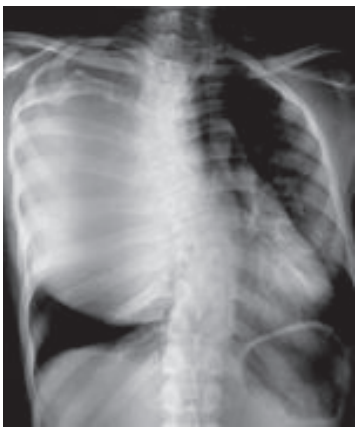
Case Report:

A 15-year-old girl presented with a one month history of progressive onset lower extremity weakness. The patient had no other significant complaints. On examination, she was unable to stand or walk without support.

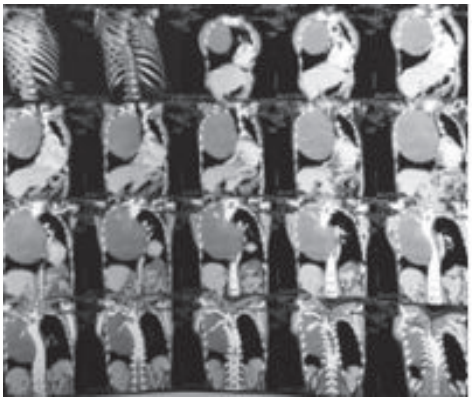
A neurological examination revealed 3/5 strength in all muscle groups in the patient's lower extremities. Sensory was markedly decreased below the nipple line. Deep tendon reflexes were hyperreflexic throughout the lower extremities along with markedly increased tone, right greater than left. Sustained clonus was noted at both ankles.

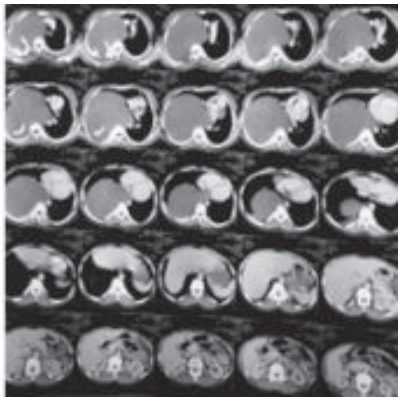
Magnetic resonance imaging showed a huge well defined mixed signal intensity oval lesion in posterior mediastinum predominantly on the right side. The lesion predominantly hypointense in T1WI and predominantly hyperintense in T2WI. Foci of signal void areas within the lesion represents calcification T1WI and T2WI increased signal intensity areas represents fatty component. The lesion causes collapse of right lung sparing basal segments of right lower lobe. Mediastinal structure are pushed of left. Epidural extension of the lesion is noted into the spinal canal causing widening of the right neural foramen resulting compression over the spinal cord and corresponding nerve root compression at D4-D5 level. Spinal cord pushed to left.

Right Posterolateral thoracotomy was done. A huge tumor occupying right paravertebral space pushing the right lung medially and inferiorly. It has a broad base, Posterior end of right 4th rib thin out and became triangular shaped. Tumor invaded in between the space.

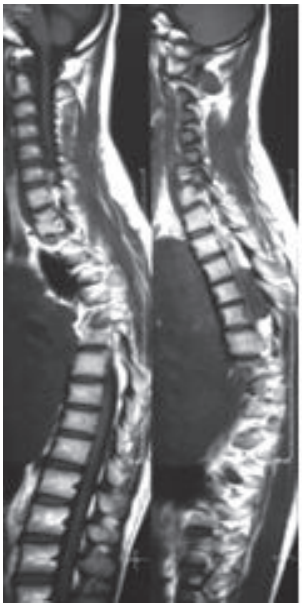


Pre-op chest x-ray

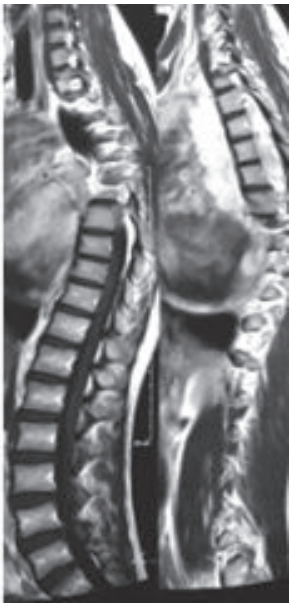




Pre-op CT scan of chest

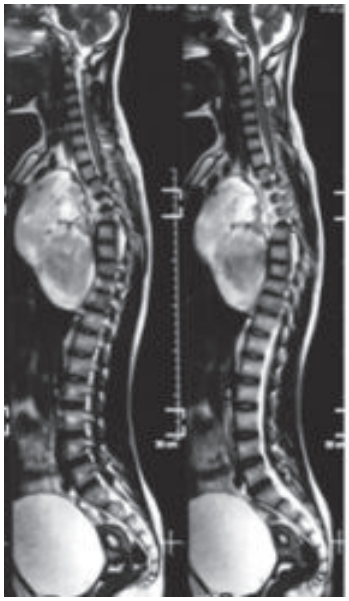


A. Non-contrast

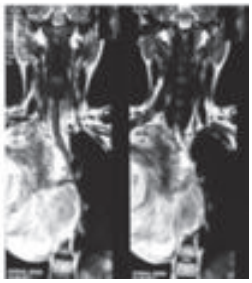


B. Contrast

Pre-op MRI of dorsal spine- sagittal (T1)

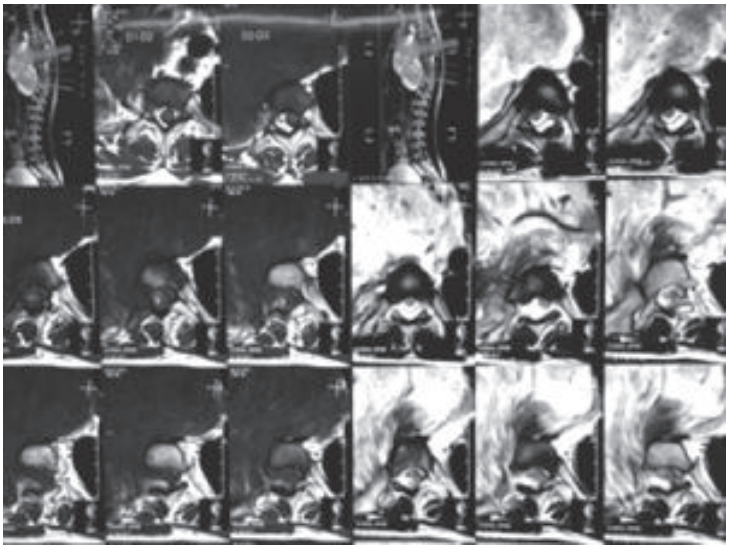


Sagittal section

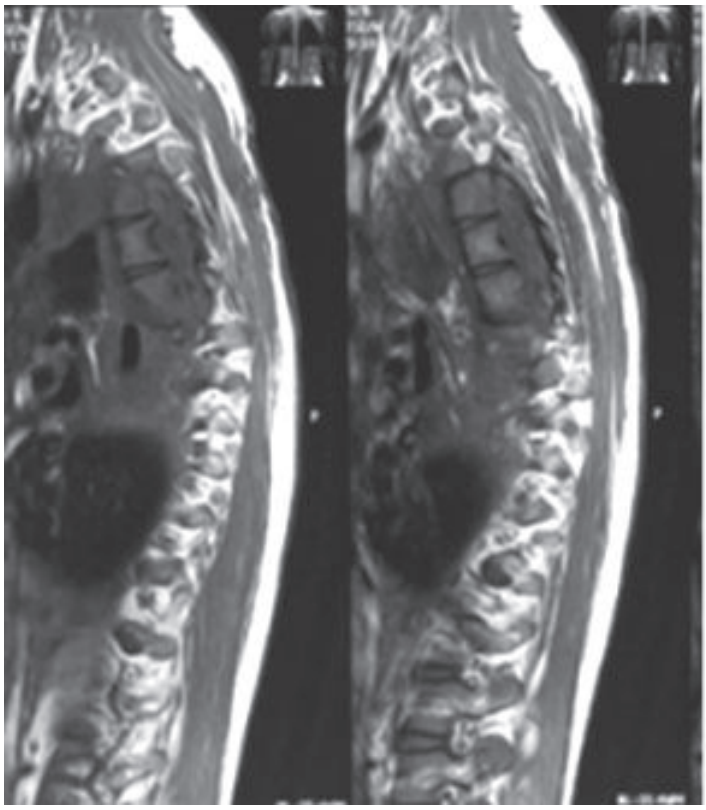


Coronal section

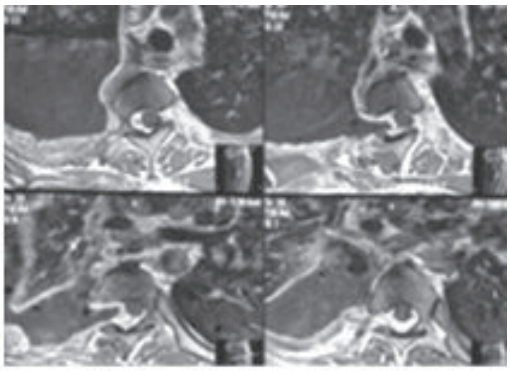
Pre-op MRI of dorsal spine- (T2)



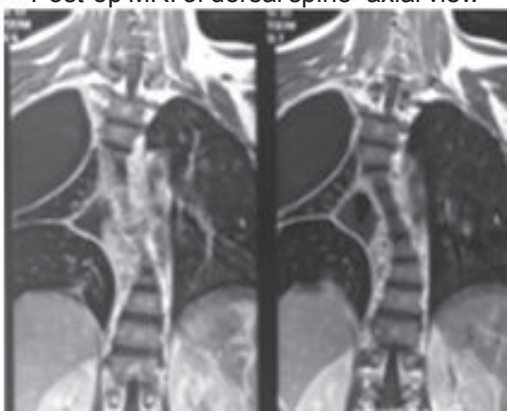
Pre-op MRI of dorsal spine Axial section



Post-op MRI of dorsal spine- sagittal view



Post-op MRI of dorsal spine- axial view



Post-op MRI of dorsal spine- coronal view

Discussion:

About 10% of Ganglioneuromas may involve the spinal canal[5]. Paraspinal Ganglioneuromas can extend into the spinal canal, forming dumb bell 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 polypetide, 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[6]. 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[7].

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[8].

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.

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