



Ganglioneuroblastoma: Spinal with Thoracic Extension- A Rare Diagnosis

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Abstract

Background: Ganglioneuroblastoma are rare tumors with origin from the neural crest cell. Spine is very rare site for their occurrence. In the spinal canal, the most common location is thoracic, intramedullary site. Our case is significant because it was located in the thoracic spine extending extradurally into the thoracic cavity, first reported in the literature.

Case Presentation: We report a 13-year old female admitted with paraplegia and urinary retention. Sensations were lost below D8 level. Magnetic resonance imaging showed an irregular enhancing lesion in extradural location from D4-D9 vertebra with intrathoracic extension on right side. Laminectomy (D5-D9) and right rib resection (rib 7 and 8) was done. Subtotal excision of the thoracic mass was achieved. D5,6-D9,10 right transpedicular fixation was done. Adjuvant radiotherapy was advised. After 8 months of follow up, the patient's power in both lower limbs was grade 2.

Conclusion: Ganglioneuroblastoma in children and young adults should be included in the differential diagnosis of thoracic cord tumors. Definitive diagnosis depends on post-operative histological examination. Curative treatment should be a complete resection of the tumor. Also, in partially resected cases, adjuvant radiotherapy may become necessary, along with close follow up.

Keywords: Ganglioneuroblastoma; Spinal; Thoracic

Introduction

Ganglioneuroblastoma are rare tumors with origin from the neural crest cell. They lie between benign gangliogliomas and frankly malignant neuroblastoma with respect to their malignant potential [1]. Primary occurrence of the tumor in spinal cord is rare [2,3]. We report a case of thoracic extradural ganglioneuroblastoma with intra thoracic extension and discuss its management.

Case Report

A 13-year old female was admitted with a history of mild upper back pain with burning pain in both the lower limbs for 3 months. She started developing weakness in both the lower limbs and 20 days prior to admission to our hospital; she was paraplegic with grade 0 power in both the lower limbs. Simultaneously, she was catheterized for urinary retention. Muscles were grossly hypertonic with exaggerated jerks and clonus. All modalities of sensations were lost below D8 level.

T1-weighted magnetic resonance imaging showed an irregular hypo intense lesion in extradural location from D4-D9 vertebra with intrathoracic extension on right side. On T2-weighted images the lesion was intense relative to normal cord. After contrast administration, there was strong enhancement (Figure 1 and 2).

Operative: Laminectomy (D5-D9) and right rib resection (rib 7 and 8) was done. On extracavitary approach tumor was extradural, firm, moderately vascular, compressing the spinal cord and extending through the neural foramina into the thoracic cavity. Also collected hematoma inside the mass was aspirated. Subtotal excision of the thoracic mass was achieved. D5,6-D9,10 right transpedicular fixation was done (Figure 3).

Hematoxylin-eosin stained sections showed dural tissue with adherent tumor with areas of hemorrhage and necrosis and a small round morphology and focal neurofibrillary rosette's. Foci of calcification were identified (Figure 4A). Other areas show sheets of ganglion cells on a fibrillary background (Figure 4B). Immunohistochemistry revealed a negative staining for glial fibrillary acidic protein, whereas NSE immunoreactivity was positive in most of the cells and Ki-67 positive in 60%.

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Table 1: Cases of thoracic ganglioneuroblastoma in literature.

No.	Study	Age/sex	Diagnosis	Treatment	Outcome
1	Patnaik et al. [1]	25 years/male	T2 Extradural mass	Excision. Radiotherapy refused by patient	Mild improvement at 4 months
2	Sibilla et al. [2]	42 year/male	T5-T8 Intramedullary mass with paraparesis	Excision	Mild improvement
3	Tripathy et al. [3]	15 year/male	T2 Intramedullary and extradural mass	Excision + Radiotherapy	No improvement at 4 months
4	Akgun et al. [8]	8 months/male	T5-T10 Intradural intramedullary	Excision	No post op deterioration

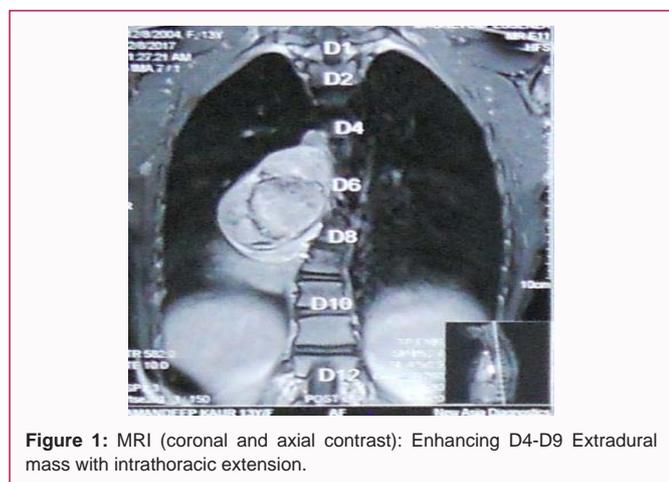


Figure 1: MRI (coronal and axial contrast): Enhancing D4-D9 Extradural mass with intrathoracic extension.

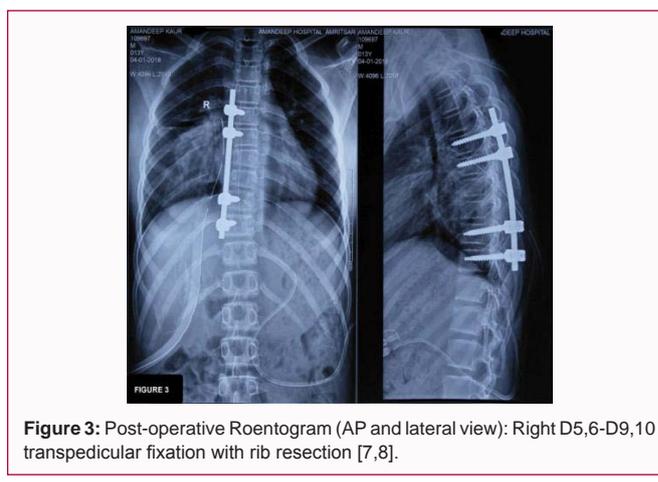


Figure 3: Post-operative Roentogram (AP and lateral view): Right D5,6-D9,10 transpedicular fixation with rib resection [7,8].

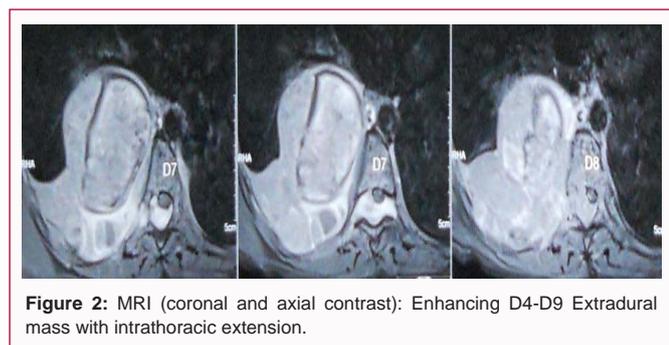


Figure 2: MRI (coronal and axial contrast): Enhancing D4-D9 Extradural mass with intrathoracic extension.

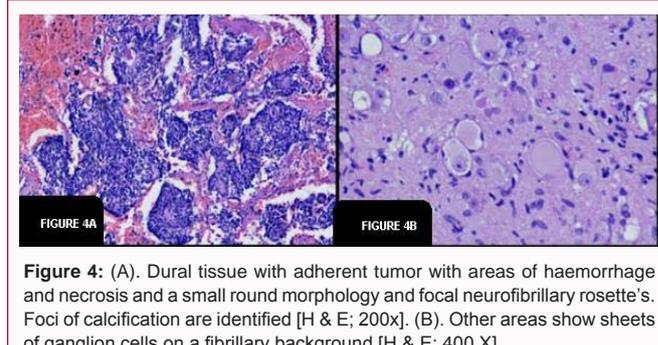


Figure 4: (A). Dural tissue with adherent tumor with areas of haemorrhage and necrosis and a small round morphology and focal neurofibrillary rosette's. Foci of calcification are identified [H & E; 200x]. (B). Other areas show sheets of ganglion cells on a fibrillary background [H & E; 400 X].

The final diagnosis was ganglioneuroblastoma of the spinal cord.

Post-operatively the patient's spasticity in both the lower limbs decreased and there were small flickering movements in both feet. Adjuvant radiotherapy was advised. After 8 months of follow up, the patient's power in both lower limbs was grade 2.

Discussion

Cerebral ganglioneuroblastoma is a term used for tumors which histological consist of undifferentiated small round cells in all stages of neuronal differentiation [4,5]. These are grouped as Primitive Neuroectodermal Tumors (PNETs)

- a) Peripheral or pPNET- those that arise from the sympathetic nervous system
- b) Central or cPNETs- those that arise within the CNS [6].

Three types of embryonal tumors of the CNS have been distinguished by the World Health Organization (WHO) (2007)-medulloblastomas, the PNETs, and the atypical rhabdoid/teratoid tumors. All three types histological correspond to malignant grade 4 tumors. The PNETs include the (ICD-O code 9490/3), medulloepithelioma and ependyoblastoma [4,6]. CNS neuroblastoma and ganglioneuroblastoma, which were earlier,

classified as both neuronal and embryonal tumors by WHO, are now classified only as embryonal tumors [7]. These tumors are usually found in the cerebrum, cerebellum, and suprasellar region. The other common location is posterior mediastinum and retroperitoneum. Spine is very rare site for their occurrence. In the spinal canal, the most common location is thoracic, intramedullary site [1].

Our case is significant because it was located in the thoracic spine extending extradurally into the thoracic cavity, first reported such case in the literature. Other reported cases were localized to the spinal compartment only (Table 1).

Probably the mass eroded the neural foramina and entered the thoracic cavity as was seen intra-operatively.

Ganglioneuroblastoma present from 4 weeks to 20 years. Clinical symptoms are related to the site of origin. In our case, we present a patient with compressive myelopathy at D4-D9 level.

All patients with confirmed diagnosis of ganglioneuroblastoma should receive radio chemotherapy. Radiotherapy should be given as adjuvant treatment even if there is complete excision in patients over 12 years of age. Prognosis remains unfavorable due to aggressive, malignant nature of the tumor with frequent recurrences as well as distant metastases [1]. There was only one reported case of primary

ganglioneuroblastoma in an extradural location in the literature [1]. Our patient had a history of very short duration indicating the aggressive and malignant nature of the tumor. There was no primary lesion either in the brain or retroperitoneum. Ganglioneuroblastoma should therefore be considered in the differential diagnosis of extradural thoracic tumors in a young adult.

Conclusion

Ganglioneuroblastoma in children and young adults should be included in the differential diagnosis of thoracic cord tumors. Definitive diagnosis depends on post-operative histological examination. Curative treatment should be a complete resection of the tumor. Also, in partially resected cases; adjuvant radiotherapy may become necessary, along with close follow up.

Declarations

Ethics approval and consent to participate

This case report has been approved by the Ethics Committee of the hospital and patient party provided written informed consent.

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