

Rare Atypical Presentation of a Neuroblastoma of Posterior Mediastinum

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Abstract

Introduction Neuroblastoma is the third most common malignancy and one of the most common neurogenic tumors of posterior mediastinum in the pediatric age group. A posterior mediastinal tumor causing spinal cord compression is an important example of an oncologic emergency arising from a neurogenic tumor. We present a rare case of posterior mediastinum neuroblastoma, with exclusive lower limb weakness presenting at an unusually older age.

Case Presentation A 6-year-old girl reported with upper back ache and sudden onset progressive bilateral lower limb weakness, with difficulty in urination for 10 days. The weakness had progressed to paraplegia on the day of presentation. On examination, the child was observed to have loss of tone and power in both lower limbs and bilateral extensor plantar reflex. Chest X-ray showed heterogeneous opacity involving most of the left lung. Contrast-enhanced MRI showed posterior mediastinal tumor, extending into spinal canal. Patient underwent D3 to D6 laminectomy with subtotal tumor excision and spinal decompression. Crush biopsy showed round cell tumor, and final histopathological examination revealed poorly differentiated neuroblastoma, which was strongly positive for neuron-specific enolase on immunohistochemistry. The surgery was followed by chemotherapy, and the patient remained symptom free on 2-year follow-up.

Discussion The presenting symptoms in case of thoracic neuroblastoma may include airway obstruction and chronic cough. Sometimes, it may present with weakness, limping, paralysis, and bladder and bowel disturbances in case of spinal cord involvement. Posterior mediastinal neuroblastoma presenting with paraparesis is uncommon. In our case, the child presented with isolated lower limb flaccidity with no other classical symptoms. To the best of our knowledge and thorough study, only four cases of posterior mediastinum neuroblastoma presenting with lower limb paralysis has been published in the English literature to date.

Conclusion Tumor usually presents with pressure symptoms, caused by the mass in the posterior mediastinum, but it's almost exclusive occurrence in the pediatric population warrants a thorough investigation in patients presenting with not only a symptomatic mass but also exclusive lower limb neurological symptoms.

Keywords

- ▶ neuroblastoma
- ▶ paraplegia
- ▶ atypical presentation
- ▶ rare

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Introduction

Neuroblastoma is the third most common malignancy in childhood and one of the most common neurogenic tumors of posterior mediastinum in the pediatric age group.¹ Children commonly present at the age of 2 years, with respiratory symptoms being the usual presentation.² A posterior mediastinal tumor causing spinal cord compression is an important example of an oncologic emergency arising from a neurogenic tumor.³ We present a rare case of posterior mediastinum neuroblastoma, with exclusive lower limb weakness presenting at an unusually older age.

Case Presentation

A 6-year-old girl child with no previous significant medical history reported to our OPD with upper back ache and sudden onset progressive bilateral lower limb weakness, with difficulty in urination for 10 days. The weakness had progressed to paraplegia on the day of presentation. She gave a history of fall from height a month back, following which she was asymptomatic for 20 days.

On physical examination, the child was observed to have loss of tone and power in both lower limbs and bilateral extensor plantar reflex. Chest X-ray showed heterogeneous opacity involving most of the left lung (►Fig. 1). Contrast-enhanced MRI showed posterior mediastinal tumor extending into spinal canal (►Figs. 2 and 3). The blood investigations were normal.

Patient underwent D3 to D6 laminectomy with subtotal tumor excision and spinal decompression. The crush biopsy showed round cell tumor, and final histopathological examination revealed poorly differentiated neuroblastoma, which was strongly positive for neuron-specific enolase on immunohistochemistry. The surgery was followed by chemotherapy, and the patient remained symptom free on 2-year follow-up.



Fig. 1 Chest X-ray.

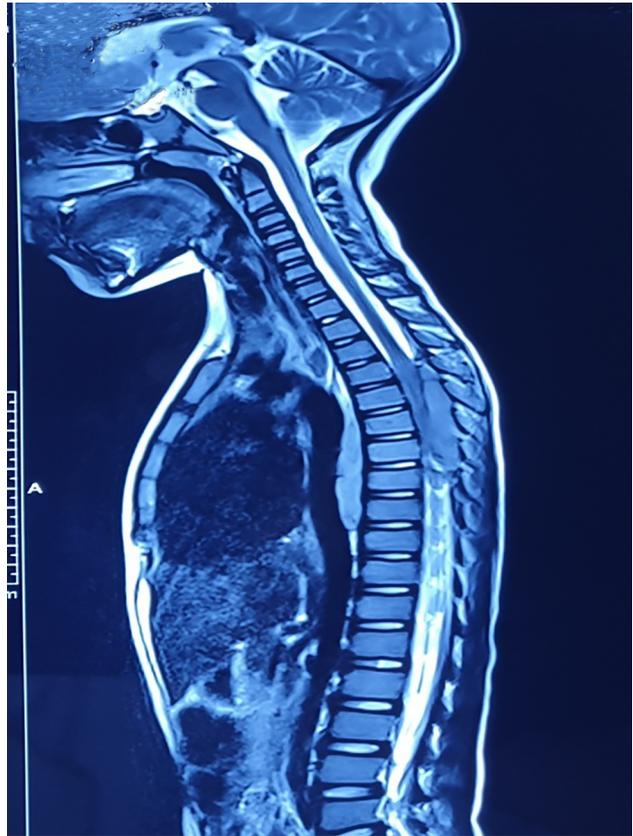


Fig. 2 MRI dorsal spine T2-weighted sagittal image.

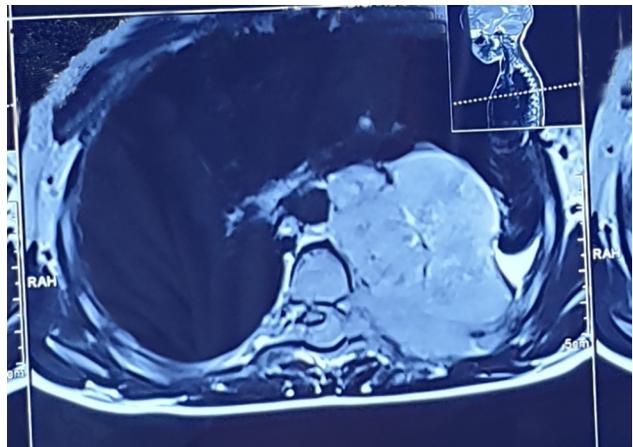


Fig. 3 MRI D6 vertebra T2-weighted axial image.

Discussion

Neurogenic tumor is one of the most common causes of a posterior mediastinal mass, accounting for almost 90 percent of cases. It is an embryonal tumor of the sympathetic nervous system.⁴ Between 7 and 15% of children with neuroblastoma present with spinal cord involvement.⁵

It is difficult to detect neuroblastoma compressing the spinal cord in young children due to apparently slow onset, and progress of paralysis delaying the diagnosis and treatment in

Table 1 List of published cases of neuroblastoma with lower limb paralysis

S. no.	Author	year	Age/sex	Presenting complaints	Treatment	Outcome
1	Mclatchie et al ²	1980	16 months/M	Progressive paraplegia	Thoracotomy and tumor excision	Died at 27 years of age
2	Latchaw et al ¹¹	1981	a)18 months/M b)11 years/M	a) Progressive paraplegia b) Weakness of lower limbs with incontinence	a) Decompression b) Decompressive laminectomy	a) Not mentioned b) Not mentioned
3	Rajparath et al ⁹	2016	11 months/M	Irritability and difficulty in standing	Chemotherapy only	Good
4	Present case	2021	6 years/F	Upper back ache and sudden onset progressive bilateral lower limb weakness	Subtotal tumor excision with decompression for the benefit of chemotherapy	Good

previous cases published.^{6,7} In our case, since the age at presentation was atypically a bit higher, we could diagnose the spinal compression early with early intervention.

The presenting symptoms in case of thoracic neuroblastoma may include airway obstruction and chronic cough. Sometimes, it may present with weakness, limping, paralysis, and bladder and bowel disturbances in case of spinal cord involvement.⁸ Posterior mediastinal neuroblastoma presenting with paraparesis is uncommon.⁹ In our case, the child presented with isolated lower limb flaccidity with no other classical symptoms.

Emergency MRI is the imaging of choice. Chemotherapy and laminectomy is the modality of treatment for neuroblastoma causing spinal cord compression.⁷ In our case, we got the MRI as soon as the child presented to us in the OPD, which helped in early laminectomy and tumor excision followed by chemotherapy.

Usually, most recurrences are seen within 2 years of treatment, and if there is no recurrence detected in 5 years after treatment, the disease is said to be cured.¹⁰ We have not found any recurrence on radiological surveillance till 2 years posttreatment and the child is on follow-up to date.

To the best of our knowledge and thorough study, following are the cases of posterior mediastinum neuroblastoma presenting with lower limb paralysis in the English literature to date (► **Table 1**).

Conclusion

Although this tumor usually presents with pressure symptoms caused by the mass in the posterior mediastinum, the fact is that, although this tumor is a common cause of posterior mediastinal mass, the tumor itself in the general population is rare. However, it's almost exclusive occurrence in the pediatric population warrants a thorough investigation

in patients presenting with not only a symptomatic mass but also exclusive lower limb neurological symptoms.

Conflict of Interest

None declared.

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