

A Rare Pediatric Tumor of the Posterior Mediastinum: Ganglioneuroblastoma

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ABSTRACT

Ganglioneuroblastoma is a rare malignant pediatric tumor located in the posterior mediastinum. A giant mass lesion located in the posterior mediastinum and extending into the spinal canal was observed in a 4-year-old girl admitted to our hospital with walking difficulties. The mass was completely excised with a joint operation of neurosurgery and thoracic surgery. After the operation, the patient regained walking functions.

INTRODUCTION

Neurogenic tumors are the most common mediastinal tumors in children, 20% have a malignant course, and neuroblastomas are the most common malignant neurogenic tumors.^[1] Neurogenic tumors of the mediastinum in children and adults may originate from the nerve sheath, sympathetic ganglion, paraganglion cells, and peripheral neuroectodermal tissue.^[2] Ganglioneuroblastomas are malignant lesions originating from the sympathetic ganglion.

Mediastinal tumors are mostly asymptomatic. However, as the size of the tumor increases, pressure-related symptoms occur. In addition to symptoms such as dyspnea, cough, and chest pain, neurologic symptoms may occur after invasion into the surrounding tissue.^[3]

Pathology Neuroblastomas show S-100 expression in immunohistochemical staining, stained with CD34 positive.^[4] Ganglioneuroblastomas are rarely seen in the mediastinum and are located mainly in the adrenal glands.^[4]

Definitive diagnosis and treatment of the giant tumors of the posterior mediastinum are performed through surgical excision.^[3]

CASE REPORT

A 4-year-old girl weighing 18 kg, complaining of numbness in the feet and inability to walk for 3 months, had no known history of disease or medication use. On physical examination, the patient was in good general condition, conscious, and vitals were stable. Respiratory sounds were reduced in the upper right zone, and there was no cyanosis. A decrease in muscle tone in the lower extremities was determined, the motor tone in the upper extremities was normal, and the anal tone could not be assessed.

Posteroanterior chest radiography showed a solid homogeneous opaque mass in the upper zone of the right lung extending from the apex to the sixth intercostal space. Thoracic computed tomography revealed that the mass

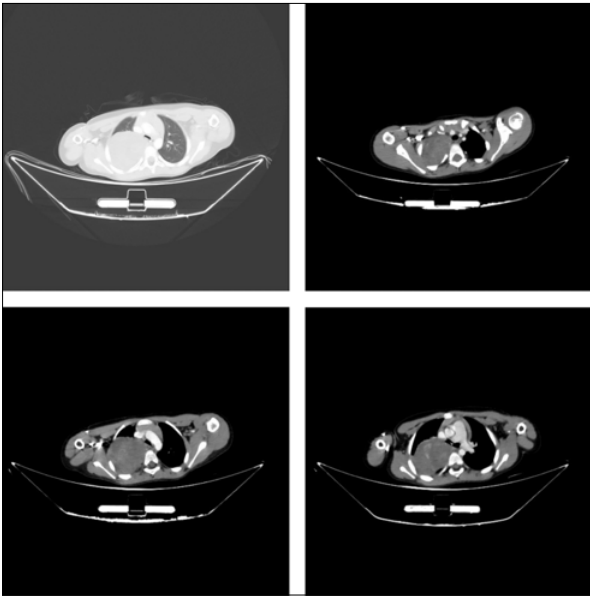


Figure 1. Sections of thorax computerized images of masses located in the posterior mediastinum.

was located in the posterior mediastinum at the C7-T6 level with an approximate size of 7×6.5 (Figure 1). No cranial pathology was observed in the magnetic resonance imaging (MRI), and it was indicated that the mass extended into the spinal canal at the C7-T4 level (Figure 2).

Transthoracic fine-needle aspiration biopsy of the mass was performed by interventional radiology, and the pathology result was reported as stromal cells.

It was decided to operate the patient. In the case coordinated with neurosurgery, C7-T4 hemilaminectomy was performed in the prone position. Then, posterior mediastinal mass excision was performed with lateral thoraco-

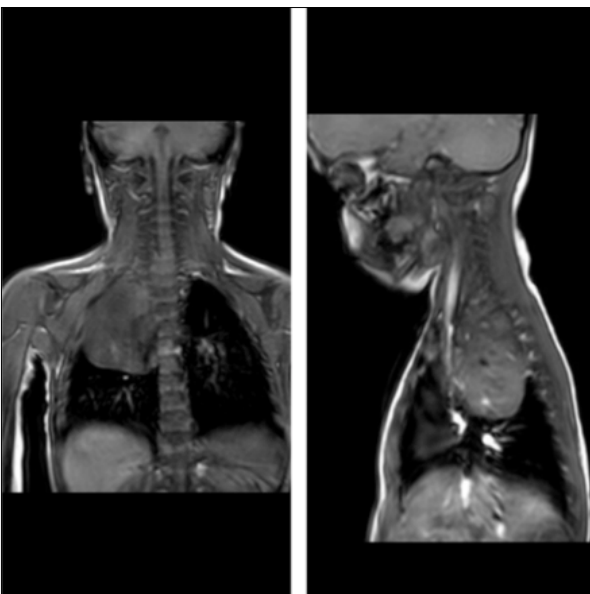


Figure 2. Magnetic resonance imaging sections of the mass located in the posterior mediastinum.

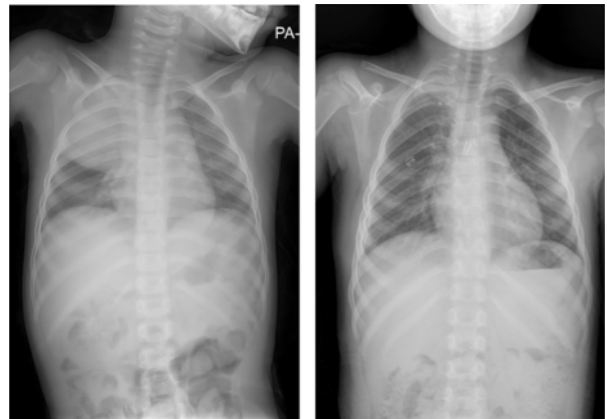


Figure 3. Preoperative and postoperative postero-anterior lung radiographs of the patient.

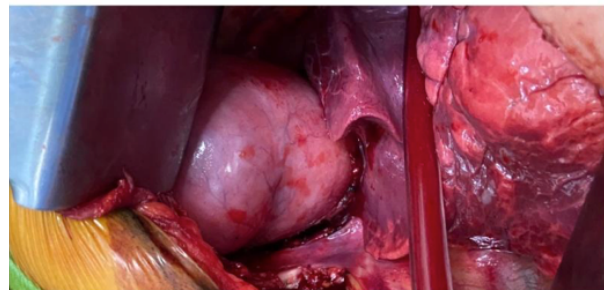
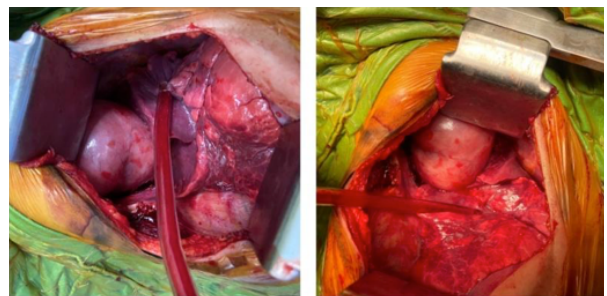


Figure 4. Peroperative photographs of the case.

tomy in the left lateral decubitus position (Figure 3).

In the final pathology, the mass size was measured as 8×5×3 cm, and ganglioneuroblastoma diagnosis was applied. CD56, neuronal specific enolase (NSE), S100, and Synaptophysin were found to be positive in the tumor cells.

The patient was discharged with recovery after 6 days in the intensive care unit and 24 days in the clinical ward in the post-operative period. At the 1st-month post-discharge follow-up visit, the patient's gait was ataxic, but motor strength was normal (Figure 4).

DISCUSSION

The mediastinum is divided into three compartments: Anterior, posterior, and middle. Neurogenic tumors constitute 80% of posterior mediastinal tumors.^[3] Ganglioneuroblastomas are found in the adrenal gland (35%), retroperitoneum (30%), posterior mediastinum (20%), and pelvis (23%) in order of frequency.^[5] Our case is a case of ganglioneuroblastoma located in the posterior mediastinum.

Ganglioneuroblastoma is mainly observed in pediatric patients aged 1 and 2 years, the mean age is 22 months, and it is usually diagnosed at the age of 10 years.^[6] Our patient was 4 years old and was diagnosed early according to the general literature.

In imaging methods, they appear as opacities on direct radiography, they are located in the posterior mediastinum in thoracic computed tomography, but the tomography findings are not specific enough.^[5] Neuroblastomas and ganglioneuroblastomas contain coarse calcification, while gangliogliomas contain 20% fine punctate calcification.^[7] MRI shows high signal intensity on T1-weighted imaging and low signal intensity on T2-weighted imaging and evaluates invasion better.^[5] In the computed tomography scans, our case did not show any calcification. In the MRI, extension into the spinal canal was observed.

Our patient underwent pre-operative transthoracic fine needle aspiration biopsy but could not be diagnosed. Histopathologic diagnosis is not mandatory for a surgical decision.^[3] Chemotherapy and radiotherapy are among the treatment options in cases where surgical treatment is not possible.^[5]

Diagnostic markers of ganglioneuroblastomas include catecholamines, valinmandalic acid, and homovalenic acid.^[8] Biochemistry markers were not studied in our patient because the operation was scheduled in the early period.

In a reported case, a 4-year-old pediatric patient underwent an operation due to a mass located in the posterior mediastinum causing a cervical intradural extension, and the patient regained upper and lower extremity strength after the operation.^[9] In another case report of ganglioneuroblastoma in an 8-month-old child, the patient regained muscle strength in the feet with laminectomy and excision.^[10] In our case, lower extremity motor power was fully restored with successful surgery.

The final pathology of our case was reported as nodular-type ganglioneuroblastoma (Schwannian stroma-rich/dominant and Schwannian stroma-poor). The International Neuroblastoma Pathology Classification based on the Shimada system is used in the classification of neuroblastomas.^[11] In neuroblastomas, synaptophysin, chromogranin, CD56, NSE, and protein gene product 9.5 (PGP9.5) are stained positive, although not specific for diagnosis.^[11] In our case, CD56, chromogranin, NSE, S100, synaptophysin are stained positive.

Conclusion

Our case report is presented to draw attention to rare pediatric neurological tumors. We believe that successful surgery in mediastinal neurological tumors of pediatric patients performed by experienced centers would yield satisfactory results.

Peer-review

Externally peer-reviewed.

Authorship Contributions

Concept: A.Ö., S.K., R.D.; Design: A.Ö., S.K., R.D.; Supervision: A.Ö., S.K., R.D.; Fundings: A.Ö., S.K., R.D.; Materials: A.Ö., S.K., R.D.; Data: A.Ö., S.K., R.D.; Analysis: A.Ö., S.K., R.D.; Literature search: S.K., A.Ö., R.D.; Writing: S.K., A.Ö., R.D.; Critical revision: S.K., A.Ö., R.D.

Conflict of Interest

None declared.

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Posterior Mediasteninin Nadir Görülen Çocukluk Çağı Tümörü: Ganglionöroblastoma

Ganglionöroblastoma, posterior mediastende yerleşimli çocukluk döneminin nadir görülen malign bir tümördür. Yürüme güçlüğü ile hastanemize başvuran 4 yaşındaki kız çocuğunda posterior mediasten yerleşimli spinal kanal içine uzanan dev kitle lezyon izlendi. Beyin cerrahisi ve göğüs cerrahisini ortak operasyonu ile kitle total eksize edildi. Ameliyat sonrası hasta yürüme fonksiyonlarını tekrar kazandı.

Anahtar Sözcükler: Çocukluk çağı tümörü; ganglionöroblastoma; posterior mediasten tümörü.