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Case Report

Thoracic Ganglioneuroma in a Seven-Year-Old Boy: A Case Report

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Abstract

Introduction: Thoracic ganglioneuroma is a rare benign tumor arising from the ganglia of the sympathetic nervous system. Most present asymptomatically as an incidental radiographic finding, but some cases present with symptoms.

Case Presentation: A 7-year-old boy presented with dyspnea. Computed tomography and magnetic resonance imaging studies showed a large lesion in the posterior mediastinum extending to the spinal canal at the T3 and T4 levels. An image-guided biopsy confirmed the diagnosis of ganglioneuroma. A two-stage surgical tumor resection performed. The first stage approached the tumor posteriorly and freed the mass from the spinal cord; the second stage debulked the mass anteriorly through a lateral thoracotomy. The patient has improved after surgery in terms of his difficulty in breathing and back to his normal activity. The two-stage surgical resection had no complications. The patient has been followed up clinically and radiologically.

Conclusion: A multidisciplinary approach is highly recommended in large-size tumors. Although complete resection has a favorable outcome in most cases, it may be difficult to achieve this in case of large tumors that are adjacent to vital structures.

Keywords: *Ganglioneuroma; Spinal Tumor; Spinal Cord Tumor; Nerve Sheath Tumor; Thoracic Tumor*

Introduction

Ganglioneuromas (GNs) are rare benign tumors that usually arise from the ganglia of the sympathetic nervous system [1,2]. They are most commonly found in the peripheral nervous system [3]. The thoracic GNs are commonly located in the posterior mediastinum, retroperitoneum, and adrenal gland, but rarely in the neck [1,4]. Although GNs typically involve the paraspinal region and are associated with intraspinal extension, they can invade the spinal canal [1,3].

Most are asymptomatic and found incidentally on radiographic imaging studies, however some present with chest discomfort [5], back pain [1], scoliosis [6,7], neurological symptoms [8,9]. We present our approach in a seven-year-old boy with a thoracic GN who presented with dyspnea on exertion.

Case Report

Presentation

A seven-year-old boy presented with a 3-month history of exertional dyspnea. His review of systems was unremarkable and without constitutional symptoms. There was no family history of chest disease. On physical examination, the patient had a normal facial appearance without dysmorphic features. Auscultation of the chest showed decreased breath sounds over the left lung. He was neurologically intact with normal deep tendon reflexes in the upper and lower extremities and normal gait.

Investigation

A chest radiograph was obtained and showed a large well-defined soft tissue opacity in the left hemithorax measuring about 14 x 8.9 cm (Figure 1).

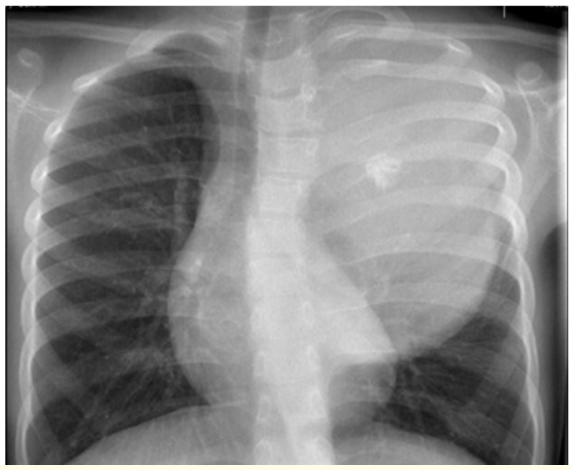

Figure 1: *Plain chest x-ray with left thoracic mass.*

Subsequent chest and thoracic spine CT showed a large left extrapulmonary mass with central calcification causing mass effect upon the mediastinal structures and heart, which were mildly displaced to the right, and complete atelectasis of the left upper lobe (Figure 2). The upper border of the mass extended to the lower aspect of the left neck, while the medial anterior border crossed the midline in the mediastinum. The large mass extended into the left T3 and T4 neural foramina shifting the spinal cord to the right.

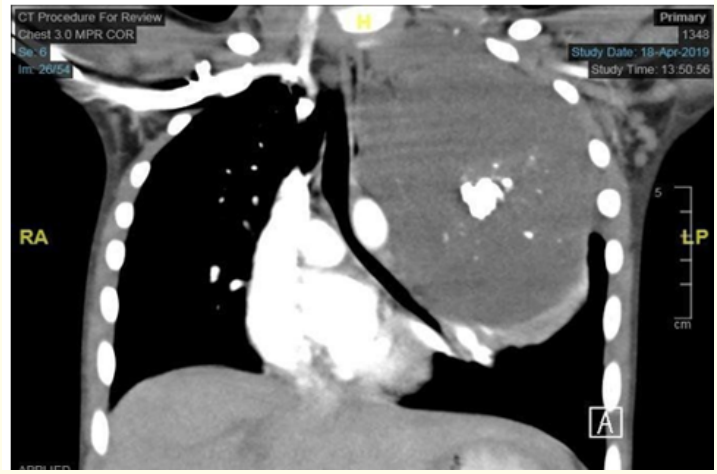

Figure 2: *Coronal chest CT showing central calcification and mass effect on the mediastinum.*

The patient then underwent total spine MRI with contrast which showed a large well-delineated and slightly lobular left posterior mediastinal mass measuring 137.5 × 62.5 × 79.5 mm in the chest cavity that extended into the left T3 and T4 neural foramina and caused posterior T3 scalloping. The mass was isointense to muscle on T1-weighted images and had heterogeneous signal intensity on T2-weighted images. The spinal cord was displaced toward the right without evidence of signal abnormality or distortion in shape (Figure 3 and 4).

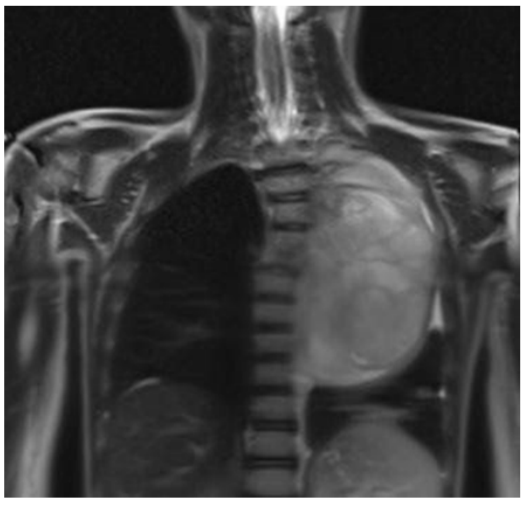

Figure 3: *Coronal MRI of the thoracic mass.*

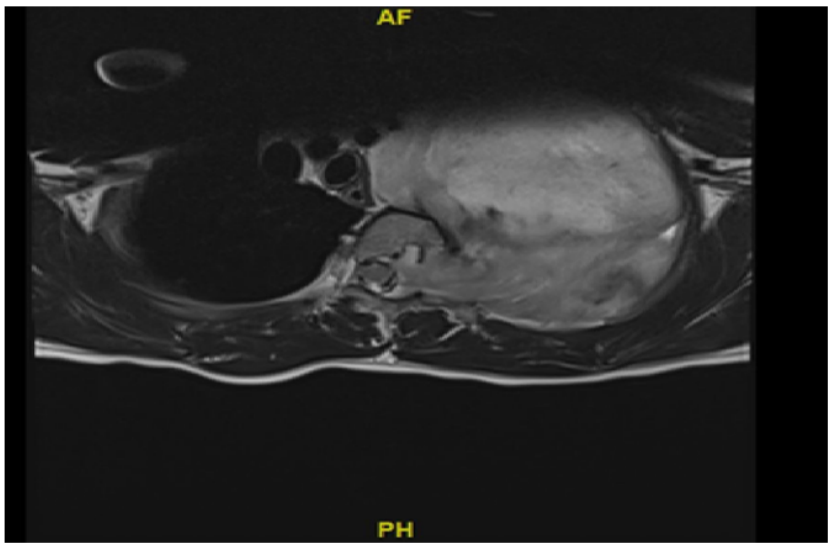

Figure 4: *Axial MRI showing left T3 foraminal extension of the mass.*

An image-guided biopsy of the left mediastinal mass was performed. Histopathologic examination of the specimen showed Schwannian stroma with numerous mature ganglion cells, consistent with GN.

Management

The patient was managed using a multidisciplinary approach that included pediatric surgeons, spine surgeons and oncologists. Chemotherapy and radiation therapy were deferred at this stage given the biopsy results. Surgical resection was planned, involving both the pediatric and spine surgery teams. The first stage was performed by the spine surgeons under general anesthesia using intraoperative motor and somatosensory evoked potential monitoring. A posterior thoracic spine approach with the patient in the prone position was used. T2–T4 laminectomies were performed under the microscope for tumor visualization. The mass within the spinal canal extended caudally to T4 and was approximately 5 × 4 cm in dimension. It appeared bulky with clear margins and was adherent to the spinal cord and the left T3 nerve root. The nerve root was ligated and resected to avoid traction or tension on the spinal cord during the second stage of tumor resection (Figure 5).

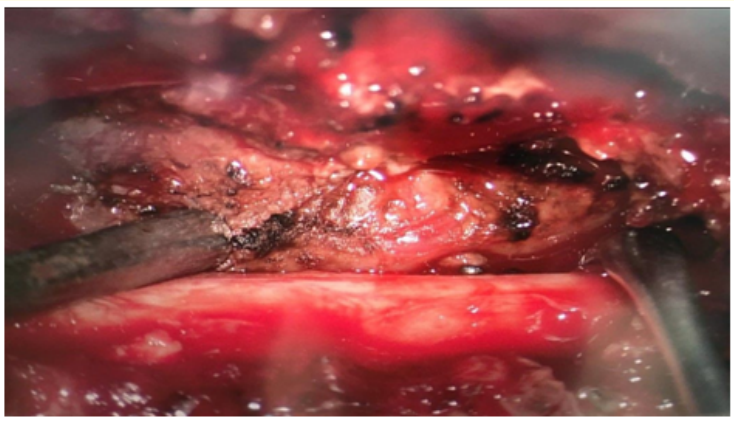

Figure 5: *Intraoperative photograph showing the bulky tumor and ligated left T3 nerve root.*

The mass was carefully dissected from the spinal cord and remaining nerve roots then debulked using the Cavitron Ultrasonic Surgical Aspirator (CUSA) and bipolar electrocautery. Because the anterior and middle columns of the spine appeared stable, we decided to defer spinal fixation and stabilization and closely monitor the patient postoperatively for the development of spinal deformity.

The pediatric surgical team performed the second stage of resection. With the patient in the right lateral decubitus position, a left lateral thoracotomy was performed to access the tumor through an incision in the left fifth intercostal space. No fluid or adhesions were found between the mass and the pleura anterolaterally, but posterior adhesions between the mass, ribs, and the vertebrae were present and dissected. Due to the small working space, the mass was resected in piecemeal slices until the thoracic inlet was reached. The total weight of the resected mass was approximately 460 grams. At the end of the procedure, a 24 French chest tube was inserted and connected to an underwater seal. The patient was then transferred to the pediatric intensive care unit intubated with stable vital signs.

Two tumor specimens were sent for histopathologic examination, one from the T3 mass within the spinal canal and one from the mass in the left thorax. The T3 mass consisted of multiple fragments of gray-tan, firm tissue measuring 1 × 1 × 0.3 cm in aggregate. The thoracic mass was comprised of multiple fragments of white-tan, rubbery firm tissue measuring 14 × 13 × 5 cm in aggregate with a homogenous, smooth, and shiny outer surface. Microscopically, both samples showed a Schwannian stroma-rich tumor mixed with mature ganglion

cells (Figure 6). No immature neuroblastic component was seen. Immunohistochemistry showed that the Schwannian stromal cells were positive for S100 and GFAP (Figure 7). The ganglion cells were positive for chromogranin, synaptophysin, and neuron-specific enolase (Figure 8). The Ki-67 proliferative index was approximately 1%. Based on the above findings, the final diagnosis was GN.

The patient was extubated two days after surgery. Neurological examination at this time showed no motor or sensory deficit. On the fourth day, he was transferred to the general ward and began to mobilize. He was then discharged home in stable condition within a week. Since then, the patient has followed up regularly for one year and no longer complains of exertional dyspnea. CT and MRI have shown no sign of tumor recurrence.

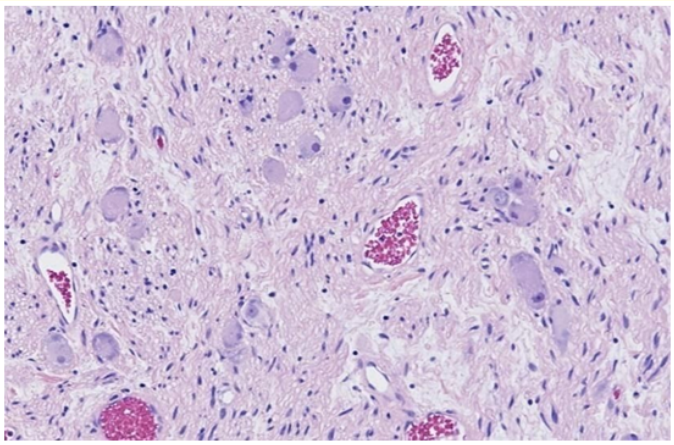

Figure 6: *(20X) Hematoxylin and eosin (H&E) stained section showing scattered mature ganglion cells with intervening Schwannian stroma.*

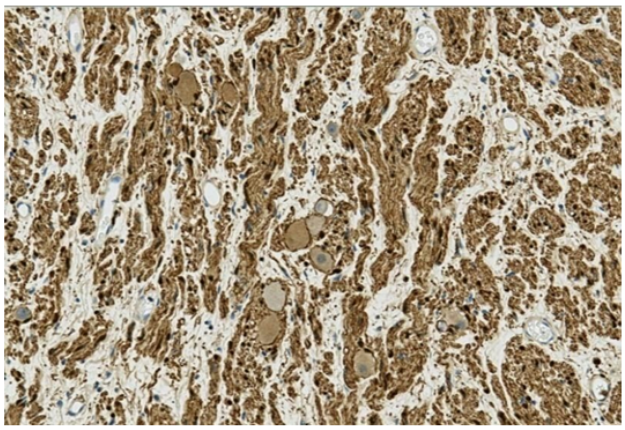

Figure 7: *Ganglion cells and Schwann cells are both positive for S100.*

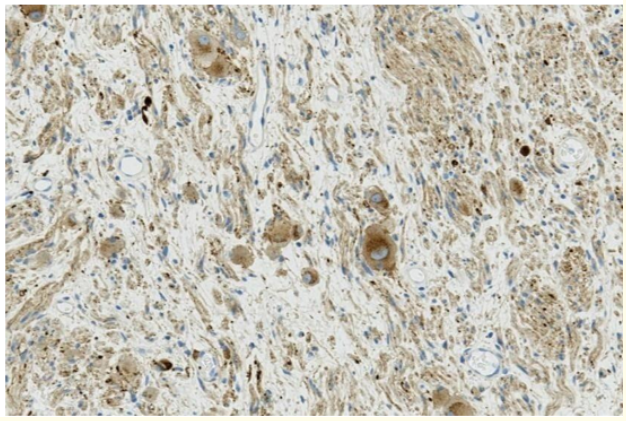

Figure 8: *Ganglion cells and Schwann cells are both positive for synaptophysin.*

Discussion

GNs are one of the few tumors that arise from neural crest cells and may occur at any peripheral autonomic ganglion site [10]. They can appear as a posterior mediastinal mass, which has a broad range of differential diagnoses; ganglioneuroma is only one of many to be considered, including malignant peripheral nerve sheath tumor, schwannoma, neurofibroma, ganglioneuroblastoma, neuroblastoma, primitive neuroectodermal tumor, lymphoma, pheochromocytoma, paraganglioma, fibroma, fibrosarcoma, liposarcoma [10-13].

Based on our literature review, only twelve cases of extradural thoracic GNs have been previously reported. With their benign background, GNs are capable of impressive growth [5]. This thoracic GN is large in size; it occupied more volume percent of the thorax since it occurred in a pediatric patient. Although most previous cases were found incidentally, they can present with radicular pain, chest discomfort due to tumor mass compression, or hydrops fetalis in neonates [5,14].

Radiographic and histopathologic findings play an important role in diagnosis. In this case, the patient's symptoms along with a plain chest radiograph that showed a large, well-defined soft tissue opacity led to further investigation with CT and MRI, which provided more detailed information. The spinal cord appeared displaced toward the right side with no evidence of shape or signal abnormality. These findings of spinal canal involvement are rare, and they are among 10% of the cases of GN [3,8]. GNs characteristically appear hypodense and hypointense on plain CT and MRI, respectively. Contrast-enhanced CT and MRI studies show no or slight enhancement in the arterial phase and progressive mild enhancement in the delayed phase [15]. It also showed coarse calcification that is generally present in approximately in 20% of GN cases [16].

The diagnosis is made histologically based on the observation of mature ganglion cells within the tumor [15]. Although most of the previous thoracic GNs reports did not describe histopathology findings in detail, our findings were in agreement with those that did: a Schwannian stroma-rich tumor admixed with mature ganglion cells; stromal cells positive for S100 and GFAP; ganglion cells positive for chromogranin, synaptophysin, and neuron-specific enolase; and low proliferative index. These together with the radiographic findings were our cornerstone in establishing the diagnosis.

Surgical excision is the definitive treatment of GNs [15]. In our case, the mass occupied a portion of the spinal canal and two-thirds of the left hemithorax adjacent to the aorta and major vessels. A multidisciplinary approach and two-stage resection were key. The first stage was directed at the posterior intraspinal tumor. We first freed the spinal nerve root from the tumor and debulked the tumor inside

the capsule under neurophysiologic monitoring. We sacrificed the left nerve root because the tumor had extended to the canal, which was different from previously reported tumors that remained extraforaminal. No instrumentation was needed in most of the previously reported cases, as in our case, except for those patients with associated scoliosis or those who underwent single-stage posterior resection alone that resulted in iatrogenic spinal destabilization.

The second stage of tumor resection involved thoracotomy for resection of the anterior intrathoracic tumor. After access to the tumor was achieved, it was debulked as much as possible due to its attachment to vital neck structures. Postoperatively, pulmonary edema resulting from re-expansion of a chronically compressed lung may occur and close monitoring is crucial [17,18]. In our patient, follow-up CT and MRI two weeks postoperatively showed good lung expansion and a significant reduction in the size of the mass (Figure 9 and 10). He had no oxygen requirement or respiratory complaints and remained neurologically intact.

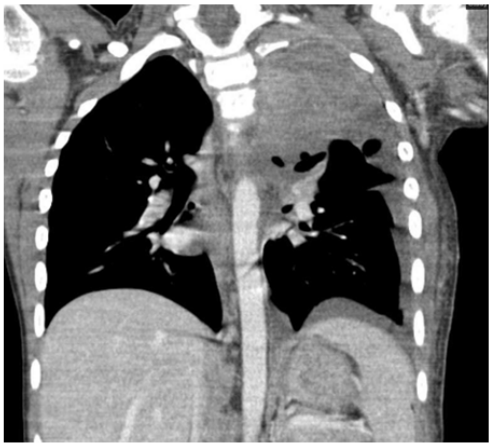

Figure 9: *Coronal CT after surgery.*

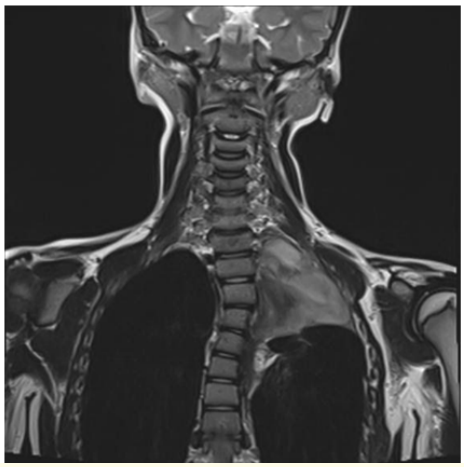

Figure 10: *Coronal MRI 5 months after surgery.*

Conclusion

GNs are benign tumors with a low recurrence rate. A multidisciplinary approach is highly recommended for large size tumors. Although complete resection has a favorable outcome in most cases, it may be difficult to achieve with large tumors adjacent to vital structures. Reporting such a case helps to provide more data regarding the disease and establishes a guide for future diagnosis and management.

Conflicts of Interest

None of the authors have any conflict of interest.

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