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Thoracic Ganglioneuroma: A Rare Neural Tumor (Case Report and Literature Review)

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Abstract

Ganglioneuroma is an extremely rare tumor that is derived from neural crest. Many ganglioneuroma cases are detected incidentally unless they are large enough to cause compressive symptoms. We report an 18-year-old patient with posterior mediastinal ganglioneuroma which was abutting the descending aorta. The patient underwent successful resection by thoracoscopic approach and was followed up for one year with no complications. In summary, a detailed review with experts in both radiology and pathology is mandated to diagnose these tumors. Informed consent was obtained from the patient.

Keywords

Neural Cell Tumors, Ganglioneuroma, Mediastinal Mass

1. Introduction

Neural tumors in the chest cavity are generally rare. However, they represent about 75% of posterior mediastinal tumors [1]. Differential diagnoses for posterior mediastinal masses are vast and can be categorized into neural lesions such as nerve sheath tumors (schwannomas), neural crest tumors (ganglioneuroma), parasympathetic ganglion (paraganglioma), or non-neurogenic lesions such as chondroma, sarcoma, lymphoma, Ewing sarcoma, and invasive thymoma. Ganglioneuroma is one of these neural tumors which is considered rare with few cases reported in the literature [2]. They are benign tumors derived from peripheral nerve sheaths [3]. Many ganglioneuroma cases are detected incidentally unless they are large enough to cause compressive symptoms. They appear as solid, well-defined lesions with a characteristic appearance on CT (Computed

Tomography) scans that require a specialized radiologist to distinguish them from other lesions and their differential diagnosis includes neurilemmoma (Schwannoma), neurofibroma, paraganglioma, and neuroblastoma [2]. In this paper, we present a case of posterior mediastinal mass which was revealed to be a ganglioneuroma.

2. Case Presentation

An eighteen-year-old gentleman who is not known to have any medical issues was referred to our clinic after an incidental finding of a posterior mediastinal lesion on a chest CT that was performed in an outside hospital as a routine new job investigation. Initially, there was a suspicious shadow of a mediastinal mass on a regular X-ray. The patient was completely asymptomatic. Physical examination and laboratory workup were unremarkable. Based on the CT chest, the mass was in the left posterior mediastinal paraspinal region and measured approximately 2 x 3.2 x 6 cm in transverse, AP (anteroposterior), and craniocaudal dimensions respectively (**Figure 1**). Further imaging was requested to delineate the relationship to surrounding tissues. A chest MRI (Magnetic Resonance Imaging) revealed the mass with specific measurements as (1.7 x 3.2 x 6 cm) paralleling the third thoracic vertebrae and extending to the sixth thoracic vertebrae. In addition, it was longer in the vertical direction, and there was a 2 cm horizontal extension below the fourth rib, at the level of the aortic arch abutting the descending aorta, with no aortic or neural foramina invasion. As per the MRI, the mass showed a low signal intensity in T1 and heterogenous high signal intensity in T2 weighted images, with evidence of delayed enhancement on post-contrast images (**Figure 2**). Workup continued with Pulmonary Angiography CT that confirmed the patency of the pulmonary trunk, right and left pulmonary arteries, along with lobar and segmental branches (**Figure 3**).

The decision was made to go for resection. A left thoracoscopic approach was made under general anesthesia, and a left-sided double-lumen tube was used for intubation. The patient was positioned in right lateral decubitus. Using three small ports; the camera port in the eighth intercostal space at the posterior axillary line, and two other ports; one anterior to the level of the fourth intercostal space and another posterior at the sixth intercostal space. Initially, we were faced with some difficulties in identifying the origin of the lesion, but after meticulous attempts at dissection using both sharp (hook) and blunt dissection, we were able to resect the lesion completely (**Figure 4**, **Figure 5**). The rest of the operation was unremarkable and continued smoothly. The patient was extubated immediately after the operation and shifted to the recovery unit in a stable condition, with an immediate chest X-ray obtained (**Figure 6**). Postoperatively, the patient was clinically doing well and improving. The chest tube was removed on the third postoperative day and the patient went home on the fifth postoperative day in excellent condition (**Figure 7**). The patient was seen regularly in the clinic and was doing well up to one year after surgery (by the time of publishing this paper). We will continue to follow the patient in the clinic in case of any late complications.

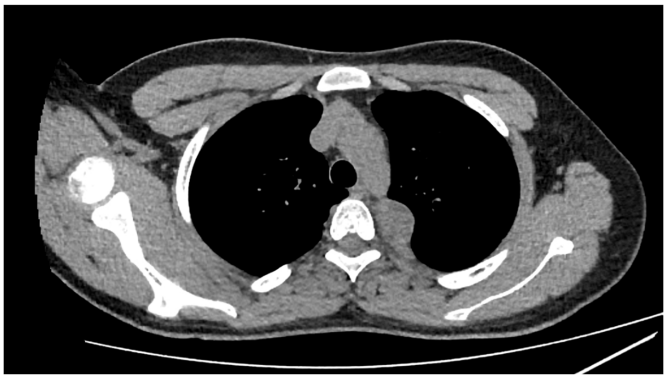

Figure 1. CT chest showing a left posterior mediastinal paraspinal soft tissue mass.

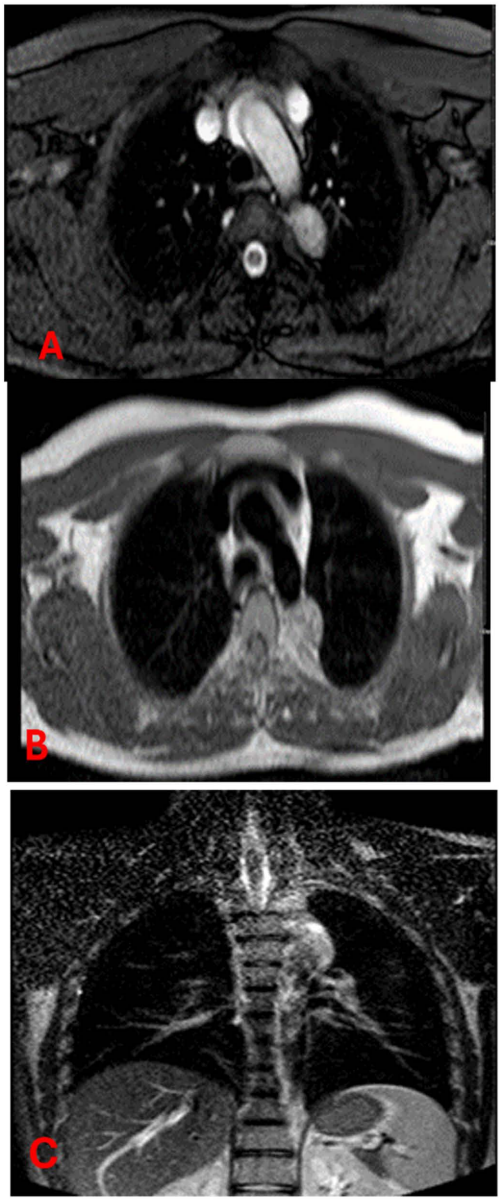

Figure 2. Chest MRI showing left posterior paraspinal soft tissue mass (A = T2 image; B = T1 Image; C = coronal cut view).

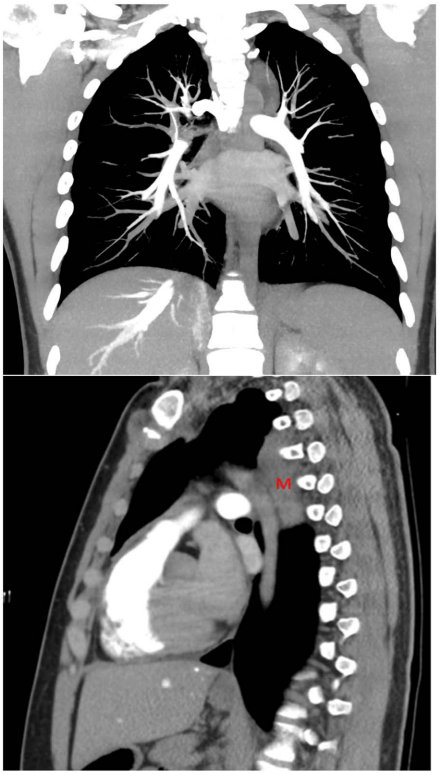

Figure 3. Pulmonary Angiography CT (M = Mass).

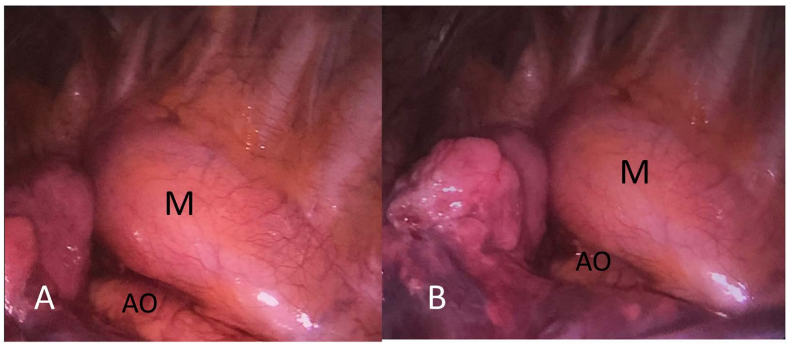

Figure 4. Operative findings (M = soft tissue mass, AO = Aorta).

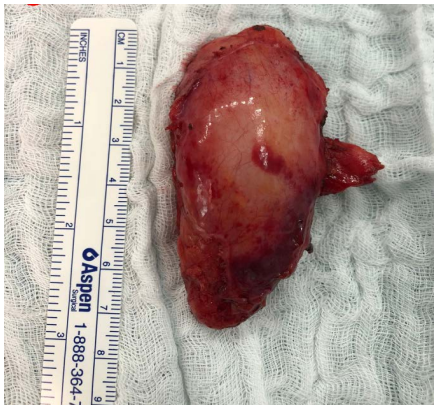

Figure 5. Gross appearance of the mass post-resection.

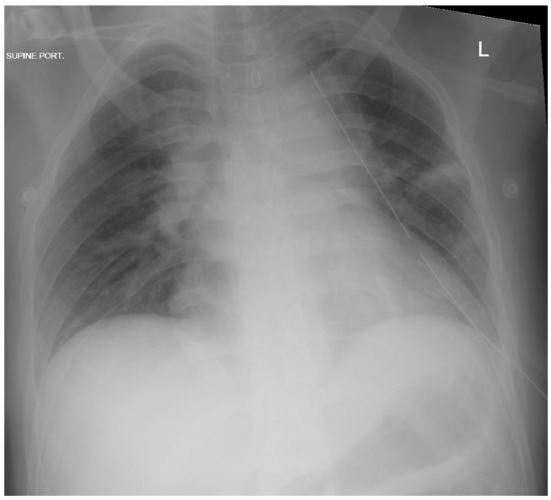

Figure 6. Post operative chest x ray.

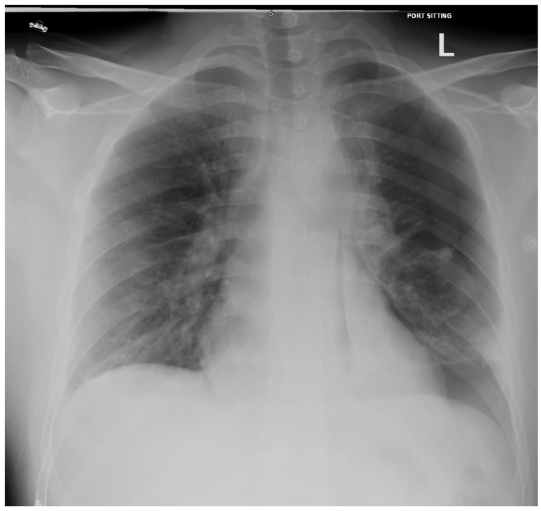

Figure 7. Chest x ray after chest tube removal on day 3.

Pathology review showed a posterior mediastinal mass that was identified as a mature ganglioneuroma, measuring 7.5 x 3.5 x 2.5 cm on gross description. The specimen consists of a tan-brown, ovoid firm, and well-defined homogenous mass. Microscopically, the immunohistochemical stains highlight the Schwann cells with S100. Also, mature ganglionic cells were interspersed throughout the tumor. Immunohistochemical stains for desmin, smooth muscle actin, and calretinin were all negative.

3. Discussion

The diagnosis of ganglioneuroma is not always easy due to its rarity. It is a benign lesion that originates from neural crest cells in the sympathetic ganglia or adrenal medulla. It is often discovered incidentally; it is usually detected in healthy and younger ages, but some reported cases are of patients 80 years old [3]. The size of the lesion varies from small asymptomatic lesions to large symp-

tomatic ones reaching up to 23 x 10 x 10 cm [2]. Symptoms mainly will be related to adjacent organ compression including but not limited to pain, shortness of breath, cough, hemoptysis, and post-obstructive pneumonia. In this case, the lesion was found in the posterior mediastinum however ganglioneuromas can be found in other sites such as the retroperitoneum, adrenal glands, intracranial, and ureter [4]. There are three cases reported in the literature where ganglioneuroma was detected in endobronchial locations [3]. These endobronchial lesions often require sampling to differentiate them from other lesions such as adenoid cystic, endobronchial lipoma, and hamartoma. Imaging plays a very important role in the diagnosis of these lesions and the planning for their management. Ganglioneuromas appear as hypodense and homogeneous round or elliptical lesions on a CT scan with a lower density when compared to surrounding muscles. On MRI, they have a low signal on T1-weighted images and a high signal on T2-weighted images with an occasional whorl appearance, and enhancement of the edge due to the capsule [5] [6]. Malignancy should be ruled out when atypical findings are detected on radiology. It is rare for ganglioneuroma to erode into adjacent bony structures such as ribs and vertebral bodies however it was reported in the literature, which reflects that in certain circumstances there will be an invasive characteristic [7] [8]. To differentiate ganglioneuroma from other mediastinal diseases such as lymphoma, thymoma, neurofibroma, and others it is necessary in certain cases to obtain pathological samples through needle biopsies which are 90% sensitive, keeping in mind the possible complications after such procedures like bleeding, pneumothorax, seeding of the disease through the biopsy site or pleura and others [2] [7].

Histologically they are described as a nerve fiber with a wave pattern under the microscope with mature ganglion cells. Positive staining with Vim, S-100, neuron filament protein, myelin basic protein, and neuron-specific enolase shows the nerve origin of the tissue [9]. Ganglioneuroma has been reported to be associated with rare paraneoplastic syndromes. Dr. Tiwari *et al*. report a case of thoracic ganglioneuroma associated with Rapid-onset Obesity with Hypoventilation, Hypothalamic dysfunction, and Autonomic Dysregulation Syndrome (ROHHAD), recently called ROHHAD-NET (ROHHAD-Neuro Endocrine Tumors) associated with well-defined left paravertebral enhancing solid mass lesion extending from the second to fifth thoracic vertebra seen on MRI, that turned out to be a ganglioneuroma after a successful thoracoscopic excision. Further, it was described as well-circumscribed, smooth, gray-white, firm in consistency, and had a whorled appearance [10]. Although when we talk about ganglioneuroma we are speaking about a benign entity it's worth mentioning that some reports are mentioning the presence of malignancy. Pi-Yu Chen reported a case of a 34-year-old man with an anterior mediastinal tumor which was labeled as a mixed germ cell tumor based on pathology. In association with elevated serum α-fetoprotein (AFP) and β-human chorionic gonadotropin (β-hCG). The patient received neoadjuvant chemotherapy followed by surgical removal. A final pathologic examination of the excised specimen showed pre-

dominantly malignant ganglioneuroma and small residual foci of teratoma. That was the first reported case of a malignant ganglioneuroma from a mediastinal germ cell tumor [11]. Many surgical approaches are used to resect ganglioneuromas. However, the majority can be removed by minimally invasive techniques like in our case. A paper written by Okui *et al*. mentioned the use of modified Trapdoor thoracotomy to resect a cervicomediastinal ganglioneuroma [12].

4. Conclusion

In conclusion, rare neural chest tumors have been reported in the literature. Ganglioneuromas are rare benign peripheral nerve tumors which are presented in this case report. In this paper, an 18-year-old medically free, asymptomatic patient discovers an incidental posterior mediastinal mass during a routine checkup. He underwent an uneventful left thoracoscopic resection of the mass. Patient was followed in clinic at regular intervals with no reported complications up to one year post-surgery. We concluded that challenges in diagnosis are attributed to its rare entity, and symptoms vary according to size difference. Despite its rarity, neural cell tumors must be considered as one of the differentials in cases of posterior mediastinal mass. A detailed review of both radiology and pathology with experts in the fields is mandatory to classify and manage these tumors.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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