Case report - Thoracic oncologic

Giant ganglioneuroma of the posterior mediastinum

Javed Hayat*, Riaz Ahmed, Shereen Alizai, Muhammad Umer Awan
Division of Cardiothoracic Surgery, Shifa International Hospital, Sector H 8/4, Islamabad, Pakistan
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Abstract

We report the case of a 45-year-old female referred to us with progressive shortness of breath and a huge left mediastinal mass. Computed tomography of the chest revealed a mass occupying the posterior mediastinum and extending from the apex caudally to the left hemidiaphragm. Further magnetic resonance imaging demonstrated tumor extension to the left intervertebral foramina of T5–T6 and T6–T7. Excision of the mass was performed through a left posterolateral thoracotomy. Histology confirmed a mediastinal ganglioneuroma. This is an unusual tumor with more than one extension in the spine. Ganglioneuromas are rare tumors of the peripheral nervous system. Most of these tumors are, however, retroperitoneal and are more common in children and young adults. Ganglioneuromas arise from neural crest cells. These tumors are mostly asymptomatic, but some may present with hypertension and flushing. Massive tumors can present with pressure symptoms.

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1. Case report

A 45-year-old female had been visiting her general practitioner for the previous 10 years with hypertension. However, over a period of four months, she developed progressive shortness of breath. She also had a single episode of hemoptysis. She had no loss of weight, and her appetite was good. Her clinical examination was unremarkable except for reduced air entry to her left mid- and lower chest. This prompted her general practitioner to do a chest X-ray, which showed an opaque left hemithorax. It also showed splaying of the left ribs, with scoliosis. Laboratory test results were within normal limits.

Computed tomography of the chest depicted a huge mass occupying the left posterior mediastinum and extending from the apex to the hemidiaphragm, with a shift of the midline. The intervertebral foramen between T5 and T6 was widened, with the mass extending into it. Because of spinal involvement, magnetic resonance imaging of the spine was performed, which confirmed a mass with an epicenter at the left intervertebral foramina of T5–T6 and T6–T7 (Fig. 1). Since this tumor had more than one extension in the spine, suspicion of malignancy was raised. Ultrasound-guided fine-needle aspiration cytology was performed. Cytology of the specimen revealed benign mesenchymal cells composed of spindle-shaped Schwann cells and collagenous bundles with no evidence of malignancy.

The patient was taken to the operating room, and a left posterolateral thoracotomy was performed. A huge mass was found to be occupying almost the whole of the left hemithorax. It was covered with mediastinal pleura and adherent to the chest wall, pulmonary hilum, left lung and descending aorta. However, the mass was not invading any of these structures. It had rich blood supply from the mediastinum. Careful dissection of all the adhesions and division of all the feeding vessels led to two pedicles extending into the thoracic spine. Traction on the tumor was kept to a minimum to prevent neurological complications. Preparation

*Corresponding author. Tel.: +92-51-4603666; fax: +92-51-4863182. E-mail address: javedhayat2002@yahoo.co.uk (J. Hayat).

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was made for neurosurgical resection of these pedicles. A neurosurgeon assessed the tumor, and since both the pedicles were extradural, it was decided not to proceed with laminectomy in the prone position. These pedicles were carefully dissected and excised from the thoracic spine with the help of the neurosurgeon. Hence, complete en bloc resection was achieved. A postoperative chest X-ray revealed normal lung expansion. The postoperative recovery was uneventful, with no neurological deficit, and the patient was discharged home on day 4.

Gross examination revealed a mass measuring 21 cm × 14 cm × 9 cm and weighing 1320 g. It also had two pedicles measuring 4.5 cm × 2.8 cm and 4.4 cm × 2.6 cm, respectively (Fig. 2). On thin sectioning, the surface was found to be a homogeneous grayish-tan color. Histologically, it showed a non-encapsulated spindle cell lesion. The spindle cell's contained markedly elongated nuclei with wavy serpentine configuration and pointed ends. These cells were arranged in fascicles, nerve bundles, ganglion cell neurons and fibroblasts on a fibromyxoid background. These findings led to the final diagnosis of mediastinal ganglioneuroma.

The patient is alive with no signs of recurrence two years after her surgery.

2. Discussion

Ganglioneuromas are tumors arising from the neural crest cells. These tumors belong to a group that also includes peripheral nerve sheath tumors, melanomas and neuroendocrine tumors [1]. Ganglioneuromas by and large either occur in the posterior mediastinum or are more commonly retroperitoneal. About 40% of these tumors occur in the posterior mediastinum [2]. Mediastinal neurogenic tumors are more common in infants and children, and are mostly malignant [3]. Ribet and Cardot reported 84.8% of nerve cell tumors to be in infants and children, of which 67.2% were malignant [3]. Although nerve cell tumors were more common in the pediatric population according to this report, the majority (73.5%) of nerve sheath tumors were found in adults. Ganglioneuromas have also been reported to occur rarely in other locations, such as the tongue, bladder, uterus, bone and skin [4].

Ganglioneuromas can remain asymptomatic for a number of years, to be discovered incidentally on a chest radiograph. Big tumors can cause pressure symptoms and will, if they produce catecholamines, present with hypertension and flushing [5]. Shimada et al. published a histological classification of neuroblastic tumors in a large series of cases [6]. According to this classification, ganglioneuromas should have a schwannian stroma-dominant maturing histology.

There is no medical treatment for these tumors. Mediastinal ganglioneuromas, although benign, can grow aggressively and invade mediastinal structures and the spine. Patients treated surgically for a benign neurogenic tumor have an excellent prognosis [7]. A multidisciplinary approach involving a neurosurgeon is important before embarking upon surgery. Our patient had long-standing hypertension, but her pressure symptoms prompted further investigations. Following excision of the tumor, her antihypertensive medication requirements were reduced.

The ganglioneuroma in our patient was unusual that it was massive in size with more than one tumor pedicle attached to the vertebral bodies. To our knowledge, this has not previously been reported and hence, raised a suspicion of malignancy. A thorough preoperative assessment is mandatory to help plan surgery accordingly. If cytology reveals both spindle and ganglion cells, it is diagnostic of ganglioneuroma [8]. In conclusion, surgical excision of these tumors is safe and curable.

References