



**A CASE REPORT: POSTERIOR MEDIASTINAL GIANT VERTEBRAL CANAL-
ASSOCIATED GANGLIONEUROMA**

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ABSTRACT

This is the case study about Ganglioneuroma in a 23-year-old Chinese female. Ganglioneuroma is a rare benign neurogenic tumor originating from the sympathetic nervous system and occurring in youth and adulthood. Computer tomography plain scan showed soft tissue mass on the right side of posterior mediastinum and in front of the spine.

INTRODUCTION

Case study: The patient 23-year old Chinese female. 1 years ago, there was no obvious reason for recurrent paroxysmal cough with yellow sputum, no hyperthermia, no low fever, night sweats, and no fatigue. Before February, the patient coughed and sputum increased and sputum was bloodstained. Color Doppler echocardiography showed congenital heart disease (atrial septal defect type II). Preoperative mediastinum occupying was found after physical examination. CT plain scan showed soft tissue mass on the right side of posterior mediastinum and in front of the spine. The spindle shape with chest 9 as the center was about 19 cm in upper and lower diameter, and the maximum transverse diameter was about 10 cm *6 cm. The mass extends downward to the retroperitoneum. A small amount of spotted calcification was seen in the mass, and multiple vertebral bodies and ribs showed erosive bone destruction, and multiple intervertebral foramens were enlarged. The mass was slightly enhanced in three-phase enhanced CT scan, and delayed enhancement was slightly more obvious than arterial and venous phase (CT value increased by about 9 Hu on average). MRI showed an equal sign on the T1WI on the mass and uneven and slightly hyperintense on the T2WI and T2WI lipid sequences. The lump is intruded into the spinal canal through multiple intervertebral foramens, and the local spinal cord is compressed and displaced. The lumen moved forward to move the inferior vena cava forward. MR enhanced scan showed a slightly heterogeneous plaque enhancement.

Transthoracic biopsy pathology: nerve fiber hyperplasia, structural disorders, rich blood vessels, ganglion cells were found in some areas. Diagnosis: posterior mediastinal spinal ganglion cell neuroma.



Figure 1: CT reconstruction of the right side shows a spindle like soft tissue mass on the right side of the spine, with a small amount of spotted calcification.



Figure 2: Coronal T1WI showing a heterogeneous signal on the paraspinal mass.



Figure 3: coronal T2WI showed uneven hyperintensity and multiple intervertebral foramen enlargements.



Figure 4: MRI coronal enhanced scan showed heterogeneous enhancement and local chest compression.



Figure 5: CT enhanced axial scan showed slight enhancement of the lesion and erosion of the rib and thoracic vertebrae.

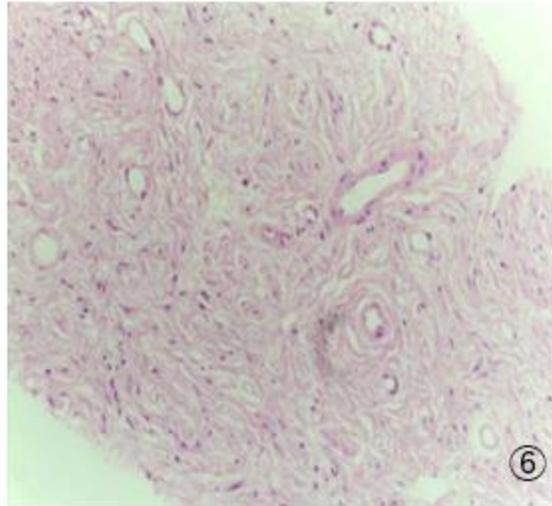


Figure 6: Ganglion cells in some areas (HE staining, * 200)

Cause:

The mediastinum may be divided into compartments, as in adult chest disease, but the causes of mediastinal masses are different (Table 1).

Table 1: CAUSES OF MEDIASTINAL MASS, BY COMPARTMENT.4 CAUSES ARE
Anterior (superior)
Normal thymus** Thymic infiltration (leukaemia, lymphoma, histiocytosis)** Nodal mass** Thyroid Cystic lymphangioma/haemangioma Thymic cyst or thymoma Teratoma Plexiform neurofibroma
Anterior (inferior)
Cardiac* Pericardial cysts, tumours, and fat pads Morgagni hernia (gut or omentum)
Middle

Nodal mass (as above), granulomatous disease (TB)** Vascular and aortic ring anomalies* Bronchogenic cyst (foregut duplication, neurenteric cyst)* Venous anomalies (left superior vena cava) Plexiform neurofibroma
Posterior
Sympathetic chain tumours (neuroblastoma ganglioneuroma sequence)** Hiatal and diaphragmatic hernia (intrathoracic kidney)** Spinal related tumours (plus neuroblastoma from abdomen)* Spinal sepsis (staphylococcal, tuberculosis)* Bronchopulmonary sequestration* Oesophageal duplication, neurenteric cyst Neurofibroma Pheochromocytoma Extension of abdominal pathology (e.g. pancreatitis with pseudocyst) Extramedullary haematopoiesis (haemoglobinopathy)
** = Common; * = occasional; others are uncommon.

DISCUSSION

Ganglioneuroma (GN) is a rare benign neurogenic tumor originating from the sympathetic nervous system and occurring in youth and adulthood. Mediastinal masses are conventionally divided by location into anterior, middle, or posterior mediastinal compartments. This division into compartments is for descriptive convenience only, since it is not based on anatomical boundaries that limit spread; nor do radiologists in general use these terms in the way they are defined by anatomists. Localization of a mass to one of these compartments is a useful step towards reaching the most appropriate diagnosis or differential diagnosis; the age of the patient and such characteristics as the presence of calcification, fat, fluid, or soft tissue within the mass, invasion of the mediastinal fat (indicating malignant rather than benign disease) and contrast enhancement characteristics on computed tomography (CT) and magnetic resonance imaging (MRI) are important in narrowing the differential diagnosis. Retroperitoneal and posterior mediastinum were the most common sites, accounting for 32% ~ 52% and 39% ~ 43% respectively, while neck and pelvic cavity were rare, accounting for 8% ~ 9% [1]. GN is soft, the capsule is intact and smooth, and the section is gray-yellow, microscopically composed of a large number of mucous matrix, nerve fiber cells and a relatively small number of ganglion cells. About 10%~25% can be accompanied by calcification and cystic degeneration and fatty degeneration but is rarely seen [1]. About 25% of GN can contain small amounts of other tumor cell components

such as neuroblastoma, ganglioneuroblastoma or pheochromocytoma. Patients with pheochromocytoma are older, while those with neuroblastoma or ganglioneuroblastoma are younger [1]. Ganglioglioma and Gangliocytoma, Gangliogliomas are uncommon low grade tumors consisting of a mixture of neoplastic astrocytes and dysplastic neurons. Slow-growing neoplasms with well-differentiated ganglion cells and a low-grade astrocytomatous stroma,[7] these tumors are considered WHO grade I or II. [8,9]Gangliocytomas (also known as ganglioneuromas) are relatively less common and contain only neuronal elements; they are classified as WHO grade I. [10] Together, these tumors account for 1.3% of all primary brain tumors. [9] Gangliogliomas and gangliocytomas occur in young people (peak incidence in the second decade of life) and constitute approximately 3% to 4% of primary brain tumors in children. [11,12] They are found most commonly in the temporal lobes but may occur anywhere in the cerebral hemispheres. [12,14] The clinical presentation is usually with seizures of long duration, and gangliogliomas are considered the most common cause of chronic temporal lobe epilepsy. [13] These tumors are usually well circumscribed solid masses but may include one or more cysts. [8,20] Gross total resection of a tumor is the treatment of choice. Overall, gangliogliomas and gangliocytomas have a 92% 3-year survival. [11] In approximately 10% of cases, however, the tumor is more aggressive; in these cases, the malignant element is always glial. [8] Diagnostic imaging findings are relatively nonspecific and are similar to those in other low-grade intracerebral neoplasms. [9,10]Noncontrast CT scans demonstrate relatively well-circumscribed hypodense (occasionally isodense with adjacent brain) lesions typically located in the periphery of the hemisphere with little associated mass effect or surrounding vasogenic edema. Foci of calcification are identified in 33% to 40% of cases, and cysts in 50% of cases.[11,12,15,16] Hemorrhage is rare in these tumors. [12]GN usually has no obvious clinical symptoms and can be found only in local discomfort or pain, occasionally due to tumor secretion of catecholamine, vasoactive intestinal peptide or androgen and cause hypertension, diarrhea or feminization [1]. This patient was diagnosed with congenital heart disease by preoperative physical examination and huge lump. CT plain scan showed homogeneous and slightly soft tissue density and a small amount of spotted calcification. The enhanced scan showed slight enhancement and linear and strip enhancement in the mass. The delayed scan showed more obvious enhancement than the early one, which was consistent with the literature [2, 3]. MRI scan showed moderate T1WI signal and T2WI showed an uneven and slightly hyperintense signal. Because of the long time of MR scan, the enhancement of lesions is more obvious than that of CT scan, which indicates that GN has the characteristics of delayed enhancement. It is generally believed that the presence of a large number of mucous matrix in the stroma of tumor cells leads to the increase of extracellular space, which leads to the progressive accumulation of contrast agents in the space, and is the cause of the slight enhancement in the early stage of tumor and the gradual enhancement after delaying [4,5]. In addition, the growth pattern of GN is very characteristic. According to whether it involves adjacent spinal canals, it can be divided into spinal canal correlation and non-spinal canal correlation [6]. Imaging techniques Mediastinal masses are often incidentally detected on chest radiograph. Despite diagnostic limitations the chest radiograph is also important for detecting and localizing mediastinal masses when suspected clinically. Ultrasound of the mediastinum, including echocardiography and

endoscopic ultrasound, may be of use in selected patients, in particular for distinguishing cystic from solid mediastinal masses and for distinguishing cardiac from paracardiac masses. Ultrasound may also be used to guide mediastinal biopsy. Ganglioneuromas and ganglioneuroblastomas usually arise from the sympathetic ganglia in the posterior mediastinum and therefore usually present radiologically as well-defined elliptical masses, with a vertical orientation, extending over the anterolateral aspect of three to five vertebral bodies [17,18]. Calcification occurs in approximately 25%. CT appearance is variable[17].On MRI ganglioneuromas and ganglioneuroblastomas are usually of homogeneous intermediate signal intensity on T1- and T2-weighted images. Neuroblastomas are typically more heterogeneous due to areas of haemorrhage, necrosis, cystic degeneration and calcium. They may be locally invasive and have a tendency to cross the midline[19]. In this case, tumors embedded in multiple intervertebral foramens belong to spinal canal correlation, which is rarely reported in GN literature. Growth pattern and enhancement characteristics are favorable evidence for the diagnosis of GN, but pathologically, if there are few ganglion cells in ganglioneuroma, it is similar to neurofibroma, and the diagnosis can be made only after the ganglion cells are found carefully.

Invertigation of mediastinal mass:

Table 2: INVESTIGATION OF A MEDIASTINAL MASS
1. Plain radiography. Chest PA and lateral, localized spine and rib views if posterior
2. US if mass anterior, pericardial or basal (solid or cystic mass, nodal or vascular/cardiac)
3. CT. Post intravenous contrast medium. Gives details of location and the relationship to vessels and extension. CT essential for lung detail and metastases.
4. MRI. Pre- and post-gadolinium. MRI is used if there is a posterior mass, associated spinal anomaly or clinical evidence of cord involvement. Axial, sagittal and coronal sections are essential.
Additional imaging
5. Contrast studies: Oesophogram if the mass involves the middle or posterior mediastinum or clinical symptoms of dysphagia or stridor.
6. Radionuclide imaging
99mTc-Pertechnetate: duplication cyst of the oesophagus
123I-MIBG: neuroblastoma-like tumour, whole body

CONCLUSION

Ganglioneuroma (GN) is a rare benign neurogenic tumor originating from the sympathetic nervous system and occurring in youth and adulthood. The mediastinum may be divided into compartments, as in adult chest disease, but the causes of mediastinal masses are different (Table 1). Investigation of mediastinal mass on (table 2). Transthoracic biopsy pathology: nerve fiber hyperplasia, structural disorders, rich blood vessels, ganglion cells were found in some areas. Diagnosis: posterior mediastinal spinal ganglion cell neuroma.

Keyword: ganglioneuroma; posterior mediastinum; Ganglioneuroblastoma ; Neuroblastoma ;CT;MRI

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Conflicts of Interest:

The authors declare no conflicts of interest.

Abbreviations:

GN: Ganglioneuroma; **CT:** Computed tomography; **MRI:** Magnetic resonance imaging; **WI:** Weighted image.

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