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## Recommended Citation

Khan, H., Khan, F. W., Fatimi, S. (2017). Giant ganglioneuroma in a 5-year child. *J Coll Physicians Surg Pak*, 27(3), S16-S17.  
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# Giant Ganglioneuroma in a 5-Year Child

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## ABSTRACT

We report a 5-year boy presenting with pain in the lower chest and upper abdomen. On evaluation with computed tomography scan of the chest, he was found to have a large 16 x 14 cm posterior mediastinal mass compressing the inferior vena cava and liver, and shifting the heart to the opposite side. Tumor was approached via right postero-lateral thoracotomy and dissected off from esophagus, heart, inferior vena cava and lungs. The entire tumor was resected without any complications. Postoperative course was unremarkable. Resection of the tumor was successful. The histopathology showed it to be giant ganglioneuroma with no evidence of malignancy.

**Key Words:** *Giant ganglioneuroma. Pediatric. Posterior mediastinum.*

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## INTRODUCTION

Ganglioneuromas are well-differentiated, slow-growing tumors that have their origins from neural crest cells and arise commonly from sympathetic ganglion cells.<sup>1</sup> They are benign in histology and are composed of Schwann cells, ganglion cells, and fibrous tissue. They belong to a family of neurogenic tumors that are more commonly classified as pediatric mediastinal tumors.<sup>2</sup> Ganglioneuromas arise typically in infants and young children, occurring more in females than males with a ratio of about 3:1.<sup>3</sup> They are usually asymptomatic, and the most common sites are posterior mediastinum and retroperitoneum 37.5% each.<sup>1</sup> The incidence of reported ganglioneuroma is 1 in 1,000,000.<sup>4</sup>

Treatment is complete surgical resection of the tumor, after preoperatively localising the tumor and evaluating its extension *via* CT or MRI because that contributes to the surgical planning. Through this report, the authors aim to inform the reader how a pediatric ganglioneuroma might present, what the diagnostic modalities are, and the standard of treatment followed.

## CASE REPORT

A 5-year boy presented to the physician's office with pain on the right lower chest and upper abdomen. The pain lasted less than a minute occurring once in 5 - 6 days. It was mild in intensity and did not radiate. There were no aggravating or relieving factors and the pain was sharp in nature. There was no history of fever, diarrhea, vomiting, hemoptysis, cough, shortness of breath, headache or weight loss. The mother reported no

complications during pregnancy of the child and a normal vaginal delivery. All his developmental milestones were normal and his vaccinations on schedule. There was no history of a similar case in the family. Physical examination was unremarkable and no swelling was noted. The child was sent home on paracetamol.

On the third episode of pain, an ultrasound and PA chest X-ray (Figure 1) were performed, revealing a well delineated, radiopaque homogenous mass in the posterior mediastinum. The patient was then referred to our cardiothoracic surgery department. CT scan chest (Figure 2) was done, revealing a non-enhancing 16 x 14 cm well-defined, rounded, predominantly hypo-dense mass in the posterior mediastinum on the right side in the right paravertebral location. The mass was causing basal atelectasis of the lung and mild pleural effusion on the right side which was compressing the inferior vena cava and liver, pushing the heart to the opposite side. However, it was not invading any of the structures.

An elective surgery was performed to resect the tumor (Figure 3) via right postero-lateral thoracotomy. It was excised off from esophagus, heart, inferior vena cava and lungs, without any complications. The treatment was tolerated well, and the postoperative course was unremarkable. Histopathological examination of the mass confirmed it to be ganglioneuroma, verifying that it was benign. The patient was discharged within a week and followed for another month, during which recovery was complete and no active complains reported.

## DISCUSSION

Ganglioneuromas are mostly asymptomatic and are revealed when the patient is being assessed for another disorder.<sup>5</sup> Majority of the cases are diagnosed by 10 years of age, however, some may be discovered later in life.

The most common site is mediastinum; however, they might also be found in the gastrointestinal tract, bones, parapharyngeal and supraclavicular regions.<sup>6-9</sup>

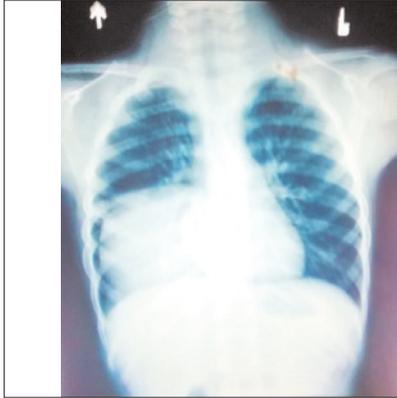
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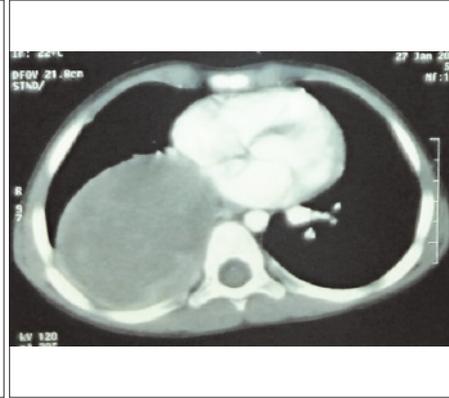
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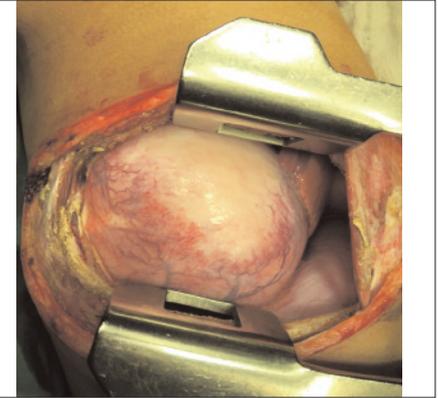
*Received: February 09, 2016; Accepted: October 27, 2016.*



**Figure 1:** Chest X-ray showing large opacity in the right lower chest.



**Figure 2:** CT chest with contrast: Well defined hypo-dense mass in the right posterior mediastinum.



**Figure 3:** Ganglioneuroma measuring 16x14cm as seen through right postero-lateral thoracotomy.

Symptoms are governed by the position of the tumor compressing the surrounding structures. Cough, chest pain, dyspnea and pneumonia by mediastinal location of the tumor, vomiting, constipation, abdominal pain and weight loss by abdominal location of the tumor.<sup>1</sup> Horner's syndrome, if compressing the cervical ganglion.<sup>9</sup> If the retroperitoneal space is disturbed, abdominal pain and bloating may occur. If the tumor is near the spinal cord, compression may result in pain and loss of sensation and movement in the arms, legs or both. Hormones/catecholamines released by the tumors can cause diarrhea, enlarged clitoris (in women), sweating and high blood pressure.

Imaging studies prove very useful in diagnosing ganglioneuroma preoperatively. A study by Guan *et al.* showed that, on plain CT thoracic ganglioneuromas present as homogenous or heterogenous hypodensity mass with a Hounsfield value of 20 - 40 HU with punctate calcification as an additional finding.<sup>10</sup> With contrast, there is no or slight enhancement in the arterial phase and a progressive enhancement in the delayed phase after 120 seconds. On T1 weighted MRI, the images show homogenous hypointense signals and on T2 the images demonstrate heterogenous hyperintense signals. Furthermore, in T1, T2 and Gadolinium enhanced MRI there is non-enhancement or slight enhancement in the artery phase and progressive mild enhancement in the delay phase, similar to what is observed in the CT contrast. Thoracic ganglioneuroma can mimic cystic teratoma, neurilemmoma, lymphangioma cysticum or bronchocele on imaging and needs to be well differentiated to avoid misdiagnosis.<sup>10</sup>

Diagnosis can be proved via presence of neuroblasts in the biopsy. Surgical excision is the treatment of choice

for ganglioneuroma, especially if there are symptoms due to the tumor compression, marked increase in size or encroachment on the vertebral foramina.

## REFERENCES

1. Geoerger B, Hero B, Harms D, Grebe J, Scheidhauer K, Berthold F. Metabolic activity and clinical features of primary ganglioneuromas. *Cancer* 2001; **15**:1905-13.
2. Bower RJ, Kiesewetter WB. Mediastinal masses in infants and children. *Arch Surg* 1977; **112**:1003-9.
3. Joshi VV. Peripheral neuroblastic tumors: Pathologic classification based on recommendations of international neuroblastoma pathology committee (modification of shimada classification). *Pediatric and developmental pathology. Pediatr Dev Pathol* 1999; **3**:184-99.
4. Shetty PK, Shetty PK. Ganglioneuroma always a histopathological diagnosis. *Online J Health Allied Sci* 2011; **9**:4.
5. Cataldo AND, Lanteri RA, Trombatore GI, Licata AN. Mediastinal ganglioneuroma: A rare and often asymptomatic tumour. A case report. *Chir Ital* 2005; **57**:403-5.
6. Rao BS, Ravi G. Ganglioneuroma of small intestine presenting with perforation peritonitis. *Indian J Gastroenterol* 2000; **20**:31-2.
7. Mithöfer K, Grabowski EF, Rosenberg AE, Ryan DP, Mankin HJ. Symptomatic ganglioneuroma of bone. A case report. *JBJS Case Connector* 1999; **1**:1589-95.
8. Albonico G, Pellegrino G, Maisano M, Kardon DE. Ganglioneuroma of parapharyngeal region. *Arch Pathol Lab Med* 2001; **125**:1217-8.
9. McRae D, Shaw A. Ganglioneuroma, heterochromia iridis, and Horner's syndrome. *J Pediatr Surg* 1979; **14**:612-4.
10. Wang YM, Li YW, Sheih CP, Hsu JC. Magnetic resonance imaging of neuroblastoma, ganglioneuroblastoma, and ganglioneuroma. *Zhonghua Min Guo Xiao Er Ke Yi Xue Hui Za Zhi* 1994; **36**:420-4.

