BACKGROUND: Intrathoracic tumor is a rare entity in the pediatric age and neurogenic tumors account for 40-50% of childhood intrathoracic tumors. They generally result severe symptoms, such as respiratory distress, neurological dysfunction and metabolic disturbances. Posterior mediastinal ganglioneuroma (GN) usually occurs in children and can be found accidentally. Precise preoperative diagnosis is very difficult and has a great influence on surgical intervention.

CASE REPORT: A 12 Year old boy was investigated for recurrent cough of short duration and found to have huge posterior mediastinal mass on chest radiograph. There was no history of fever or respiratory difficulty. Clinical examination revealed decreased air entry on the left side. All the other Systems were normal. Routine haematological and Biochemical investigations were within normal limits. CT Chest showed homogenous mass in the left paraspinal region of 10 X 7.2 X 7.5 cm size with peripheral calcification and thickening of third and fourth ribs and the spine was normal. CT guided FNAC suggestive of benign lesion. Thoracotomy and excision of the mass was done. Histopathology showed a maturing ganglioneuroma.

DISCUSSION: Ganglioneuroma is a very rare tumour characterised by a slow evolution and arising from cells originating in the neural crest. This type of tumour occurs most commonly in the posterior mediastinum and is considered malignant in childhood and benign thereafter. Ganglioneuromas may also arise in the retroperitoneum, adrenal gland, and retropharyngeal soft tissues. The totally asymptomatic clinical picture could be explained by the very slow growth of this type of tumour which can displace the surrounding anatomical structures without infiltration.

However, some patients who had increased diameter of the tumor may be presented with cough, neck swelling, dyspnea, chest pain, Horner’s syndrome and back pain. Approximately 10% of tumors may extend to spinal channel and cause neurologic symptoms due to compression. These tumors are called as dumb-bell or hour-glass tumors. Rarely present with labile hypertension and flushing, as these tumors may produce excessive catecholamines. Unlike the case with neuroendocrine tumors, catecholamines are metabolized in the tumor, and this fact probably accounts for the relative lack of symptoms. Ganglioneuromas have also been associated with a syndrome of chronic diarrhea, which is usually found in children and is possibly mediated by vasoactive intestinal peptide. This symptom as well as hypertension resolve after removal of the tumor.
Macroscopically, ganglioneuromas are well-circumscribed, unencapsulated, benign tumors with a firm gray to yellow surface. Microscopically, the tumor is composed of mature ganglion cells admixed with spindle cells of Schwannian origin and nerve fibers. Mature ganglion cells consist of a large nucleus with nucleoli and a granular, basophilic cytoplasm, containing basophilic granules. Malignant form of ganglioneuroma is ganglioneuroblastoma. One fourth of the tumors demonstrate calcification and rib or vertebral erosion that is subtle but often apparent.

Magnetic resonance imaging (MRI) and computed tomography (CT) scanning are the preferred methods for imaging ganglioneuromas and ganglioneuroblastomas. MRI is the modality of choice for evaluating the extension of spinal tumors. In general, neuroblastic or neurogenic tumors appear radiologically as well-circumscribed, smooth or lobulated masses that may contain calcifications. The benign (ganglioneuromas) and malignant (ganglioneuroblastomas) forms of these tumors are virtually identical radiologically. The only differentiating factor is the possibility of distant metastases with malignant ganglioneuroblastomas.

If a ganglioneuroma is confirmed by pathology, surgical excision alone is curative. Postoperative adjuvant radiotherapy is controversial. In the tumors of "hourglass" that has spinal extension, it should be done bilateral laminectomy and fusion with bone graft in the first intervention and then excision via thoracotomy.

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