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Case Report

Doege Potter Syndrome

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ABSTRACT

Doege Potter Syndrome is a benign solitary fibrous tumor of pleura having a paraneoplastic presentation in the form of recurrent hypoglycemic episodes. An elderly male presented to us with recurrent attacks of hypoglycemic seizures. On imaging studies, the patient was found to have a large mass extending into right middle and lower lobes of lung. A CT- guided biopsy was performed, which on microscopy and immunohistochemistry, was suggestive of a benign solitary fibrous tumour of pleura. Surgical resection of the tumor is the treatment of choice and is curative. Unfortunately, our patient succumbed to an episode of hypoglycemia before operative intervention could be undertaken.

Key-words : Doege Potter Syndrome, solitary fibrous tumour, hypoglycemia, paraneoplastic manifestation.

Introduction :

Solitary fibrous tumors are uncommon soft tissue tumors of mesenchymal origin initially reported as a pleura based lesion but now found to occur at any site. They account for about 5% of all pleural tumors.[1,2] About 80% are benign and 50% present as an asymptomatic mass.[3] 10-20% cases present with hypertrophic osteoarthropathy, also known as Pierre-Marie-Bamberg syndrome. In 2-4% cases, it presents as recurrent attacks of hypoglycemia as was in our case. It is then called Doege Potter Syndrome. Hyopoglycemia occurs due to increased production of insulin-like growth factors, chiefly IGF-2. Symptoms are directly related to size of tumour. Immunohistochemical markers used for confirmation include vimentin, CD34 and BCL-2. Surgical resection is curative.

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Case Report :

A 62 year old male patient came with chief complaints of generalized seizures since two months with a frequency of 4-5 episodes per day. The patient always had a documented low blood sugar (40-50 mg/dl) at the time of seizures. The seizures were aborted on administration of intravenous dextrose. There was a history of removal of a mass from right thorax twenty years back, details of which were not available. The patient was non-diabetic. On physical examination, he had a pulse of 80/min, respiratory rate of 14/min, blood pressure of 140/90 mmHg. There were no signs of superior vena caval obstruction, pallor or clubbing. He had a postoperative scar in right subcostal region extending upto right infrascapular area. Trachea was central. There was dull note on percussion and decreased intensity of breath sounds in right mammary, axillary and infrascapular areas.

The hemogram, liver function test and renal function test were normal. Glycosylated haemoglobin was 5.5%, fasting insulin, serum cortisol and C-peptide were normal. Chest roentgeogram revealed a mass in right middle and lower zone ***(Figure 1).***

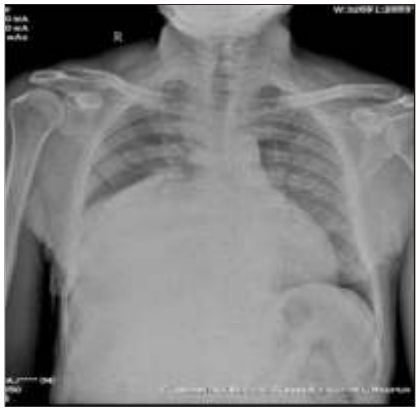

Figure 1 : X-ray chest (PA view) showing homogenous radio-opacity in right lower zone of lung

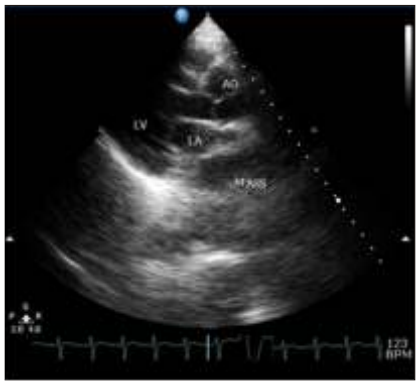

Figure 2 : Echocardiogram showing supra-cardiac mass compressing craniocaudally

On echocardiography, a supracardiac mass was seen compressing craniocaudally ***(Figure 2).***

A CT scan of thorax and abdomen showed a large well defined heterogenously enhancing mass of size 15 x 16 x 13 cm having necrotic area within involving right middle and lower lobes of lung with perihilar and mediastinal extension displacing cardiac chambers towards left. Liver and pancreas were normal ***(Figure 3).***

A CT-guided biopsy of the tumor was performed which on histopathology showed slender spindle cells in fascicles and bland nuclear features with intervening collagenous stroma with no mitosis or necrosis suggestive of benign fibrous tumor arising from pleura ***(Figure 4).*** On immunohistochemistry, the tumor was positive for CD34, vimentin, BCL-2, KI-67 and negative for cytokeratin further confirming the diagnosis.

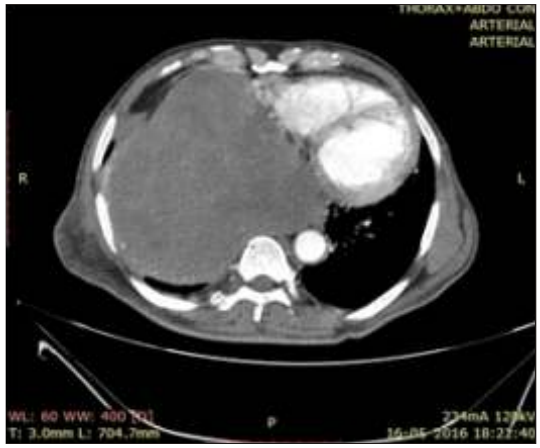

Figure 3 : Contrast enhanced computerized tomography scan of thorax showing a large mass occupying right hemithorax compressing the heart

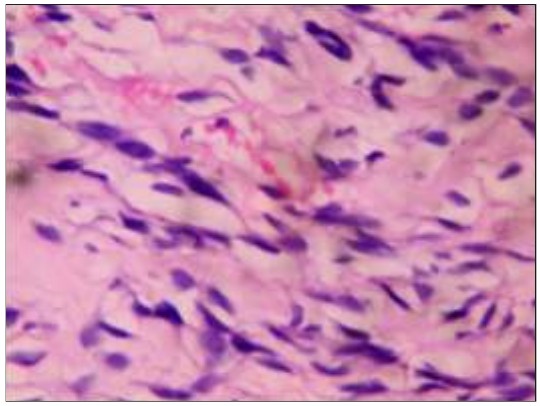

Figure 4 : Histopathology showing spindle cells with intervening collagenous stroma with no malignant features

Unfortunately, the patient succumbed to an episode of hypoglycemia before surgical resection.

Discussion :

Solitary fibrous tumor of pleura (SFTP) are rare tumors of mesenchymal origin which are often mistaken for mesothelioma. They were first reported in 1931.[4] The incidence is < 5% of all pleural neoplasms. Mostly these neoplasms are considered benign but large tumors have a tendency to show malignant potential.

Most patients are asymptomatic and present to the clinician only if the pleural tumor starts compressing on lung parenchyma causing symptoms in the form of chest pain, breathlessness and in some cases, hemoptysis.

An alternate presentation is in the form of paraneoplastic syndromes, the commonest of which is Hypertrophic Pulmonary Osteoarthropathy (HPO), also called Pierre-Marie-Bamberg syndrome, having an incidence of 22%.[5] The cause for HPO remains unknown, some postulate it to be due to ectopic growth hormone production.

In only 3-4% cases, patients present with tumor-associated hypoglycemia. This was first reported by Doege and Potter in 1930, hence the name Doege Potter syndrome. Hypoglycemia occurs more frequently in females, tumors located in right hemithorax(as was in our case), in large SFTs (> 20 cm) and in SFTs with high mitotic rate.[6] Hypoglycemia occurs due to increased production of pro-IGF- II and IGF-II.

Both the paraneoplastic syndromes disappear after tumor resection.

Diagnostic modalities include imaging studies, histopathology and immunohistochemistry. A chest computed tomography scan gives details regarding the size and location of tumor. Magnetic resonance imaging is seldom required if invasion is suspected. Histological features show cellular areas with intermittent hyalinized or necrotic areas with fibroblasts and mesothelial cells. On immunohistochemistry, the tumor stains positive for vimentin and negative for cytokeratin differentiating it from mesothelioma.in addition they are CD34-,CD99- and bcl-2 positive.

Complete surgical resection is curative. However, the tumor has a high recurrence rate, 8% in benign neoplasms and 63% in case of malignant tumors.[3] Re-resection of recurred tumors can be undertaken. Our patient also had a history of removal of mass from right hemithorax.

Conclusion :

Solitary fibrous tumor of pleura is a curable entity. Paraneoplastic syndromes disappear after complete surgical resection. The SFTs have a high recurrence rate. Hence the patient needs to be kept in follow up.

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