

A Rare Case of Large Anterior Mediastinal Mass Presenting As Right Ventricular Outflow Obstruction and Congestive Hepatopathy

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Abstract

Thymoma is a rare tumour yet the most common neoplasm of the anterior mediastinum, accounting for 20-25% of all mediastinal tumours. Its incidence peaks in the 4th and 5th decades of life. Usually thymoma is diagnosed incidentally on chest imaging, when investigating patients with myasthenia gravis, with chest symptoms such as cough, chest pain, or dyspnoea. This case report was a 50year old female patient who presented to OPD with gross abdominal distention, bilateral pedal oedema, facial puffiness and easy fatigability. Chest radiograph showed widening of mediastinum, USG abdomen showed gross ascites following which patient shifted for CECT chest & abdomen which revealed a large bilobed heterogenous thick walled anterior mediastinal lesion which extends to either side of midline. The lesion was seen to cause severe compression over the right ventricle resulting in dilatation of right atrium, SVC and IVC. On left side the lesion was seen to compresses the left ventricle, the main pulmonary artery, left superior pulmonary vein,

and left main bronchus. Features of congestive hepatopathy were seen. This is, to our knowledge, the second case of thymoma to present this way with right-side heart failure like symptoms.

Keywords: Large anterior mediastinal mass, Thymoma, Right ventricular outflow obstruction, Refractory ascites, Congestive hepatopathy.

Introduction

Thymoma originating from the epithelial cells of the thymus was an uncommon tumour ^[1]. It is best known for its association with myasthenia gravis (MG) which occurs in about 50% of patients with thymoma. Most cases occur between the ages of 40 & 60 years ^[2] with a mean age at presentation being 52 years. There are three types of main presentations of thymoma first one it presents as an asymptomatic anterior mediastinal mass on chest roentgenogram in 30-50% of patients. Second one during the evaluation of MG or other paraneoplastic syndromes such as pure red cell aplasia or acquired hypogammaglobulinemia ^[3] in one third of patients and third one presents with local symptoms

such as cough, superior vena cava syndrome, chest pain, or dysphagia in one third of patients [1]. This report describes thymoma in a 50 year old female patient who presented with a picture of right-side heart failure with gross ascites and pedal oedema. However thymoma presenting as right ventricular outflow obstruction very rare.

Case Report

A 50 year old female patient admitted in general medicine department with chief complaints of gradual onset of abdominal distension, bilateral pedal edema, and facial puffiness since 1 year. Chest radiograph & Ultrasound abdomen were advised which showed widening of mediastinum (Fig: 1), dilated hepatic veins and IVC in liver & gross ascites (Fig: 2). After that CECT chest& abdomen were requested which showed a large bilobed heterogeneous thick walled anterior mediastinal lesion measuring 11.6x 15x9.6 cm (CCxTRxAP) of predominantly fluid attenuation with some high attenuation areas within (avg HU 10-40), which extends to either side of midline. On non-contrast study wall of the lesion showed calcifications (Fig: 3). Gross ascites was present.

On contrast study during arterial phase reflux of contrast material into the inferior venacava and hepatic veins, due to high right atrial pressure (Fig:4).The lesion was seen to cause severe compression of the right ventricle with resultant dilatation of right atrium, SVC and IVC(Fig:6). During portal venous phase liver showed heterogeneous enhancement pattern giving nutmeg appearance due to congestive hepatopathy (Fig:5).On left side the lesion was seen to compresses left ventricle, the main pulmonary artery, left superior pulmonary vein, and left main bronchus(Fig:7).

Fat planes were lost with major vessels (SVC, Pulmonary artery) and also with left ventricle.

Figures

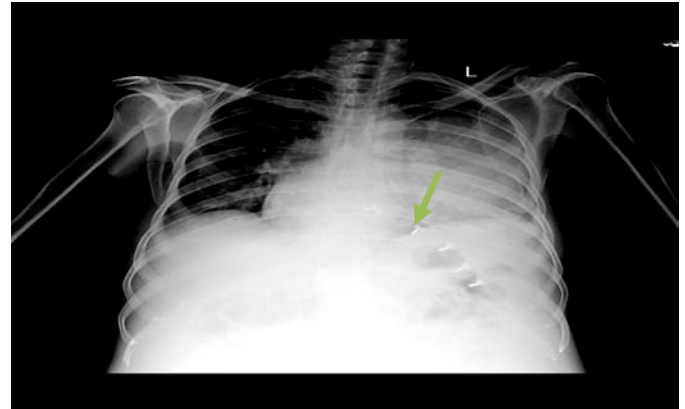


Fig: 1 Frontal chest radiograph showed widening of mediastinum.

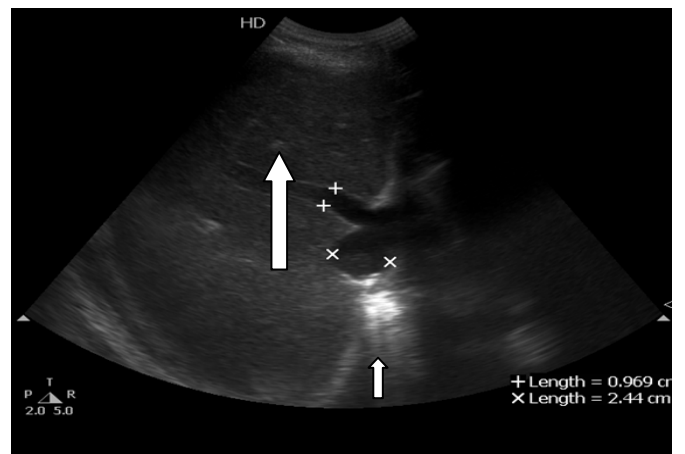


Fig: 2 USG abdomen showed dilated hepatic vein (large arrow) & dilated IVC(small arrow).



Fig: 3. On NCCT A large bilobed lesion in anterior mediastinum with peripheral wall calcifications.

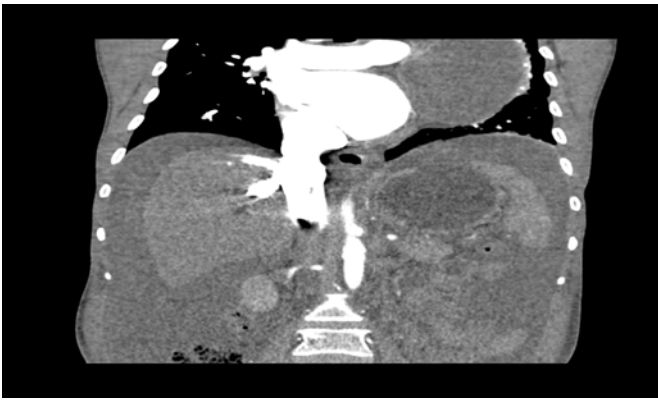


Fig : 4.CECT abdomen coronal section arterial phase showed Retrograde opacification of IVC and hepatic veins during arterial phase.

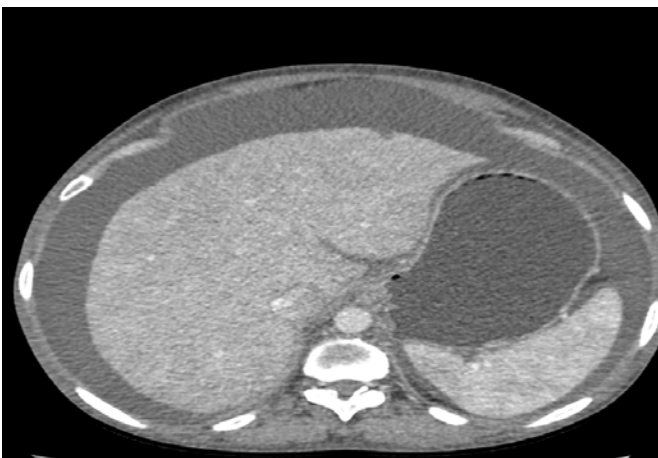


Fig : 5. CECT abdomen axial section portal venous phase Diffuse heterogenous enhancement of liver.



Fig: 6.CECT chest & abdomen during portal venous phase, severe compression of the right ventricle (white arrow) resultant dilatation of right atrium.

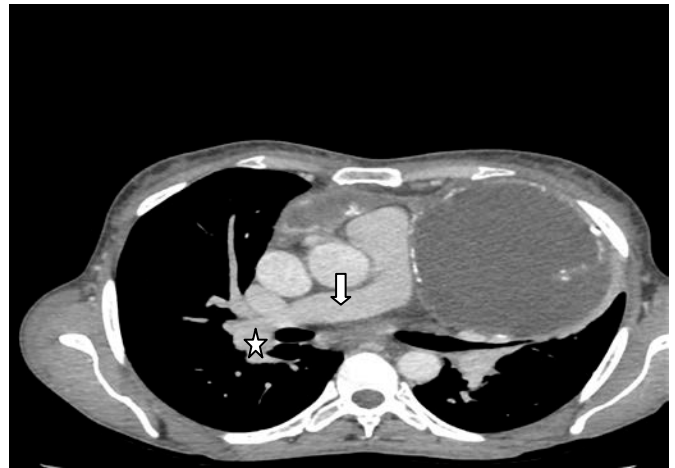


Fig: 7. Dilatation of SVC (asterisk) and compression of the main pulmonary artery (arrow), superior pulmonary vein and main bronchus.

Discussion

A massive mediastinal mass represents an uncommon but potentially deadly clinical entity. Anterior mediastinal mass presents with facial swelling from impaired venous drainage from the superior vena cava, oesophageal narrowing causing dysphagia, or dyspnea due to pericardial effusion, cardiac compression, or dynamic tracheal stenosis [4]. A mediastinal mass is associated with a pericardial effusion, and the mediastinal mass may results in tamponade-like physiology by causing direct compression of the right ventricle, superior vena cava, or right ventricular outflow tract, resulting in tamponade-like physiology [4].

Thymomas usually present in one of three major ways as an incidental finding on thoracic imaging in an asymptomatic patient or because of local (thoracic) symptoms such as chest pain, cough, and shortness of breath or during the evaluation of a paraneoplastic syndrome such as MG, pure red cell aplasia, and hypogammaglobulinemia among others [5].

A patient with suspected thymoma should be initially evaluated by a thoracic imaging like computed tomography scan and/or magnetic resonance imaging,

which establishes the presence of an anterior mediastinal mass and delivers the initial information about the resectability of the tumour. The differential diagnosis includes lymphoma, retrosternal thyroid, and mediastinal germ cell tumour [6]. On CT teratomas shows fat, fluid, soft tissue components and calcifications. Retrosternal thyroid appears high attenuation on unenhanced CT & intense and prolonged enhancement on CECT, Thus preoperative assessment may include germ cell tumour markers (BHCG and AFP) and thyroid function tests.

Surgery with radical excision of the tumour is usually the first step in the management of thymomas, followed by radiotherapy and chemotherapy according to the stage of the disease [7]. For those patients thought to have a tumor that is amenable to complete resection, the initial step in management is surgical resection, which will establish the diagnosis and provide the primary treatment. In contrast, for patients who have a disease that is not considered amenable to complete resection or for whom surgery is contraindicated for any reason, a tissue diagnosis using either a core biopsy or an open biopsy is required prior to therapy [8,9].

In this case report 50 years female presented with large anterior mediastinal mass and gross abdominal ascites, bilateral pedal edema, resembling the presentation of a right-side heart failure. Most cases of thymoma occur between the fourth and fifth decades of life, the patient had none of the usual presenting symptoms of thymoma neither she had the symptoms of MG. Instead, she presented with features of right ventricular outflow obstruction. On CECT imaging features were similar to the thymic mass rather than any other anterior mediastinal masses. In this case tissue diagnosis done by core biopsy, surgery was contraindicated due to

stage IIIB thymoma (Involvement of Major vessels). Radiotherapy and Chemotherapy was given.

Conclusion

Thymoma presenting with features of right ventricular outflow obstruction extremely rare. To our knowledge there are only few cases of such unusual presentation of thymomas reported in English literature. The main purpose of this report is to discuss the rare presentation of thymoma and the role of imaging in aiding the diagnosis. CECT very useful not only for characterization, mass effects, extensions of the lesion and staging but also elicits features of right ventricular out flow obstruction caused by lesion such as congestive hepatopathy well delineated.

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