Case Report

A rare clinicoradiological presentation on giant thymoma extending bilaterally in both hemi-thoraces: Case report with review of literature

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ABSTRACT

We present the case of a giant thymoma in a 70-year-old lady presenting as an intrathoracic mass lesion extending bilaterally, filling most of the space in both hemi-thoraces. The rarity of the occurrence of giant thymoma and unusual clinicoradiological presentation makes it an interesting case report. It is indicated that the diagnosis of thymoma should be considered in any intra-thoracic anterior mediastinal mass lesion.

Key words: Median sternotomy, Mediastinal tumors, Thymoma

Mediastinal masses comprise a variety of benign and malignant lesions. A thymoma is a very rare tumor that originates from the epithelial cells of the thymus gland situated in the anterior mediastinum [1]. Thymomas are typically found on one side of the mediastinum. Extension to both hemi-thoraces makes it a very rare type [2]. Thymomas are frequently associated with variety of paraneoplastic syndromes, the most common being myasthenia gravis [3]. It is found in 20% of patients with myasthenia gravis [4]. Thymoma occurs in patients of all ages, with a peak incidence between the ages of 40–60 years. It may be identified incidentally on imaging performed for unrelated reasons. Diagnostic modalities include history, physical examination, imaging, and histopathology of the tumor. There are mainly three histologic types (type a, b, and AB). Masoaka-Koga classification is used for staging of thymoma [5]. Surgical resection is the mainstay of treatment.

We are reporting this case because giant thymoma is an extremely rare clinical condition and is very uncommon for a thymoma to reach both hemi-thoraces [2].

CASE REPORT

A 70-year-old lady presented with the complaints of fever, cough, running nose, and body aches for 2 days. There were no neurologic, respiratory, cardiac, or other complaints.

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Her vitals were normal with a blood pressure of 124/78 mmHg, respiratory rate of 20/min, and pulse of 82/min. Her body temperature was 99.5°F. There was no pallor, cyanosis, icterus, pedal edema, clubbing, or vascular engorgement. Examination of the chest revealed dull note in mid and lower zones and diminished breath sounds on the anterior side, as well as on the posterior side bilaterally. There were no added sounds. Cardiac sounds, S1 and S2, were normal, and there was no murmur. Abdomen was soft and non-tender with no organomegaly. There was no drooping of eyelids or double vision. There was no dysphagia, hoarseness, impaired speech, or changes in facial expressions.

Laboratory investigations were as follows: Total leukocyte count – 5600/µL, polymorphs 68%, lymphocytes 28%, eosinophils 3% basophils 1%, and platelets 2.3 × 10^6/µL. Hemoglobin was 10 g/dL and blood sugar (R) was 136 mg/dL. Renal functions and liver functions were within normal range. Acetylcholine receptor (AChR) antibody level was normal (0.01 nmol/L). Patient could not do pulmonary function test. Her X-ray chest revealed a big mediastinal mass extending in both hemi-thoraces (Fig. 1). Computed tomography (CT) scan showed a giant tumor of size 18 × 9.5 × 16.8 cm, occupying the anterior mediastinum and extending in both hemi-thoraces (Fig. 2). No invasion of adjacent structures by tumor was noted. Percutaneous needle biopsy of the tumor was taken and subjected to histopathology examination. Histopathology showed oval epithelial cells, indicating a B-Type

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thymoma (Fig. 3). Echocardiography of the patient revealed mild pericardial effusion, which was considered due to the mass effect of the tumor compressing major vessels. Her fever was considered viral fever, which subsided the next day without any medication.

Surgical resection of the tumor was performed through median sternotomy. She recovered well and was discharged in satisfactory condition.

DISCUSSION

Benign or malignant mediastinal masses can develop from structures that are normally located in the mediastinum or that pass through the mediastinum during development. The anterior mediastinum is the most common location where mediastinal masses occur in adults. The most commonly found anterior mediastinal masses are thymoma, teratoma/germ cell tumor, lymphoma, and thyroid tissue.

Thymoma originates from epithelial cells of the thymus gland. Thymomas account for approximately 20–25% of all mediastinal masses [6-8]. The overall incidence of thymoma in the US is 0.13/100,000 person-years [9]. Vasudevan *et al*. also reported a similar incidence in India [10]. Thymomas occur in patients of all ages with a peak incidence between the ages of 40–60 years [6-8]. Giant thymomas in adults are very rare [1,2,11]. Thymomas are typically found on one side of the mediastinum. Our case had bilateral thymoma, which made it a very rare case. To the best of our knowledge, only one case of bilateral thymoma has been reported in the literature [2]. They grow very slowly and expansively. The most cases, including our case, were asymptomatic [12]. Symptoms, if present, may be due to direct mass effect. Direct involvement or compression of normal mediastinal structures causes a wide variety of symptoms, including chest pain, dyspnea, cough, stridor, dysphagia, and facial or neck swelling due to vascular compression (e.g., superior vena cava syndrome). Systemic symptoms may be due to a variety of paraneoplastic syndromes, such as myasthenia gravis or less commonly, hypogammaglobulinemia and pure red cell aplasia [3,13].

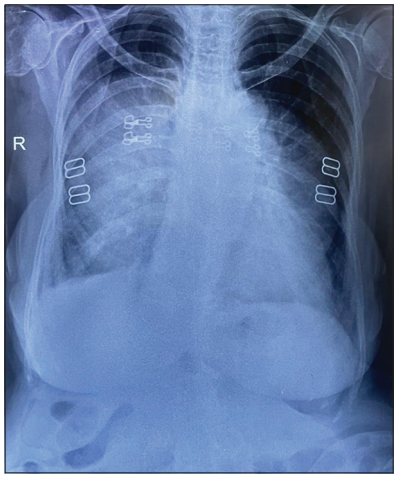

Figure 1: X-ray chest showing a giant mass in the mediastinum extending in both right and left hemi-thoraces

The diagnostic evaluation of thymoma includes a thorough history and physical examination, imaging, laboratory studies, and tissue sampling. The chest X-ray is a basic imaging technique which can show masses in the chest. For more detailed information, a contrast-enhanced CT of the chest is required which can establish presence of anterior mediastinal mass, distinguish between thymic malignancy and other benign etiologies such as thymic cyst or thymic hyperplasia, provide initial evidence to distinguish between thymoma and thymic carcinoma, and demonstrate staging of thymoma [14,15]. Magnetic resonance imaging and positron emission tomography/CT scans can also aid in distinguishing between thymoma and thymic carcinoma [16,17].

The diagnosis of a thymoma is confirmed by a tumor tissue sample and histopathological analysis of the sample. For patients amenable to complete resection based on imaging studies, the diagnosis can be confirmed through surgical resection. If the imaging results are inconclusive or if the patients are not amenable to complete resection, a tissue diagnosis can be established with a core needle biopsy or an open thoracoscopic biopsy [18].

According to the updated World Health Organization classification, there are three principal histological types of thymoma, depending on the appearance of the cells by microscopy [1,19,20]. The classification is as follows: Type A: If the epithelial cells have an oval or fusiform shape (spindle bland – looking) neoplastic epithelial cells; Type B: If epithelial cells have an epithelioid shape, round or polygonal (Type B has three subtypes: B1 (lymphocyte-rich), B2 (cortical) and B3 (epithelial)

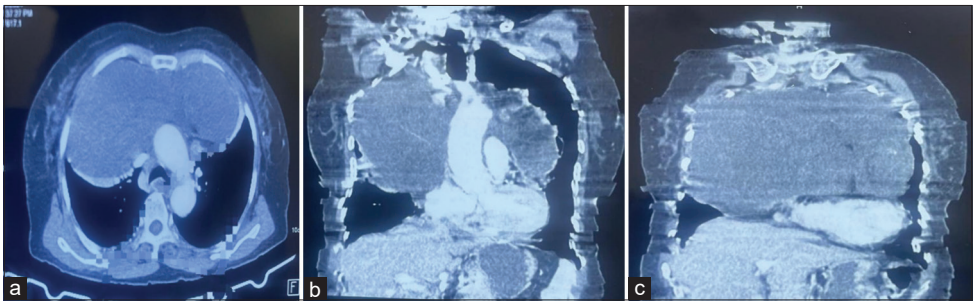

Figure 2: (a) Computed tomography (CT) scan showing a large capsulated mass in the anterior mediastinum extending in both hemi-thoraces; (b) CT scan with contrast enhancement showing an encapsulated giant mediastinal mass extending in both hemi-thoraces; and (c) CT scan with showing an encapsulated giant anterior mediastinal mass occupying both hemi-thoraces

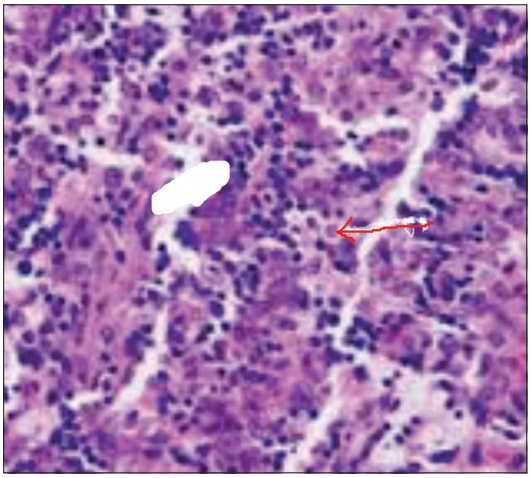

Figure 3: Histopathology of tumor showing round epithelial cells (arrow) indicating a B type thymoma

depending on the extent of the lymphocytic infiltrate and the degree of atypia of the neoplastic epithelial cells; Type AB if the tumor contains a combination of both cell types. In our case, histology revealed a B-type lymphoma.

The International Thymic Malignancy Interest Group has selected the Masaoka system [21] with the modification proposed by Koga *et al*. [22] as the one that will be used [5]. It is mainly based on the invasiveness of the tumor through the capsule into surrounding tissues and neighboring structures. The stages are 1–4. It is stage 1 if the tumor is microscopically and grossly completely encapsulated, while in stage IV, there is pleural or pericardial metastasis or there is evidence of hematogenous or lymphogenous metastasis [21,23]. The clinical stage in our case was stage 1, as the tumor was completely encapsulated without any evidence of invasion of any adjacent structures.

Some laboratory tests should be done to look for associated problems or possible tumor spread. These tests include: Complete blood count, protein electrophoresis, anti-AChR antibodies (indicative of myasthenia), electrolytes, liver enzymes, renal function, etc. [3]. In our case, there was no evidence of any associated problem or tumor spread.

As far as the treatment of thymoma is concerned, surgery is the mainstay of treatment. If the tumor is apparently invasive and large, pre-operative neoadjuvant chemotherapy and/or radiotherapy may be used to decrease the size before surgery is attempted. Invasive thymoma may require additional treatment with radiotherapy and chemotherapy [23] The type of thymectomy depends on the invasiveness of the tumor. In simple thymectomy, there is a complete removal of the thymus gland, in extended thymectomy, there is removal of gland along with surrounding tissues and lymph nodes, while in radical thymectomy, there is removal of invaded tissues, including parts of the lungs or other structures [24]. The surgical approach includes open surgery median sternotomy. However, now the surgical approach is shifting toward minimally invasive techniques such as video-assisted thoracic surgery and robotic surgery [25,26]. Surgical removal of thymoma in our case was performed through median sternotomy, as though thymoma was very giant, but it was without any evidence of invasion of surrounding structures. Median sternotomy involves making a vertical incision through the sternum to allow for the resection of the thymoma. It is the standard approach for many thymoma cases due to its broad exposure and ability to address various tumor sizes and locations [27]. The patient recovered well and was discharged after 7 days.

CONCLUSION

Thymomas are very rare epithelial tumors of the thymus gland. They have a wide spectrum of morphological, pathologic characteristics, and clinical presentations. Although it is a benign tumor, it can invade surrounding structures and metastasize. Hence, clinicians should have a high index of suspicion for early diagnosis and treatment.

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