

# Case report of 49-year-old man with unusual invasive thymoma

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## Abstract

Invasive thymomas are rare anterior mediastinal tumor, representing 50% of anterior mediastinal masses and about 20%–30% of all mediastinal tumors. They are of unknown etiology; about 50% of patients with thymomas are diagnosed incidentally with chest radiography. Thymoma is classified into different stages, which determine the prognosis and type of management, the standard primary treatment for these tumors is thymectomy.

**KEY WORDS:** Mediastinal tumors, anterior mediastinal masses, tumors of the thymus, invasive thymomas

## Introduction

The thymus is an anterior mediastinal lymphoid organ that is mainly composed of epithelial cells in the outer cortex, myoid cells, and lymphocytes in the germinal center. Being located in the upper anterior mediastinum and lower part of the neck, the thymus is active during childhood and involutes after puberty being replaced by adipose tissue gradually thereafter, although it never disappears completely.<sup>[1]</sup> The thymus is responsible for the processing and maturation of T-lymphocytes. After birth, the thymus starts to grow to reach its maximum weight of 40 g around puberty, and then it involutes and persists in an atrophic state into old age. T-lymphocytes develop from a common lymphoid progenitor in the bone marrow that also gives rise to B-lymphocytes, but those progeny destined to give rise to T-cells leave the bone marrow and migrate to the thymus. This is the reason they are called thymus-dependent (T) lymphocytes or T-cells.<sup>[2]</sup>

Thymomas are relatively rare mediastinal tumors arising from epithelial cells and accounting for approximately 0.2%–1.5% of all malignancies.<sup>[3]</sup> It is the most common neoplasms

arising in the thymus originating from epithelial cells of thymus, it accounts for about 25% of all mediastinal tumors with a peak incidence between 40 and 50 years of age. Thymus originates in the embryo from the ventral ring of third and fourth pharyngeal pouches and ectoderm endoderm of the cervical sinus, as epithelial outgrowths on each side.<sup>[4,5]</sup>

Thymic epithelial tumors originate from the thymic epithelium and are distinct from nonepithelial primary thymic tumors such as lymphomas or germ cell tumors. Thymic epithelial tumors have traditionally been classified into three subgroups including benign thymoma, type I malignant thymoma (invasive thymoma), and type II malignant thymoma (thymic carcinoma). More recently, however, they have been reclassified according to prognostic implications into thymomas, atypical thymomas, and thymic carcinomas.<sup>[6]</sup>

Thymoma is generally asymptomatic, but it may present with local symptoms related to encroachment on adjacent structures, as cough, chest pain, or superior vena cava syndrome. In case of disseminated disease, the most common manifestations are pleural or pericardial effusions, which may be associated with thoracic symptoms. The most common presenting symptom of an ectopic cervical thymoma is an enlarging neck mass and is frequently misdiagnosed on fine-needle aspiration cytology. The diagnosis is difficult to make and has a major diagnostic pitfall.<sup>[7]</sup>

Imaging evaluation of the thymus is challenging because thymic size, shape, and consistency change with age, and there is a moderate degree of normal variation in glandular appearance between individuals. Knowledge of the normal spectrum of thymic appearances is important to prevent overdiagnosing or underdiagnosing thymic lesions.<sup>[8]</sup>

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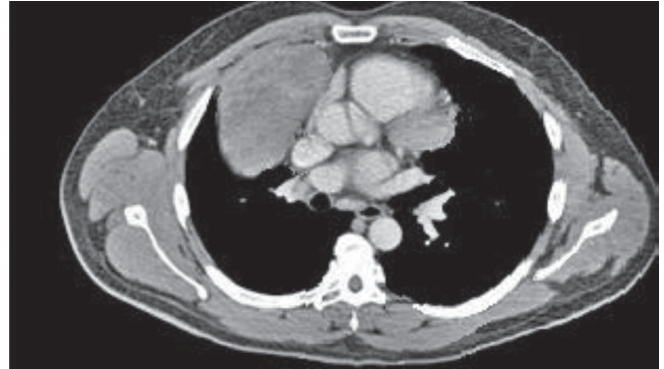
## Case Report

A 49-year-old man was referred to our department complaining of chronic cough. Chest x-ray was done revealed a well-defined lobulated right hilar mass [Figure 1]. Contrast-enhanced computerized tomography (CT) was performed with a 64-detector scanner. On the noncontrast CT images, there was a well-defined heterogeneous anterior mediastinal mass adjacent to the right border of the heart. There was no definite invasion to superior vena cava or right brachiocephalic vein. There was evidence of infiltrations into the anterior mediastinal fat but no mediastinal lymphadenopathy [Figure 2]. After contrast enhancement, the mass showed heterogeneous enhancement [Figure 3].

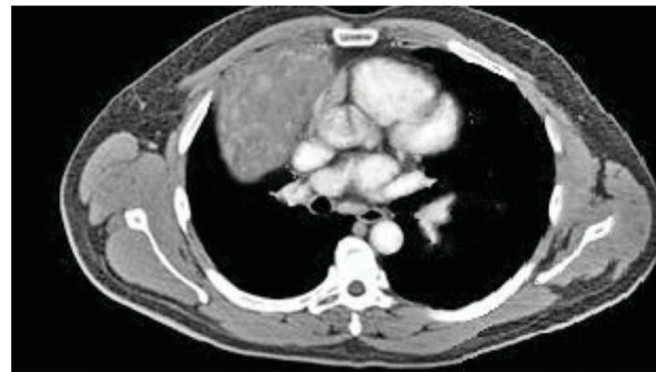
Abdominal ultrasound was done and no significant abnormality was detected. Routine laboratory investigations were done for him, which revealed a white blood cell count of  $11 \times 10^9/L$ , with lymphocyte predominance of 50%; hemoglobin level of 15 g/dL; Hematocrit of 52%; and platelet count of  $260 \times 10^9/L$ . CT guided trucut tissue biopsy and histological analysis of the mass showed that the tumor consists of neoplastic epithelial cells and non-neoplastic lymphocytes. The findings were consistent with invasive lymphoepithelial thymoma. The patient was referred to the oncology center for further management.



**Figure 1:** Anterior-posterior chest x-ray showing a well-defined lobulated right hilar mass of soft tissue density obscuring the right upper border of the heart and great vessels, no pleural effusion.



**Figure 2:** Axial noncontrast computerized tomography scan demonstrating a well-defined soft tissue mass measuring about 6 x 4 cm abutting the right boarder of the heart and the superior vena cava, no lymph nodes.



**Figure 3:** After contrast, the mass showed heterogeneous enhancement.

## Discussion

In spite, thymomas are rare mediastinal tumors, they are the most common neoplasms arising in the thymus. They may be associated with different types of paraneoplastic disorders without clear etiological factors, the most common of which is myasthenia gravis (MG), which is seen in 30%–40% thymoma cases. MG is an autoimmune disorder characterized clinically by weakness and ease of fatigue of skeletal muscles, which improve with rest. The immunopathogenesis of this disorder is well established.<sup>[9]</sup> It affects the neuromuscular junction of voluntary muscle due to interference with acetylcholine receptors. In our case no features of MG noted.

There is great variability in the appearance of the thymus on imaging studies, including features such as size, shape, attenuation on CT, and signal characteristics on magnetic resonance imaging. Due to the variety of imaging appearances, it may be challenging to differentiate the normal from the diseased thymus.<sup>[10,11]</sup> Radiographically, thymoma appears as a soft tissue mass with ill-defined borders and infiltrative

growth into the surrounding structures, mediastinal fat planes, and pleural surfaces. It may invade the trachea, pericardium, heart, and great vessels. Generally, it may not appear on chest x-ray, contrast-enhanced CT is useful in delineating the mass and in defining its vascularity and extent of invasion. Definite diagnosis of thymoma is confirmed by tissue CT-guided trucut biopsy or biopsy taken at anterior mediastinoscopy. In general, when imaging shows a convex margin, focal enlargement of a lobe, or multilobularity in an adult,<sup>[12]</sup> or when the CT attenuation is higher than fat, a mass should be suspected.<sup>[13]</sup> In our case, chest x-ray revealed right hilar mass that appeared as a well-defined mediastinal mass with heterogeneous texture.

The differential diagnosis for an invasive anterior mediastinal mass includes invasive thymoma, thymic carcinoma, lymphoma, metastasis, malignant germ cell tumors, and primary sarcomatous tumors. They show nonspecific appearance. CT features generally cannot help to distinguish them from invasive thymomas.

The prognosis and management of thymoma depends on staging of the mass.

The World Health Organization recently developed a classification system according to the histologic type of thymoma [Table 1], although most reports follow another classification system by Masaoka et al. [Table 2].

WHO classification of thymic epithelial tumors have been shown to reflect their oncological behaviors, and type A, AB, and B1 tumors have better prognosis than type B2 and B3 tumors, suggesting the significance of this classification in the clinical practice of thymomas. Type B tumors are more invasive than type A and AB tumors. Type B1 and B2 tumors are frequently associated with MG while type A and AB tumors are not. The findings of CT imaging revealed that type A and AB tumors tend to be round and have the smooth surface while type B1, B2, and B3 tumors are often flat and have irregular surface. Type AB, B1, and B2 tumors possess a significant number of CD4+CD8+ double positive T-cells in the tumor.

**Table 1:** World Health Organization classification of thymoma

Type	Histologic description
A	Medullary thymoma
AB	Mixed thymoma
B1	Predominantly cortical thymoma
B2	Cortical thymoma
B3	Well-differentiated thymic carcinoma
C	Thymic carcinoma

**Table 2:** Masaoka staging system of thymoma

Stage 1	Encapsulated tumor with no gross or microscopic invasion
Stage 2	Macroscopic invasion into mediastinal fat or pleura
Stage 3	Invasion of pericardium, great vessels, or lung
Stage 4	Pleural or pericardial metastatic spread
Stage 5	Lymphatic or hematogenous spread

These observations are supposed to be useful for preoperative evaluation of WHO classification of thymomas, and to help the clinicians decide application of preoperative therapy and the method of surgical resection including endoscopic surgery.<sup>[14]</sup>

According to stages of thymoma, stage 1 disease can be managed by complete surgical excision, stage 2 and 3 disease requires surgical excision and postoperative radiotherapy, stage 4 and 5 disease requires surgical debulking, radiotherapy, and chemotherapy.

In spite, thymoma is sensitive to both chemotherapy and radiation, the most appropriate treatment of most thymomas is thymectomy, which is usually performed via a median sternotomy.

## Conclusion

To diagnose thymoma radiographically is challenging because it appears in different shapes and sizes with different ages, moreover there are many normal variants of radiological thymic appearance. Thymomas may be diagnosed incidentally at chest imaging, patients may be asymptomatic or present with symptoms due to the presence of an anterior thoracic mass or due to paraneoplastic disorders such as MG. It is important to know the normal spectrum of radiographic appearance of thymus to avoid misdiagnosis of thymoma or any other thymic lesion.

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