

## CASE REPORT OF UNUSUAL PRESENTATION OF INVASIVE THYMOMA IN A 49 YEARS OLD MALE

Dr.Osama Abdalla Mabrouk Kheiralla

### ARTICLE INFO

Received: 27<sup>th</sup> Aug 2015

Revised: 4<sup>th</sup> Nov 2015

Accepted: 14<sup>th</sup> Nov 2015

**Author details:** Assistant Professor of Radiology, Department of Radiology, Faculty of Medicine - University of Najran

**Corresponding author: Osama**  
Assistant Professor of Radiology,  
Department of Radiology, Faculty of  
Medicine - University of Najran  
**Email:** drkheiralla@gmail.com

**Keywords:** Mediastinal tumours,  
Anterior mediastinal masses, Tumours  
of the Thymus, Invasive thymomas

### ABSTRACT

Invasive thymomas are rare tumours in the anterior mediastinum, representing 50% of anterior mediastinal masses and about 20-30% of all mediastinal tumours. 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 tumours is Thymectomy. This case study presents a 49 year-old man with unusual presentation of thymoma. On the non-contrast CT images, there was a well-defined heterogeneous anterior mediastinal mass adjacent to the right border of the heart. There was evidence of infiltrations into the anterior mediastinal fat but no mediastinal lymphadenopathy after contrast enhancement, the mass showed heterogeneous enhancement. CT guided trucut tissue biopsy and histological analysis of the mass showed that the tumour consists of neoplastic epithelial cells and non-neoplastic lymphocytes. The findings were consistent with invasive lymphoepithelial thymoma.

### INTRODUCTION

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 centre. Thymus originates in the embryo from the ventral ring of 3rd and 4th pharyngeal pouches and ectoderm endoderm of the cervical sinus, as epithelial outgrowths on each side [1, 2, 3]. The thymus is responsible for the processing and maturation of T-lymphocytes. 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 [4]. After birth, the thymus starts to grow to reach its maximum weight of 40 grams around puberty, and then it involutes and persists in an atrophic state into old age.

Thymomas are rare neoplasms arising from tissue elements of the thymus and developing in the anterior mediastinum [5, 6]. They can be associated with a variety of systemic and autoimmune disorders, such as pure red cell aplasia, pancytopenia, hypogammaglobulinemia, collagen-vascular disease, and most commonly with myasthenia gravis [5, 7, and 8]. Thymomas are uncommon tumors with an annual incidence of only 0.15 cases per 100,000 person-years [5, 9], yet representing the most frequently diagnosed tumor of the anterior mediastinum [5, 10].

Several classification systems of thymomas have been developed and described. However, clinical, pathologic, and surgical classification of thymomas remains controversial. The histomorphologic variability and

heterogeneity of cells within thymomas is a major factor guiding this intense debate [11, 12].

Currently, computed tomography is the first choice technique to characterize a mediastinal mass with regard to its anatomic dissemination and invasion of neighboring structures, as well as possible distant metastases [5]. Applying computed tomography, thymomas can often be distinguished from benign mediastinal lesions or from lymphoma in the case of multiple mediastinal abnormalities [13]. Magnetic resonance imaging is frequently implemented in the radiologic diagnosis of thymomas [14]. The major role of the magnetic resonance image seems to lie in its value for surgical planning, especially if resection of thymomas is considered that invade neighboring structures such as the great vessels or the heart [5].

As the only curative treatment, surgery remains the baseline attempt in thymoma therapy. Complete or partial median sternotomy with complete thymectomy is the operative approach of choice [15-19].

### 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 liar mass (**Fig.1**). Contrast enhanced Computerized tomography was performed with a 64-detector scanner. On the non-contrast 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 (Fig.2). After contrast enhancement, the mass showed heterogeneous enhancement (Fig.3).



**Fig 1: AP 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.**



**Fig 2: Axial Non contrast CT scan demonstrating a well-defined soft tissue mass measuring about 6X4 cm abutting the right boarder of the heart and the superior vena cava, no lymph nodes.**



**Fig 3: After contrast, the mass showed heterogeneous enhancement**

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%; haemoglobin level of 15 g/dl; Haematocrit is 52%; and platelet count of  $260 \times 10^9/L$ . CT guided trucut tissue biopsy and histological analysis of the mass showed that the tumour consists of

neoplastic epithelial cells and non-neoplastic lymphocytes. The findings were consistent with invasive lymphoepithelial thymoma. The patient was referred to the oncology centre for further management.

## DISCUSSION

Tumours of the thymus are among the rarest human neoplasms, comprising <1% of all adult cancers, with an incidence rate of 1–5 / million population / year. Thymomas are the most frequent thymic tumours in adults, followed by mediastinal lymphomas, some of which arise from mediastinal lymph nodes. In children, the mediastinum is the site of 1% of all tumours; most common are non-Hodgkin lymphomas, while thymomas are extremely rare [20].

Thymomas and thymic carcinomas are uncommon tumours with an annual incidence of approximately 1-5 per million population [21]. Thymoma is the most common neoplasms arising in the thymus originating from epithelial cells of thymus, it accounts for about 25% of all mediastinal tumours with a peak incidence between 40 and 50 years of age.

Patients with thymoma are often clinically asymptomatic [22]. However, it may present with local symptoms related to encroachment on adjacent structures, as cough, chest pain or superior vena cava syndrome. The symptomatic patients may have only local symptoms related to the presence of the tumor within the mediastinum or only symptoms related to systemic disease states that are frequently associated with the presence of thymoma or a combination of both [22]. In case of disseminated disease, the most common manifestations are pleural or pericardial effusions, which may be associated with thoracic symptoms.

Thymoma may be associated with different types of paraneoplastic disorders without clear etiological factors, the most common of which is myasthenia gravis, which is seen in 30 to 40 % thymoma cases. Myasthenia gravis is an autoimmune disease affecting the neuromuscular junction of voluntary muscle due to interference with acetylcholine receptors. Myasthenia gravis (MG) is the most common PTS encountered [22, 23, 24, 25]. This syndrome (MG) is present in approximately 30 to 59% of patients with thymoma [24, 25].

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 fine-needle aspiration. A fine-needle aspiration (FNA) biopsy is an accepted and feasible method to differentiate mediastinal lesions and to diagnose or classify thymomas histopathologically [26, 27].

The differential diagnosis for an invasive anterior mediastinal mass includes invasive thymoma, thymic carcinoma, lymphoma, metastasis, malignant germ cell tumours and primary sarcomatous tumors. They show

non-specific 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), the World Health Organization (WHO) histologic typing of tumors of the thymus (1999), based on cytologic similarities between normal thymic epithelial cells and neoplastic cells [28, 29]. although most reports follow another classification system by Masaoka et al (Table 2). The most widely used staging system acknowledging the presence of invasion and anatomic extent of involvement, both clinically and histopathologically, was defined by Masaoka and colleagues [30].

**Table 1: Who 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 tumour 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 haematogenous spread

According to stages of Thymoma, In 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

Thymomas and thymic carcinomas are rare anterior mediastinal tumours. 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 myasthenia gravis. The prognosis of thymoma or thymic carcinoma depends upon the stage of disease and the histologic type of the tumour. Thymectomy is the initial treatment for all patients with a thymoma

**Conflict of interest:** Nil

## REFERENCES

1. Bharathi M, Giri PM, Shilpa K, Geetanjali S. A rare case report of Ectopic Cervical Thymoma. *IAIM*. 2015; 2(3): 175-178.
2. Thakur A, Sebag F, Micco CD, Slotema E, Henry FJ. Ectopic cervical thymoma mimicking as papillary thyroid carcinoma: A diagnostic dilemma. *Indian J Pathol Microbiol*, 2010; 53: 305-7
3. Ramdas A, Jacob SE, Varghese RG, Dasiah S, Rai R. Ectopic cervical thymoma - the great mimic: A case report. *Indian J Pathol Microbiol*, 2007; 50(3): 553-5.
4. Jung JI, Kim HH, Park SH, Lee YS. Malignant Ectopic Thymoma in the Neck: A Case Report. *Am J Neuroradiol* 1999; 20: 1747-1749.
5. Sandra Tomaszek, Dennis A. Wigle, Shaf Keshavjee, Stefan Fischer. Thymomas: Review of Current Clinical Practice: *Ann Thorac Surg*. 2009; 87(6): 1973-1980.
6. Cowen D, Richaud P, Mornex F. Thymoma: results of a multicentric retrospective series of 149 non-metastatic irradiated patients and review of the literature. *FNCLCC trialists. Fédération Nationale des Centres de Lutte Contre le Cancer Radiother Oncol*. 1995;34:9-16.
7. Hon C, Chui WH, Cheng LC, Shek TW, Jones BM, Au WY. Thymoma associated with keratoconjunctivitis, lichen planus, hypogammaglobinemia, and absent circulating B cells. *J Clin Oncol*. 2006;24:2960-1.
8. Skeie GO, Apostolski S, Evoli A. Guidelines for the treatment of autoimmune neuromuscular transmission disorders. *Eur J Neurol*. 2006;13:691-9
9. Engels EA, Pfeiffer RM. Malignant thymoma in the United States: demographic patterns in incidence and associations with subsequent malignancies. *Int J Cancer*. 2003;105:546-51.
10. Chen G, Marx A, Wen-Hu C. New WHO histologic classification predicts prognosis of thymic epithelial tumors: a clinicopathologic study of 200 thymoma cases from China. *Cancer*. 2002;95:420-9.
11. Harris NL, Müller-Hermelink HK. Thymoma classification: a siren's song of simplicity. *Am J Clin Pathol*. 1999;112:299-303
12. Kuo TT. Classification of thymic epithelial neoplasms: a controversial issue coming to an end? *J Cell Mol Med*. 2001;5:442-8
13. Maher MM, Shepard JA. Imaging of thymoma. *Semin Thorac Cardiovasc Surg*. 2005;17:12-9
14. Tomiyama N, Honda O, Tsubamoto M, et al. Anterior mediastinal tumors: Diagnostic accuracy of CT and MRI. *Eur J Radiol*. 2009;69:280-8
15. Evoli A, Minisci C, Di Schino C, et al. Thymoma in patients with MG: characteristics and long-term outcome. *Neurology*. 2002;59:1844-50
16. Kim DJ, Yang WI, Choi SS, Kim KD, Chung KY. Prognostic and clinical relevance of the World Health Organization schema for the classification of thymic epithelial tumors: a clinicopathologic study of 108

- patients and literature review. *Chest*. 2005;127:755–61.
17. Rea F, Marulli G, Girardi R, et al. Long-term survival and prognostic factors in thymic epithelial tumours. *Eur J Cardiothorac Surg*. 2004;26:412–8
  18. Haniuda M, Kondo R, Numanami H, Makiuchi A, Machida E, Amano J. Recurrence of thymoma: clinicopathological features, re-operation, and outcome. *J Surg Oncol*. 2001;78:183–8.
  19. Zhu G, He S, Fu X, Jiang G, Liu T. Radiotherapy and prognostic factors for thymoma: a retrospective study of 175 patients. *Int J Radiat Oncol Biol Phys*. 2004;60:1113–9.
  20. Juanpere S, Cañete N, Ortuño P, Martínez S, Sanchez G. A diagnostic approach to the mediastinal masses. *Insights Imaging*. 2013;4: 29-52.
  21. Müller-Hermelink HK, Engel P, Kuo TT, Ströbel Ph, Marx A, Harris NL, Möller P, Menestrina F, Shimosato Y, Asamura H, Masaoka A, Sobin LH: World Health Organisation Histological Classification of Tumours. *Tumours of the Thymus: Introduction*. IARC Press, Lyon; 2004.
  22. Shields TW. *Thymic Tumors. General Thoracic Surgery*, 5th ed. Wolters Klumer Company; 2000. pp.2181.
  23. Kazuyo K, Yasumasa, Monden. Thymoma and myasthenia gravis: a clinical study of 1089 patients from Japan. *Ann Thorac Surg* 1978; 47:339,
  24. Maggi G. Thymoma: results of 241 operated cases. *Ann Thorac Surg* 1991; 51:152.
  25. Masaoka A, et al. Thymoma associated with purered cell aplasia. *Cancer* 1989; 64:1872.
  26. Ali SZ, Erozan YS. Thymoma. Cytopathologic features and differential diagnosis on fine needle aspiration. *Acta Cytol*. 1998;42:845–54
  27. Shin HJ, Katz RL. Thymic neoplasia as represented by fine needle aspiration biopsy of anterior mediastinal masses. A practical approach to the differential diagnosis. *Acta Cytol*. 1998;42:855–64
  28. Müller-Hermelink HK, Ströbel P, Zettl A. Combined thymic epithelial tumours. In: Travis WD, Brambilla E, Müller-Hermelink HK, Harris CC, editors. *Pathology and genetics of tumours of the lung, pleura, thymus and heart (WHO classification of tumours series)* Lyon, France: IARC Press; 2004. pp. 196–8.
  29. Travis WD, Brambilla E, Müller-Hermelink HK, Harris CC, editors. *Pathology and genetics of tumours of the lung, pleura, thymus and heart (WHO classification of tumours series)* Lyon, France: IARC Press; 2004. pp. 152–171.
  30. Masaoka A, Monden Y, Nakahara K, Tanioka T. Follow-up study of thymomas with special reference to their clinical stages. *Cancer*. 1981;48:2485–92