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Case Report

Huge Cervico-Thoracic Thymoma Associated with Liver Metastasis: A Rare Case Report

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Abstract

Thymomas are rare tumors characterized by their slow growth and their ability to directly invade by contiguity. Although distant dissemination is unusual, all thymoma subtypes have the ability to metastasize to extra thoracic organs. We report the case of a patient with a huge thymic B₂ cervico-mediastinal tumor associated with a concomitant liver metastasis. Despite the size and the significant extension of the tumor; the clinical symptoms are severe and medical treatment combining radio-chemotherapy has been able to stabilize the lesions.

Abbreviations: TET: Thymic Epithelial Tumors; WHO :World Health Organization

Introduction

Thymic Epithelial Tumors (TET) are rare, their incidence is estimated at 0.15 cases per 100,000 people/year in the United States [1] but they represent the most frequent etiology of tumors of the anterior mediastinum of adults; however, giant intra-thoracic forms represent only 4% of cases [2]. Slow development explains that the disease is frequently discovered at an advanced stage. About 70% of patients with thymomas remain asymptomatic; the other patients may present local symptoms related to a tumor developing on surrounding structures such as cough, chest pain, upper vena cava syndrome and dysphagia. Only 30% of patients with thymoma have a clinic related to severe myasthenia. Another 5% of patients have other systemic syndromes, including red blood cell aplasia, dermatomyositis, systemic lupus erythematosus, Cushing's syndrome and syndrome of inappropriate secretion of antidiuretic hormones [3].

Case Report

We describe the case of a 58-year-old man, consultant for a median cervical mass measuring 5cm in height and 4cm in width, fixed, of firm consistency (Figure 1) evolving for 5 years and gradually increasing in volume plunging into the mediastinum, its lower limit is not palpable, with no compressive signs as dyspnoea, dysphagia or dysphonia diagnosed as a goiter.



Figure 1: Anterior Cervical Mass plunging into the Mediastinum viewed from the front and in profile.

The cervico-thoracic CT scan (Figure 2) shows a well-limited heterogeneous cervico-thoracic tissue density process measuring 70mm in anteroposterior diameter, 83mm in transverse diameter and extending over 16.5cm in height; it is lateralized to the right with median and right extension, seems to push back the left thyroid lobe, at the cervical level it pushes back the visceral axis towards the right, on the anterior mediastinal stage it pushes back the arch of the aorta, laterally the supra aortic trunks. Filling of the right piriform sinus with right with thickening of the ary epi glottic folds. The study of bone windows does not show suspicious bone lesion.



Figure 2: Well limited cervical-thoracic seat tissue density process enhanced heterogeneously; lateralized to the right with median and right extension, appears behind the left thyroid lobe.

We note a hypodense lesion of the hepatic dome measuring 27mm, probably secondary metastasis (Figure 3).

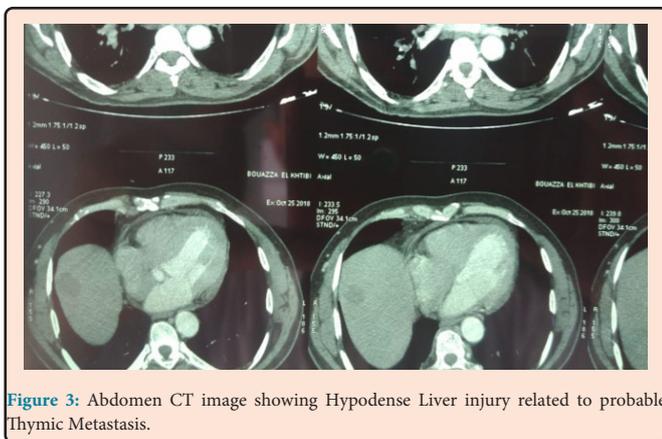


Figure 3: Abdomen CT image showing Hypodense Liver injury related to probable Thymic Metastasis.

Patient had a pan endoscopy without abnormalities, and the biopsy of the cervical mass was done under local anesthesia was in favor of a thymoma type B₂, the same histological type was found on a trans parietal liver biopsy of the suspect lesion therefore concluding with hepatic metastasis. After a multidisciplinary discussion concluding that the tumor extension does not allow complete resection as a first step; neo-adjuvant chemotherapy with multimodal strategy associated with corticosteroid therapy and radiotherapy at a dose of 50 Gy was started with regular reassessment. The patient has been followed for 2 years; he is currently experiencing a regression of cervical swelling with stabilization of the thoracic and hepatic mass.

Discussion

Thymomas are tumors which arise from epithelial cells of the thymus, they represent the histological type most frequently found in the anterior mediastinum, around 15 to 20% of all primitive mediastinal masses [4]. Patients with this tumor are often aged 50 to 60 years rarely before 20 years with a slight male predominance; no risk factor has been found, but an association with severe myasthenia or systemic disease has been described [3]. These tumor masses are characterized by their slow growth and their ability to invade adjacent organs by contiguity. On the other hand, the appearance of distant metastases is less frequent and, in the majority of cases, it is linked to thymic carcinomas [5]. Recently, the World Health Organization (WHO) has reclassified thymic epithelial tumors type A, AB, B₁, B₂, B₃ and thymic carcinomas according to the predominance of the epithelial or lymphocytic component (Table 1). In this classification, the different subtypes of thymic tumors are classified in ascending order of malignant tumors, although it has been shown that all, and not only carcinomas, have the capacity to metastasize [6]. But there exist combined forms associating several histological types, this classification has a prognostic interest thus the stage of extension and the risk of recurrence after excision increases between types A/AB, B₁, B₂ and B₃ [7].

Table 1: Classification of Thymomas (WHO 2004).

Types	Histological Features
A	A tumor composed of a population of neoplastic thymic epithelial cells having spindle/oval shape, lacking nuclear atypia, and accompanied by few or no non neoplastic lymphocytes
AB	A tumor in which foci having the features of type A thymoma are admixed with foci rich in lymphocytes
B ₁	A tumor that resembles the normal functional thymus in that it combines large expanses having an appearance practically indistinguishable from normal thymic cortex with areas resembling thymic medulla
B ₂	A tumor in which the neoplastic epithelial component appears as scattered plump cells with vesicular nuclei and distinct nucleoli among a heavy population of lymphocytes. Perivascular spaces are common and sometimes very prominent. A perivascular arrangement of tumor cells resulting in a palisading effect may be seen
B ₃	A type of thymoma predominantly composed of epithelial cells having a round or polygonal shape and exhibiting no or mild atypia. They are admixed with a minor component of lymphocytes, resulting in a sheet like growth of the neoplastic epithelial cells
C	A thymic tumor exhibiting clear-cut cytologic stypia and a set of cytoarchitectural features no longer specific to the thymus, but rather analogous to those seen in carcinomas of other organs. Type C thymomas lack immature lymphocytes; whatever lymphocytes may be present are mature and usually admixed with plasma cells

There is no official TNM classification and we often refer to Masaoka's TNM classification [8] (Table 2).

Table 2: Masaoka classification and prognostic Implication.

Stages	Definition	5-year survival	10-year survival
I	Encapsulated tumor without microscopic invasion	83-100%	86-100%
II	a) Macroscopic invasion of mediastinal cellular or mediastinal pleura b) Microscopic capsular invasion	70-100%	55-100%
III	macroscopic invasion of neighboring organs	46-70%	47-60%
IV	a) pleural or pericardial involvement b) distant metastases	11-75%	0-11%

Thus thymomas do not have a capsule, and can disseminate by lymphatic or blood route and give metastasis at a distance. Hepatic metastases although they are very rare but constitutes the 2nd extra thoracic localization. The best treatment in limited and extirpable forms is surgical excision of the entire tumor, especially in localized and encapsulated forms [9]. The conventional approach to TET is the median sternotomy; in the event of partial resection, multimodal treatment starting with chemotherapy and neoadjuvant radiotherapy is strongly recommended [9]. Extensive and inoperable forms immediately stage III and IV should be the subject of a multimodal approach if possible preoperative including poly chemotherapy; several protocols have been described, but cisplatin appears to be a pivotal drug in chemotherapy; the most often studied includes cyclophosphamide, doxorubicin and cisplatin to which are sometimes added prednisone which seems to give higher response rates by their direct cytotoxic effects on the normal lymphoid population; neoadjuvant radiotherapy may also be indicated at a dose of 24 to 30 Gy to improve the prognosis and facilitate tumor resectability [9].



Conclusion

Even if thymomas are considered slow growing tumors with predominantly local recurrences, the existence of distant metastases of any subtype of thymoma should require continuous monitoring for all patients with a history of thymoma. These tumors are sensitive to chemotherapy and radiotherapy, but complete surgical excision is the preferred treatment whenever it is technically possible.

Competing Interests

The authors declare no competing interest.

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