

CASE REPORT

Invasive thymoma

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Abstract

Thymomas are rare tumors of the thymic epithelium with a broad spectrum of morphological and clinical features. Despite a benign histological appearance, it can invade nearby structures or metastasize. The majority of patients are asymptomatic, but some may present with paraneoplastic syndromes such as myasthenia gravis, red cell aplasia, or hypogammaglobulinemia. Various staging systems of thymomas have been defined based on the degree of invasiveness. According to the WHO Classification, there are six histologic types of thymic epithelial tumors. The most important prognostic factor is the stage according to Masaoka's system. We report a case of invasive thymoma diagnosed incidentally in a patient with cardiovascular risk factors referred to the cardiologist with suspected thoracic aorta aneurism.

Keywords: thymoma, mediastinum, pericardial invasion, echocardiography.

□ Introduction

Thymomas and thymic carcinomas are rare tumors of the mediastinum. Although a vast majority of thymomas are located in the anterior mediastinum, there have been many sites of ectopic localization described. Ectopic thymomas were found in the superior and the posterior mediastinum, but also in rare locations: at the base of the skull [1], intrapericardially [2], in the lung parenchyma [3] and in the pleural cavity [4].

□ Patient and Methods

A 48-year-old patient, obese, habitual smoker (20 cigarettes per day for 20 years), diagnosed with arterial hypertension (maximum 190/120 mmHg), not treated, was referred to Craiova Cardiology Center by the family physician after a pulmonary radiography revealed an enlarged mediastinum with a suspected thoracic aortic aneurism.

The patient presented dyspnea and fatigue to usual physical activity; the symptoms have progressively worsened during the last weeks. The clinical exam did not show significant abnormalities except the high blood pressure (170/100 mmHg).

Transthoracic echocardiography found slightly thickened left ventricle walls, normal valve structure and function, a first-degree diastolic dysfunction with normal systolic function of the left ventricle. The thoracic aorta could not be viewed clearly, because the patient was obese. The next step for the exclusion of an aortic aneurism is the transesophageal echocardiography, which showed a normal thoracic aorta.

It was decided to repeat the pulmonary radiography, which demonstrated the presence of a homogenous,

clearly delineated opacity, of approximately 8 cm, located leftward, near the hilum.

Further imaging investigations were needed to establish the diagnosis.

The computed tomography showed a lobulated mass, size 3.29/2.16 cm, associated to some adjacent fibrotic constituents, tractionary on the visceral pleura, visible on anterior segments of the left inferior lobe. It was associated to a sizeable adenopathic block, 8.91/9.63 cm, located in the prevascular space, with similar adenopathies, size 2.63 cm, leftward and paratracheal, and nonspecific lymph nodes located in the aorto-pulmonary window. There was a small quantity of pericardial fluid, mostly anterior. The radiologist concluded it was a pulmonary tumor stage T2N2Mx (Figures 1 and 2, a and b).



Figure 1 – A-P Pulmonary digital radiography – enlarged asymmetrical superior mediastinum especially in the left side, with polycyclic, lobulated contours.

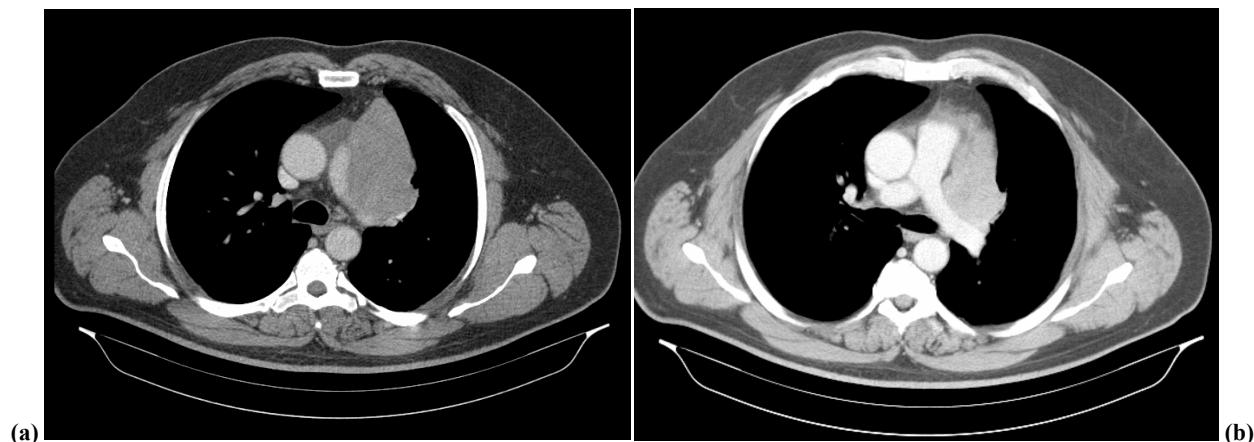


Figure 2 – (a, b) CT thoracic scan – with contrast i.v. iodinated medium – prevascular heterogeneous mass in the prevascular space, that lies near left pulmonary artery, associated with other mediastinal adenopathies; due to the presence of pulmonary fibrosis and atelectasis it was considered to be pulmonary tumor T2, N2, Mx; there was no sign of pleural and pericardial invasion in time of diagnosis.

The magnetic resonance imaging exam described a mediastinal mass, probably an adenopathic, necrotized block, in hyposignal T1, in hypersignal T2, heterogeneous, with size 7.9/9.34 cm, which entails the leftward superior and inferior prevascular and paratracheal groups. There was an adenopathic block that flanks the aortic crosa, on an over 180° sectorial segment and a distance of 6.91 cm, flanking as well the left pulmonary artery

without any sign of a definite invasion. There was no focal parietal distortion, of endoluminal formation. This sizeable adenopathic block is associated to nonspecific lymph nodes located in aorto-pulmonary window and leftward superior and inferior prevascular and paratracheal spaces, and subcarinal adenopathies, size 2.82 cm, as well as nonspecific subclavicular adenopathies, left hilar lymph nodes – 2.7 cm (Figure 3, a and b).

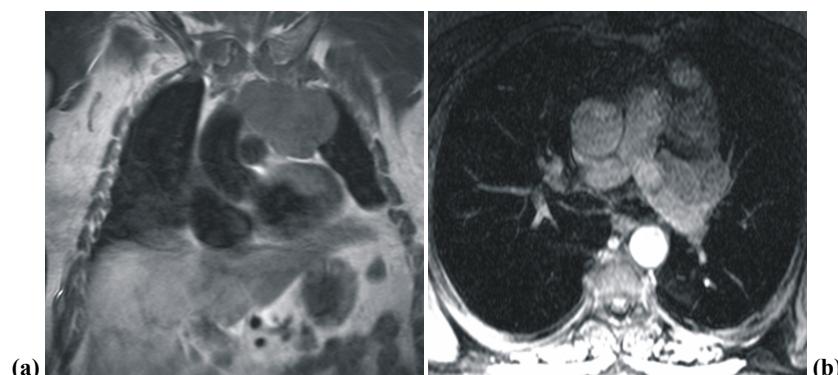


Figure 3 – Coronal T1 (a) and axial T1 postcontrast (b) IRM scans – lobulated prevascular thymic mass with heterogeneous hyposignal in T1 and T1+gado scans with the same characters associated with mediastinal adenopathy.

Results

The patient was referred for surgical intervention during which it was found a tumor formation located in the anterior mediastinum, invading the pericardium as well as the pulmonary artery and vein intrapericardially. The tumor sent for histological examination consisted of an 8/4/4.5 cm fragment, capsulated, with white color, relatively stern density, with residual tissue presenting adipose degeneration.

Microscopically it was found to be a well differentiated typical carcinoma – B3 thymoma (*WHO Classification*), consisting in a proliferation of the neoplastic epithelial cells having nuclear abnormalities and solid pattern, located in a fibrocollagenic stroma, mixed with a discrete residual lymphocytic population.

The postoperative course was uneventful; the patient was asymptomatic at one-month follow-up.

Discussion

Various staging systems of thymomas have been

defined based on the degree of invasiveness. The most common system is that of Masaoka A et al. [5]. Stage I (completely encapsulated tumor) corresponds to the benign form of thymoma, which rarely recurs; 70 to 80% of thymomas fall into this category. Stages II through IV are divided into two types. Type I tumors, also called malignant thymomas, have all the clinical, morphologic, and cytologic attributes of encapsulated thymoma but exhibit local invasion or distant metastases. Type II tumors, also called thymic carcinomas, have the cytological attributes characteristic of malignancy. These are very aggressive neoplasms, and local invasion and distant metastases are common.

A *WHO Classification* was introduced in 1999, which has been updated in 2004 [6]. Retrospective studies have shown the prognostic significance of this classification together with Masaoka's staging system and the extent of surgery.

Thymomas are slow growing tumors that tend to recur locally but are unlikely to metastasize hematogenously or to regional lymphatics. Currently, the most

important prognostic factors are disease stage according to the Masaoka's staging system, the *WHO Thymoma Classification*, and complete surgical resection [7–9].

In a study of 108 cases the 5-year survival rates of type A, AB, B1, B2 and B3 thymoma cases were 100%, 100%, 93%, 83% and 43%, respectively; while the 10-year survival rates were 100%, 100%, 81%, 70% and 33%, respectively. The median survival time of type C thymoma was 62.5 months. Type B2 and B3 thymoma cases had an intermediate prognostic ranking in comparison with type C thymoma and other groups [10].

The B3 thymoma of the *WHO Classification* are invasive in 85% and recurrent in 29% of cases, while the average stage at presentation is 2.3 with 91% of complete resections and 20-year survival in 36% of cases [11].

The expected survival of a B3 thymoma patient with pericardial extension is difficult, if not impossible, to determine, but presumably poor.

The management of most thymomas is mainly based on surgical resection [12, 13].

In advanced thymomas, Masaoka's stage III–IV, chemotherapy achieves major responses (complete and partial) in the range of 77% of cases, thereby increasing the resectability and improving survival [14].

Radiation therapy is indicated for patients with unresectable or residual disease after surgery and is sometimes used as part of an induction regimen. Multimodality treatment in locally advanced thymomas increases the survival and may sometimes be curative.

Conclusions

Although thymomas are rare they should be always taken into consideration when there is a patient presenting with an enlarged mediastinum. In conjunction with history and physical exam, imaging techniques, including echocardiography, are essential in the diagnosis and pre-operative evaluation of thymomas.

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