

Case Report

Intrathoracic goiter and invasive thymoma: rare concomitant presentation*

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ABSTRACT

We present a rare situation in which two mediastinal tumors of different topology and histology were found during the resection of an extensive mediastinal tumor in an asymptomatic patient. Different histologies within the same mass have been reported, although, to our knowledge, there have been no reports of different tumors at distinct locations. Thymomas and intrathoracic goiters account for a large proportion of the tumors found in the mediastinum. When feasible, surgical resection plays a fundamental role in effecting a cure. In order to identify concomitant lesions and perform a complete resection, detailed surgical exploration is required.

Keywords: Thymoma/complications; Mediastinal neoplasms/complications; Goite, substernal; Mediastinum/pathology; Case reports [publication type]

INTRODUCTION

Primary mediastinal neoplasias are rare, corresponding to less than 10% of all thoracic neoplasias.⁽¹⁾ Most of these tumors affect only one compartment, although they can spread to adjacent structures as the tumor grows. The anterosuperior, posterior and medial compartments are affected, respectively, in 50%, 25% and 20% of the cases. Approximately 25% of all tumors are malignant.

Thymomas represent an uncommon group of neoplasias that arise from thymus cells and are known by their association with myasthenia gravis, their

variable histology and their heterogeneous malignant behavior.⁽²⁾ Substernal goiter is the most common tumor in the superior mediastinum, appearing either as an extension of the cervical stroma, totally intrathoracic, or as ectopic thyroid tissue.⁽³⁾

The aim of this study was to present a rare situation in which, during resection of a large goiter, we found an invasive thymoma. We will also present a brief review of the literature on these tumors.

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CASE REPORT

A 70-year-old female patient was referred to the Department of Thoracic Surgery of the AC Camargo Cancer Hospital after radiographic imaging led to a diagnosis of mediastinal widening. Ten years prior, she had been submitted to right hemithyroidectomy due to a benign thyroid nodule. The results of the physical examination were normal, as were those of the laboratory analysis, including the analysis of the thyroid hormones. A computed tomography scan of the chest showed a huge mass that was in close contact with the mediastinal structures (Figure 1), especially with the superior vena cava. Due to the possibility of vascular invasion, a magnetic resonance imaging of the chest was conducted, excluding invasion of the superior vena cava.

The patient was submitted to right lateral thoracotomy, with general anesthesia and selective intubation, and the thoracic cavity was accessed via the fourth intercostal space. The examination of the cavity revealed a huge mass that was lobulated and hypervascularized, spreading from the superior mediastinum to the pericardium and compressing the superior vena cava (Figure 2). Resection of the superior vena cava adhesions was quite difficult, especially in the case of those near the pericardium. The mass did not spread to the superior thoracic aperture, and adhesions to mediastinal structures at this point were therefore easily resected. During the exploration of the mediastinum, another whitish nodular lesion of approximately 3 cm was observed in the thymic space close to the pericardial fat which, apparently, was not invaded. The appearance of the frozen biopsy sample was consistent with a thymoma (Figure 3). During postoperative recovery there were no complications, and the patient was discharged from the hospital in a good clinical state on post-operative day 4.

Anatomopathologic examination confirmed the presence of a colloid goiter, and the smallest lesion proved to be a thymoma in which the capsule had been invaded, although not breached.

After six months of follow-up treatment, the general health status of the patient was good, and she presented no signs of relapse or respiratory symptoms. As in all cases of Masaoka stage IIB thymoma,⁽⁴⁾ she was treated with adjuvant radiotherapy.

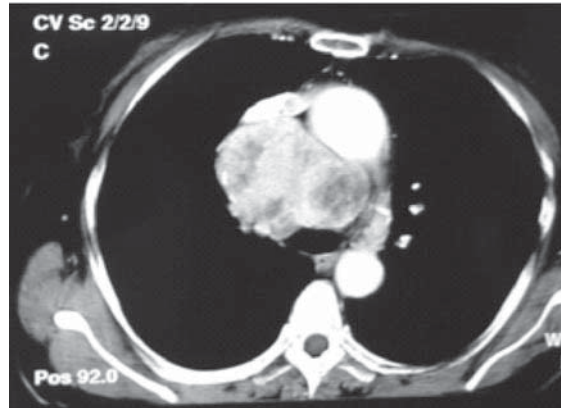


Figure 1 - Mediastinal mass compressing the superior vena cava

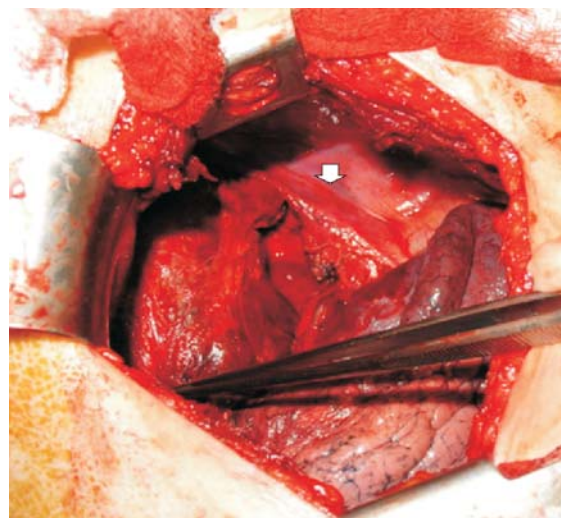


Figure 2 - The mass in relation to the thoracic structures

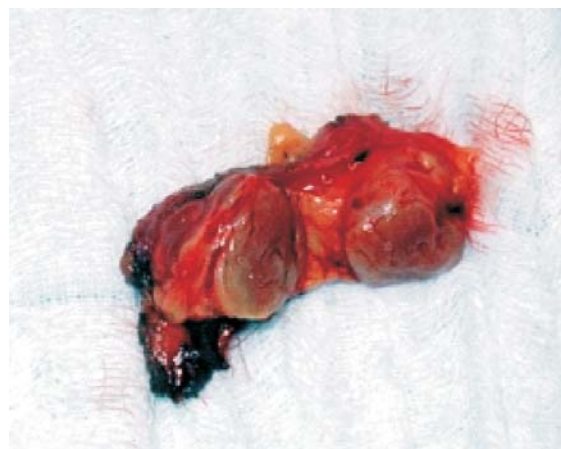


Figure 3 - Mediastinal nodule - thymoma

DISCUSSION

Tumors of different histologies in the same neoplastic mass have previously been reported. However, tumors of different lineages located in separate topographies of the mediastinum are rare. After exhaustive research, we could not find a similar situation in the medical literature.

Intrathoracic goiters are part of a group of endocrine tumors that present a relatively high incidence, accounting for 10% to 15% of all mediastinal masses.⁽³⁻⁴⁾

They are classified according to whether they extend from the thyroid into the thorax (cervicomedial goiter) or are confined to the chest, with no palpable cervical goiter (intrathoracic thyroid goiter), the latter being the case in this patient. The thyroid gland can migrate into the chest as a result of any of the following: an increase in its own weight; negative intrathoracic pressure, respiratory movements; and contraction of the neck muscles. In a series of published articles, 3% to 30% of the patients (22% of all cases) had undergone thyroidectomy prior to the study outset.⁽⁵⁻⁶⁾

Intrathoracic goiters are typically located in the anterosuperior mediastinum, although they can also be found in the medial or posterior compartments. Occasionally (in 40% of all cases), intrathoracic goiters are diagnosed in routine examinations. However, they can induce symptoms of airway compression or even evolve to superior vena cava syndrome. Chest X-rays and computed tomography scans usually provide enough information to determine the location and extent of the lesion. Magnetic resonance imaging of the chest can be used when there is suspicion of vascular invasion. The type of access should be chosen based on the position and extent of the goiter. An intrathoracic goiter can be completely removed through high posterolateral thoracotomy. However, if the situation calls for additional exploration of the mediastinum, in which the major vessels will be manipulated, sternotomy is the incision of choice.

Thymomas constitute an interesting area of study for specialists, and there is no lack of discussion on this subject. Due to their low incidence and wide histological spectrum, the histological classification and staging of thymomas have been the object of controversy. The most widely accepted staging method was proposed in 1981 and is based on

microscopic and macroscopic evaluation of the extent to which the mediastinal structures have been invaded.⁽⁶⁻⁷⁾

In 1991, the World Health Organization announced a new system of histological classification, in which thymic epithelial tumors are classified as type A, AB, B1, B2, B3 or C, the tumor type being accepted as an important prognostic factor.⁽⁸⁾

Some authors have shown that the five-year survival rate for patients presenting large masses (> 11 cm) is 58%, compared with 84% among patients with smaller tumors (5 to 11 cm).⁽⁹⁾

Thymomas can be accompanied by any of a variety of syndromes, usually auto-immune disorders. Myasthenia gravis is the most common, occurring in approximately 45% (range, 10%-67%) of all patients with thymoma.⁽¹⁰⁾

It affects males and females equally, and the incidence peaks in individuals between 30 and 40 years of age.⁽¹¹⁻¹²⁾

Approximately one-third of all patients with thymoma are asymptomatic. Among symptomatic patients, 40% present local symptoms related to the intrathoracic mass.

In most cases (90 to 95%), the tumor is localized, and surgical resection is therefore the treatment of choice.⁽¹²⁾

Complete resection is possible in nearly 100% of all patients with stage I thymoma, whereas there is a wide spectrum of resectability in patients presenting the more advanced tumor stages. Complete resection is the most significant prognostic factor, and disease-free survival is the best way to evaluate the efficacy of the surgical treatment.⁽¹⁰⁻¹²⁾

In a study involving 104 patients, of which 85.6% were submitted to complete resection and 25% received adjuvant treatment, 2 patients were treated with chemotherapy, 19 with radiotherapy and 5 received a combination of the two treatments.⁽¹³⁾

In another study, the authors evaluated a sample of 100 patients and found the ten-year survival rates to be 92%, 87%, 60% and 35%, respectively, for those with stage I, II, III and IV tumors.⁽¹⁴⁾

The mean interval of recurrence for patients with stage I tumors is ten years, compared with five years for patients with stage II, III or IV tumors. In one case, a 32-year postresection interval of recurrence was reported.⁽¹⁴⁾

The most common type is that in which there is dissemination to the pleura and pericardium,

followed by that in which the tumor growth is local and by that in which there are lung and remote (liver and bone) metastases. There is some debate regarding the benefits of subtotal resection. However, it is possible to increase survival when there is minimum residual disease, especially when the adjuvant therapy is taken into consideration.⁽¹⁵⁾

Adjuvant radiotherapy in patients submitted to complete resection remains a subject of discussion. Some authors recommend adjuvant radiotherapy for all patients submitted to surgery; others recommend it only for patients in stages II and III, as well as for patients submitted to incomplete resection. Comparing the results of several retrospective series, it has been observed that the relapse rate in stage I is so low that radiotherapy is unnecessary. In patients with stage II or III tumors submitted to complete resection, radiotherapy remains controversial. However, it has proven efficient in patients with stage III or IV tumors submitted to incomplete resection.⁽¹⁴⁻¹⁵⁾

Thymomas are sensitive to chemotherapy, responding objectively, on average, in two-thirds of all patients (range, 10%-100%) and completely in one-third of all patients (0-43%). Another important question is the heterogeneous prognosis for patients with stage III thymoma. Tumors presenting discrete invasion into the pericardium are considered stage III tumors, can be easily resected and present favorable prognoses. However, large masses that invade the lungs and major vessels are also considered stage III tumors, despite the fact that the prognosis for patients with such tumors is unfavorable.⁽¹⁶⁾

Somatostatin and somatostatin analogs have shown promise in the treatment of tumors that do not respond to chemotherapy.⁽¹⁷⁾

The present case features several interesting points: the mediastinal mass did not extend to the neck; dissection was more difficult in the inferior mediastinum than in the superior mediastinum; although it measured 3 cm in diameter, the thymoma was not detected in the pre-operative computed tomography scan; and detailed exploration of the mediastinum allowed us to find a tumor presenting a different histology in another location.

We conclude that, after the diagnosis and the surgical treatment have been defined, the surgeon should carefully explore the mediastinum during resection, considering the possibility of other concomitant lesions, as we have demonstrated in this case. In cases of thymoma, it is important to

conduct an intra-operative investigation into the possible invasion of adjacent structures. The anatomopathologic analysis can reveal, even in small-diameter lesions, whether the disease has reached an advanced stage and therefore might require adjuvant treatment.

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