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Case report

Pleural thymoma: A rare cause of pleural mass.

About one case [☆]

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ABSTRACT

Thymoma is a rare tumor arising from the epithelium of the thymus gland and is usually located in the anterior mediastinum. Ectopic thymoma is very rare and can be located in the neck, trachea, lung, and pleura. Through this manuscript, we report a rare case of a 50-year-old woman presenting with dyspnea and recurrent pleural effusion. Her computed tomography of the thorax showed a unilateral left pleural mass. Biopsy of the pleural mass confirmed the diagnosis of pleural thymoma and she was started on chemotherapy. To our knowledge, this is the first reported pleural thymoma in our region.

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Introduction

Epithelial thymic tumors (thymoma and thymic carcinoma) are rare neoplasms. Their incidence is estimated to be 0.15 cases per 100,000 persons/y and less than 1% of primary malignancies in adults [1]. It is considered however to be the most common primary neoplasm of the anterior mediastinum, accounting for 20% of tumors in this location [2]. Ectopic thymoma is exceptional and has been described in the middle mediastinum, posterior mediastinum, neck, lung, and pleura [3]. Clinical manifestations are various. Here, we report a case of pleural thymoma presenting a unilateral left pleural mass and describe the main radiological finding on X-ray, computed tomography (CT).

Observation

A 50-year-old woman with unremarkable medical history (no diabetes, blood hypertension, lung tuberculosis, or smoking history) excepted a well-managed depression presented two months ago with gradual onset worsening difficulty in breathing. It was associated to dry cough, chest pain, fatigue, and loss of weight. There were no fever, hemoptysis, or heart failure symptoms. Physical examination showed a cachexic patient (body mass index: 16) but she was comfortable and not tachypneic. Her vital signs were normal. Thoracic examination showed dullness on percussion of the left lower thoracic zone. Cardiovascular, abdominal, and neurological examinations were normal. There was no palpable lymph node.

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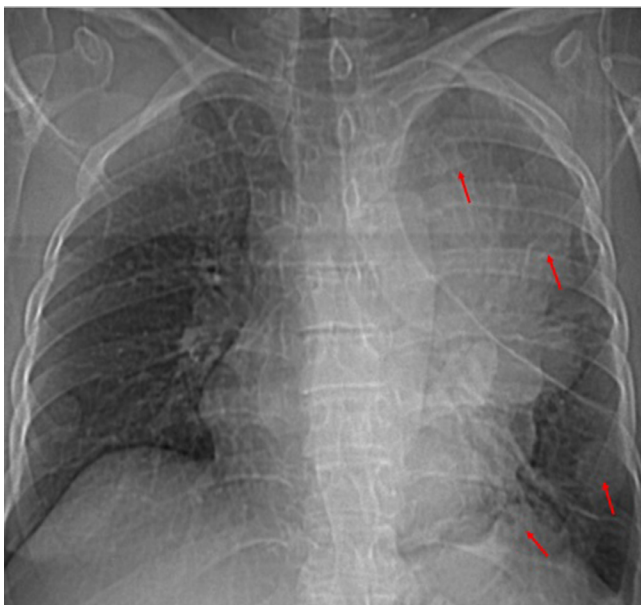


Fig. 1 – Plain chest radiography showing apical and basal extra pulmonary masses (red arrow).

There were no signs of myasthenia gravis as well. Her full blood count, renal function, and liver function test were normal. Her echocardiography was normal. Her chest radiograph showed apical and basal pleural opacities (Fig. 1) in the left hemithorax. Thoracic computed tomography showed unilateral nodular mildly enhancing pleural mass in basal, upper and mediastinal pleura. (Fig. 2). There was no pleural effusion as well. Given the CT findings, the differentials diagnostics were metastasis, lymphoma, mesothelioma, or benign tumor. Subsequently, an ultrasound-guided biopsy of the left pleural mass was done. One week later, the biopsy report returned. The biopsy showed a fragment of fibrous tissue infiltrated by sheet-like neoplastic epithelial cells admixed with non-neoplastic lymphocytes cells (Fig. 3A). Immunohistochemically study showed diffuse strong positivity to CKAE1/AE3 (Fig. 3B), CD 5 (Fig. 3C), and CD20 (Fig. 3D). It confirmed the diagnosis of pleural thymoma type B 1. The tumor was negative for CD117, excluding thymic carcinoma. A multidisciplinary team meeting was held including radiologist, oncologist, pulmonologist, thoracic surgeon. We decided to start neoadjuvant chemotherapy using cisplatin, doxorubicin, and cyclophosphamide. She tolerated the chemotherapy and she is programmed for radical surgery.

Discussion

Thymoma is a rare condition but remains the most common neoplasm of anterior mediastinum and the most frequent thymic tumor. It originates from epithelial thymic cells. It occurs in elderly patients, with male predominance, frequently between 50–60 years old. It is extremely rare before 20 years old [4].



Fig. 2 – Axial(A), coronal (B) and sagittal (C) enhanced chest computed tomography showing apical and basal pleural masses in the left hemithorax. Notice the vacuity of the anterior mediastinum excluding a pleural metastasis of a mediastinal thymoma.

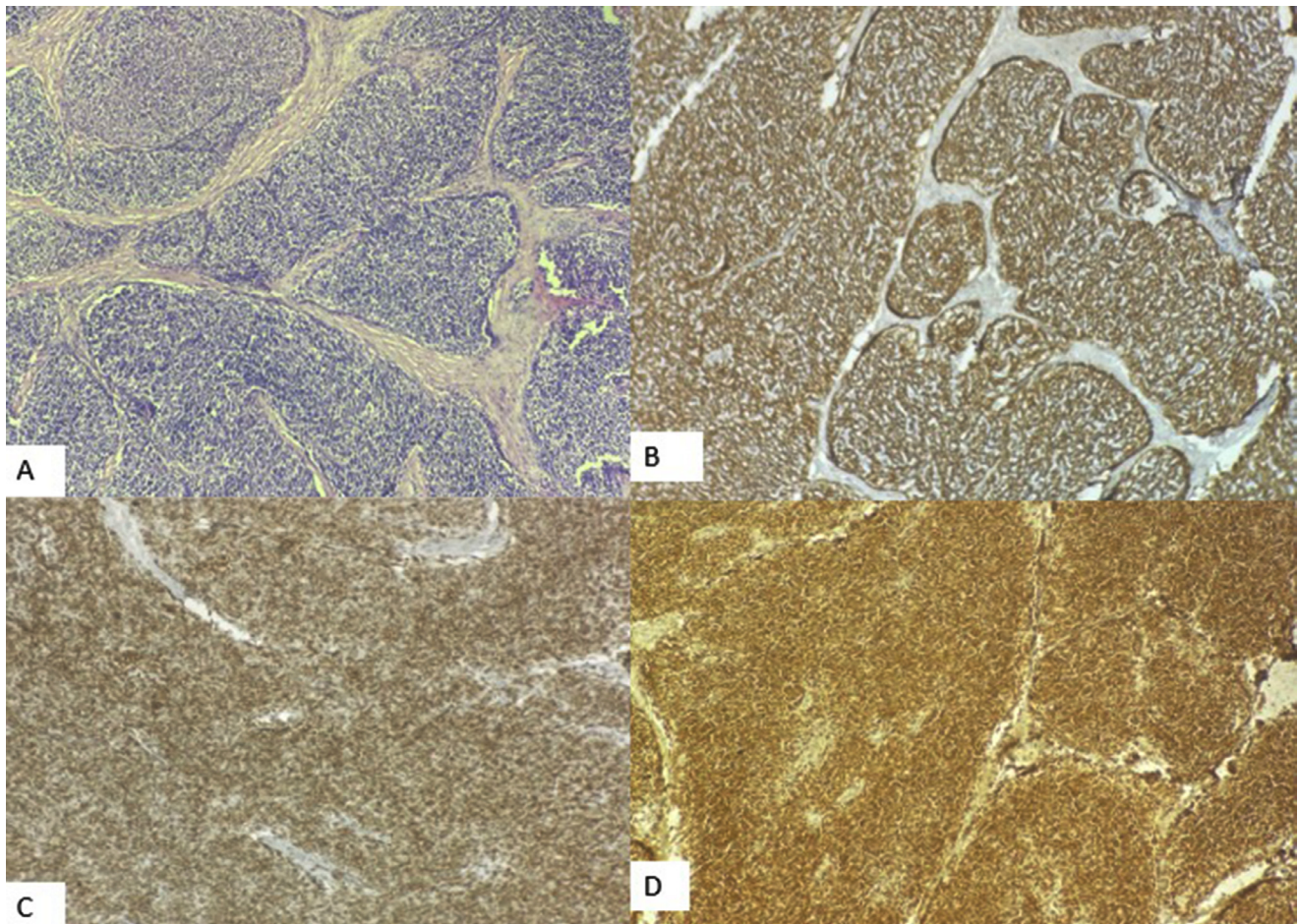


Fig. 3 – Histopathological view of the pleural mass biopsy (x10). Hematoxylin and eosin staining showing a fibrous tissue infiltrated by sheet-like neoplastic epithelial cells admixed with non-neoplastic lymphocytes cells (A). Immunohistochemically study showed diffuse strong positivity to CKAE1/AE3 (B), CD 5 (C) and CD20 (D).

The embryological origin of the thymic epithelium is the third or fourth branchial pouches. It descends caudally with the third parathyroid into the anterior mediastinum. Aberrant migration of thymic tissue can occur anywhere along this pathway which can result in ectopic thymic tissue. Thus, ectopic thymomas originate from these ectopic thymic tissues.

Thymoma originating from the mediastinum and spreading along the pleura is a common condition, however, thymomas originating from the pleura are extremely rare, and very few cases are described in the literature [5].

Clinical presentations of thymomas are various, including local symptoms related to the compression of surrounding structures like cough, chest pain, superior vena cava syndrome, dysphagia, and hoarseness of voice. Out of 30% of patients with thymoma have clinical symptoms related to myasthenia gravis. Out of 5% of patients have other systemic syndromes including red cell aplasia, dermatomyositis, systemic lupus erythematosus, Cushing syndrome, and syndrome of inappropriate antidiuretic hormone secretion [6]. About 70% of patients with thymoma remain asymptomatic.

Radiological aspects of pleural thymoma are various and not specific. They obviously share the same characteristics as

other pleural masses including mesothelioma, pleural metastasis, fibrous solitary tumor, or chest wall sarcoma. Chest radiograph typically shows pleural thickening with encasement of the lung or pleural mass lesions. On CT thorax, it usually appears as one or more unilateral pleural nodules, separate from the main mass localized in anterior-superior mediastinum. Homolateral pleural effusion could be present and indicates advanced disease. Calcification could be present. After iodinated contrast injection, they usually show a homogenous enhancement, except for huge masses that contain areas of hemorrhagic or necrotic components.

CT allows us also to appreciate the local invasion of neighboring structures. The presence of well-delimited adipose tissue between the mass and other organs and mediastinal structures suggests low aggression. The main vessels, pericardial or pleural overrun instead, hematogenous or lymphatic metastasis lean toward an invasive disease with one or more pleural nodules.

Biopsy and histopathological examination are necessary to assess the diagnostic of pleural thymoma. On microscopic examination, the tumor has a lobular configuration separated by fibrous septa associated with a dense lymphoid process.

It shows a biphasic cells population with lymphoid cells and neoplastic epithelial cells. Immunohistochemical staining can further characterize the tumor. The epithelial cells typically show immunoreactivity to CKAE1/AE3, CK5/6, and p63 protein. The T lymphocytes in the tumor express immunoreactivity towards terminal deoxynucleotidyl transferase (TdT) and CD99. On the other hand, PAX-8 and CD117 can differentiate thymoma from thymic carcinoma.

The World Health Organization histological classification system is the most used classification [7]. It is based on histological appearance and correlates with the likelihood of invasiveness and thus with staging. Type A and type AB are usually clinically benign and encapsulated, type B has a greater likelihood of invasiveness, especially type B3 and type C is almost always invasive. [8]

Therapeutic options are various. The management requires a multidisciplinary approach including a thoracic surgeon, oncologist, radiotherapist, and radiologist. Surgery remains the cornerstone of therapy except for widely metastatic disease. A complete resection represents the critical factor in determining long-term survival [9]. On the contrary, there is no benefits by debulking surgery. The role of debulking surgery should be more extensively investigated before stating that extensive surgical procedures is really necessary.

In the other hand, systemic chemotherapy, particularly the neoadjuvant setting had shown excellent results. The combination of cisplatin, doxorubicin and cyclophosphamide, the combination of cisplatin, doxorubicin, vincristine and cyclophosphamide, and the combination of cisplatin, etoposide, and epidoxorubicin have reported objective response rates ranging from 77% to 100% [10].

Thymomas are very sensitive to radiation. The radiation has been used alone or in combination with chemotherapy for unresectable thymoma. Its role as an adjuvant treatment after surgical resection is controversial and it could be useful at doses of 45–55 Gy to reduce the relapse rate only in the really invasive stage [11].

In summary, pleural thymomas are exceptional malignancies. Imaging findings are not specific but necessary for biopsy guidance, therapeutic planification, and further follow-up. X-rays is the first level exam, but the gold standard is contrast-enhanced computed tomography, allowing detection and evaluation of the response to oncologic therapy and assessing the involvement of the adjacent structures. Definitive diagnosis and staging are histological. Thymomas are chemotherapy and radiotherapy sensitive, nevertheless, complete surgical excision is the preferred treatment whenever technically feasible.

Patient consent

Consent was obtained from the patient. The study was conducted anonymously.

Availability of data and materials

The data sets are generated on the data system of the university hospital of Fez.

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