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Case Report:

A Rare Giant Pleural Thymoma in Posterior Mediastinum.

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Abstract: Thymoma is an epithelial neoplasm of the thymus, which commonly lies in the anterior mediastinum. Unusually, thymomas can also be found in other locations. Surgical excision, when feasible, appears to provide good results. We encountered a rare case of a thymoma that developed in the right thoracic cavity, and originating from the pleura in posterior mediastinum. We describe the clinical scenario, investigations, and our management of the patient.
Keywords: Thymoma; Pleural tumor; Posterior mediastinum; Surgical excision

Introduction

Thymoma is an epithelial neoplasm of the thymus, which commonly lies in the anterior mediastinum. unusually, thymomas can also be found in other locations. 75% of the thymomas are within the anterior mediastinum, 15% are in both the anterior and superior mediastinum, and 6% are located in the superior mediastinum.[1] The remaining 4% of thymomas occur ectopically, affecting the neck, middle or posterior mediastinum, and lung.[2] However, ectopic thymoma occurring in the pleura is extremely rare and has been infrequently documented.[3] In the posterior mediastinum, the occurrence of ectopic thymoma has rarely been reported. We encountered a rare case of a thymoma that developed in the right thoracic cavity, and originating from the pleura in posterior mediastinum. We describe the presentation and management of our patient and review the literature.

Case Report

Our patient was a 34 year old female who presented with history of non-productive cough since 2 months. There was no history of haemoptysis, chest pain, evening rise of temperature or past history of tuberculosis. On examination, she was well built and nourished. There was no pallor, icterus, clubbing, or generalized lymphadenopathy. Her respiratory examination revealed dullness and decreased air entry in the right lower zone. Laboratory investigation revealed a normal hemogram and normal liver and renal function.

Chest radiography showed a well-defined dense opacity in the right lower zone. Computed tomography (CT) of the thorax (Fig. 1) showed a large well circumscribed solid heterogenous enhancing mass lesion at right lower hemithorax. Inferiorly abutting diaphargamatic pleura, indentation seen with no obvious infiltration. Medially it abutts mediastinal structures with doubtful infiltration (Fig. 2). Sorrounding lung parenchyma appeared normal except superiorly some pneumonities was seen.

On Bronchoscopy, extrensic compression of right middle and lower lobe bronchi noted with no obvious endobronchial growth. Pulmonary function test shown very severe restrictive abnormality. Image guided fine-needle aspiration and core needle biopsy of the lesion gave provisional diagnosis of Thymoma or Synovial sarcoma. She was scheduled for thoracoscopy for further evaluation.

On thoracoscopy, the lesion was found to be 20*15 cm well defined growth present in posterior medistinum between right lung and diaphragm compressing right middle and lower lobe. There was no associated effusion, lymphadenopathy, or lung lesion. As the lesion was large and vascular, right posterolateral thoracotomy was performed, and the lesion was amenable to complete resection with a strip of involved pericardium. The patient had an uneventful recovery and was discharged on the postoperative day 6.

Gross examination of the lesion revealed a 20*15 cm well-encapsulated lesion with strip of pericardium. Microscopy showed neoplastic proliferation composed of a mixture of lymphocytes and keratin-positive epithelial cells with fibrous bands. High-grade tumor region shown elongated tumor cells in a trabecular arrangement with hyalinized stroma, showing region composed of spindle cells arranged in the conventional composite AB pattern with sharply delineated lymphocyte-rich and lymphocyte-poor areas consistent with thymoma (Fig. 3). Immunohistochemistry was done on the

specimen. It was positive for leukocyte common antigen and focally positive for cytokeratin. The final diagnosis of minimally invasive Thymoma Type AB was made.
Patient received adjuvant IMRT(intensity modulated radiation therapy) 54 grays in 30 fractions. Now patient is on regular follow up.

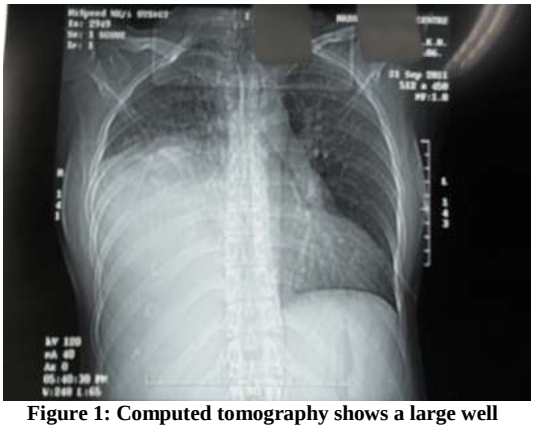

Figure 1: Computed tomography shows a large well circumscribed solid heterogenous enhancing mass lesion at right lower hemithorax.

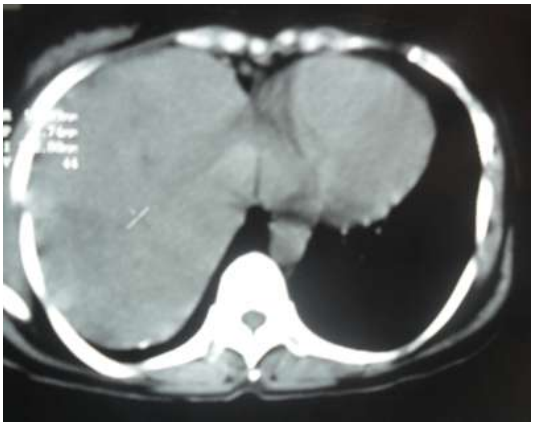

Figure 2: Computed tomography shows mass medially abutting mediastinal structures with doubtful infiltration

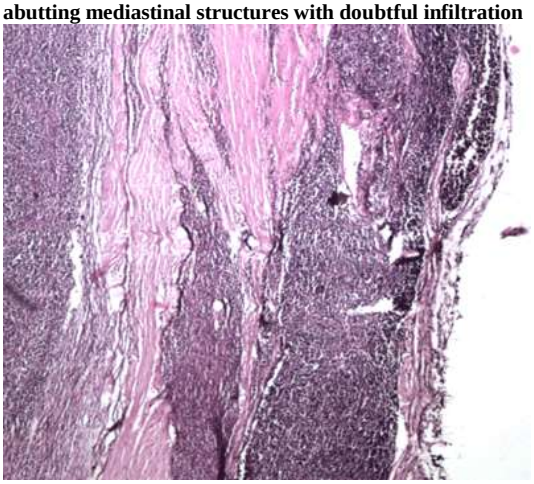

Figure 3: Histopathological specimen showing tumor composed of spindle cells arranged in the conventional composite AB pattern with sharply delineated lymphocyte-rich (*right*) and lymphocyte-poor (*left*) areas

Discussion

The thymus arises in the ventral portion of the third and fourth pharyngeal pouch. The third or fourth branchial pouches descends caudally with the third parathyroid into the anterior mediastinum by the fifth or sixth week of gestation. Any errors occurring during this phase can cause dissemination of aberrant nodules that are responsible for most uncommon thymomas. Of the thymomas, 75% are within the anterior mediastinum, 15% are in both the anterior and superior mediastinum, and 6% are located in the superior mediastinum.[1] The remaining 4% of thymomas occur ectopically, affecting the neck, middle or posterior mediastinum, and lung.[2] However, ectopic thymoma occurring in the pleura is extremely rare and has been infrequently documented.[3] In the posterior mediastinum, the occurrence of ectopic thymoma has rarely been reported.

These tumor cells spread via regional metastasis or invade surrounding structures, including the pleural space. In rare cases, advanced malignant thymoma may present as a large mediastinal mass with pleural involvement.

Most patients with invasive thymoma are typically in the fourth or fifth decade of life and present with constitutional symptoms, including fever, night sweats, and weight loss, in addition to symptoms related to local compressive mass effects, such as chest pain, dyspnea, dysphagia, and cough. Invasive thymoma also has been associated with paraneoplastic syndromes, including myasthenia gravis(35%), pure red cell aplasia, and hypogammaglobulinemia.[4]

Typical radiographic findings demonstrate a smooth or lobulated mass located in the anterior mediastinum. Other radiographic features include calcification within the mass and local invasion with pleural effusion and thickening. It is unusual for the disease to present as a massive mediastinal and pleural-based mass. MRI findings of the thymoma have the same or slightly increased intensity as that of muscle on T1-weighted images and increased intensity on T2-weighted images. Inhomogeneous signal intensity on T2-weighted images with a lobulated border, fibrous band, and lobulated internal architecture is indicative of an invasive thymoma.[5] There is a single case report of a giant thymoma completely filling the right hemithorax in a 15-year-old patient.[6]

A thoracic CT scan and tissue samples are important in determining the stage of the disease using the Masaoka staging system.[7] Stage I malignant thymoma demonstrates encapsulation without microscopic capsular invasion, and stage II disease involves invasion of mediastinum or microscopic capsular invasion. In stage III malignant thymoma, there is invasion into surrounding structures, including the pericardium, great vessels, and lung. Stage IVA malignant thymoma involves pleural or pericardial metastases, and stage IVB disease includes lymphogenous or hematogenous metastases.

A tissue sample is required for making the diagnosis of malignant thymoma and for ruling out other possible diagnoses, such as lymphoma and thymic carcinoma. Peripherally located lesions can be accessed via CT-guided core biopsy, whereas centrally located tumors may be approached via Bronchoscopy or Mediastinoscopy.

Histolological features consist of a mixture of Keratin-positive neoplastic epithelial cells and lymphocytes subdivided by fibrous bands. More recently, the frequency of the CD20 staining in spindle (medullary, WHO type A) and mixed spindle/lymphocytic (WHO type AB) thymoma has also been described as a specific feature of such thymoma types.[8]

Prognosis and outcome are directly related to the stage of the disease and the success of surgical resection. Patients with early stage I disease have excellent 5- and 10-year survival

rates of 100% and 95%, respectively.[9,10]However, in patients with advanced stage IV disease, 5- and 10-year survival rates are less than 25% without treatment.[9,10] With treatment, 5-year survival improves to 71% for those with stage IVA disease and 52% for those with stage IVB disease.[9,10] For patients with stage I disease who have had complete resection, there are no data to support the use of postoperative radiation treatment.[4] However, postoperative radiation therapy is recommended for patients with stage III and IV invasive thymoma regardless of resection status because of the high local recurrence rate.[4]

There are no large, randomized, controlled trials that demonstrate an optimal treatment regimen for advanced invasive thymoma. However, results from multiple small trials using a combination of induction chemotherapy, surgical resection, and postoperative radiation therapy followed by consolidation chemotherapy show the best outcome.[4,9,] This combination treatment plan demonstrated overall survival of 95% at 5 years and 79% at 7 years in a phase 2 study by Kim and associates10 in 2004.[11]

In patients with pleural thymoma metastases, experimental trials using cytoreductive surgery and Intraoperative hyperthermic intrathoracic chemotherapy with cisplatin and doxorubicin have demonstrated limited efficacy with a high local recurrence rate.[12] Other treatment options include the use of octreotide to suppress the somatostatin receptor on tumor cells and prednisone to inhibit growth and apoptosis.[13] Future treatment options include the use of umbilical cord blood transplant, high-dose chemotherapy followed by peripheral blood stem cell transplant, and tyrosine kinase inhibitor (gefitinib).[14]

Conclusion

Malignant thymoma is usually an indolent tumor that arises from the anterior mediastinum. In the posterior mediastinum, the occurrence of ectopic thymoma has rarely been reported. Radiographic appearance and tissue samples are very important in determining the diagnosis as well as the stage of the disease. The prognosis of malignant thymoma depends on the stage of disease and the surgical resection. In advanced Stage III or IV disease, the multimodal approach including the use of chemotherapy, radiation therapy, and surgery may result in the best clinical outcome. All patients with malignant thymoma have to have lifelong monitoring because of the risk of recurrence many years after treatment.

In summary, this report documents an extremely rare occurrence of ectopic pleural thymoma occurring in posterior mediastinum presenting as a Giant mass in the thoracic cavity.

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