Student's Corner

Extensive primary malignant Thymoma involving Pericardium, Pleura, Diaphragm and Lungs - A case report

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Abstract

Malignant thymoma is an extremely rare entity. According to a study, the annual incidence of malignant thymoma was estimated to be 0.15 per 100,000 person-years. We present the case of a 42-year-old female who came to us with a Type AB, Masaoka stage III malignant (invasive) thymoma with widespread involvement of the mediastinum.

Introduction

Invasive (malignant) thymomas are rare tumours, which represent about 20-25% of all mediastinal tumours and 50% of anterior mediastinal masses. The etiology of these types of tumours is not known.[1] In about 50% of the patients, thymomas are detected by chance with plain-film chest radiography. Standard primary treatment for these types of tumours is surgical with en masse resection for invasive tumours. Complete surgical resection is an important prognostic factor for locally advanced malignant thymoma.[2-4] We describe the case of a 42-year-old female who presented with an extensive stage III malignant thymoma involving pleura, diaphragm, and phrenic nerve invading the left lower lung and the pericardium.

Case Report

A 42-year-old non-smoking, healthy female with no known co-morbidities, presented to us with symptoms of episodic left sided subcostal chest pain for 2 months associated with backache and low-grade fever. General physical examination was unremarkable except for basal crepitations on the left side of the chest.

There was no history of previous hospitalization for any surgeries or medical problems. She was, however, being treated for a suspected pneumonia at a local hospital. During her work up for pneumonia, a chest x-ray was done which revealed a lobulated radio-opaque density on the left lower lung lobe and was subsequently referred to us.

Chest roentgenogram showed a large soft tissue density mass adjacent to left bronchus. CT scan with contrast revealed an intermediate to low-density lobulated mass in the anterior mediastinum, which was compressing the heart and major blood vessels. The heart was pushed to the right side across midline but no pericardial effusion was noted. The mass was also attached to the linguila, associated with moderate left pleural effusion. A 2cm pre-carinal node was also visible.

MRI showed a similar picture of a lobulated, solid, mildly enhancing anterior mediastinal mass measuring 12.5 × 11.1 × 7.9 cm however, another pleural based solid lesion was also appreciated along the posterior aspect of left lower lobe measuring 1.5 × 3.1 cm. Bronchoscopy demonstrated no endobronchial lesion. The culture was negative for acid-fast bacilli, other microorganisms and malignant cells. Immunohistochemistry reports were positive for Thymoma cytokeratin.

Histological analysis of CT guided trucut biopsy of the pulmonary mass showed the tumour to be consisting of neoplastic epithelial cells and non-neoplastic lymphocytes. The tumour also demonstrated signs of microinvasion of the capsule. The findings were consistent with malignant (invasive) thymoma of mixed (lymphoepithelial) variety. The diaphragmatic mass and left lateral pleural basal nodules were found to be tumour deposits. Mediastinal lymph nodes were free of tumour metastasis.

The patient underwent preoperative chemotherapy. After the shrinkage of tumour size, posterolateral thoracotomy for complete thymectomy plus excision of left lung and parietal pleural masses was done through the fifth intercostal space, and the residual tumour along with left linguila of the lung and pericardium were removed, however the phrenic nerve was cautiously preserved. Postoperative course was uneventful and the patient was discharged on seventh postoperative day.

Discussion

Thymoma is a rare tumour arising from the epithelium of the thymus gland but it is the most common neoplasm of the anterior mediastinum comprising about 20-25% of all mediastinal tumours and 50% of anterior mediastinal masses.[1] Thirty to 40 percent of these are malignant with malignancy usually defined by the surgical findings of invasion or the presence of intrathoracic or

extrathoracic metastases.[4] In a study conducted in US, the overall incidence of malignant thymoma was found to be 15 per 10000000 person-years.[5]

Malignant thymoma is detected most frequently in the fourth and fifth decade of life. Men and women are equally affected. It is rare in children.[6]

Approximately 30% of the patients with thymoma are asymptomatic at the time of diagnosis.[4] In other cases, the presenting clinical signs of these types of tumours may include coughing, chest pain, and signs of upper airway congestion. Paraneoplastic autoimmune syndromes associated with thymoma include myasthenia gravis (MG), polymyositis, lupus erythematosus, rheumatoid arthritis, thyroiditis, and Sjögren's syndrome, among others. As many as 30-40% of patients who have a thymoma also suffer from MG.[2,4] Autoimmune pure red cell aplasia and hypogammaglobulinemia affect approximately 5% and 5 to 10%, respectively, of patients with thymoma.[6]

Diagnosis is made clinically based on the radiological findings. Laboratory blood investigations are generally not indicated. Chest x-ray, including lateral view can detect most thymomas. CT scan may delineate the lesion further or detect smaller tumours and aid in characterization of thymoma to successfully distinguish it from other benign mediastinal tumours (especially lymphomas and germ cell tumours). It can also reveal features suggestive of malignancy - for instance; vascular invasion, encasement and pleural deposits. Biopsy is performed in cases where there is suspicion of malignancy in order to initiate proper treatment.[6]

The only treatment for resectable malignant thymomas is complete resection and the most common operative approach is usually median sternotomy, although we employed a left posterolateral thoracotomy due to pleural and diaphragmatic involvement and the need for excision of left lower lung lobe.[2,3] Total resection of a well-encapsulated, noninvasive thymoma is usually curative and has a risk of local recurrence of less than 2%.[3] Some reports have shown excellent long-term survival with extended resection in patients with stage III and IV disease. Their overall 5- and 10-year survival rates were 77% and 59%, respectively.[7] The value of incomplete resection is quite controversial. Many authors believe that incomplete resection has no survival or recurrence value over biopsy alone.[8] Other authors, including Mornex et al, found that survival was better after partial resection than after biopsy (5-year survival, 64% vs. 39%, respectively).[8]

Local recurrence following surgery occurs in less than 2% of patients who have surgery for encapsulated lesions and in 20% to 40% of those who undergo surgery for invasive disease.[2]

Adjuvant radiation therapy is definitely indicated for completely or incompletely resected stage III or IV thymomas.[2,6,8] The role of chemotherapy is yet not established.[8]

Several retrospective studies have attempted to define clinicopathologic variables that can be used to predict prognosis in patients with thymoma. In one multivariate analysis conducted at John Hopkins, four factors proved to have an independent influence on overall survival: the extent of surgical resection, the presence of MG, the histology, and age at initial diagnosis. MG was found to be a positive prognostic factor where as age, gender and race failed to reach statistical significance as independent predictors of both overall and thymoma-related survival.[6] The only variable that proved to be a significant independent predictor of decreased thymoma-related death was complete surgical resection independent of Masaoka stage. These results support aggressive surgical intervention, even in patients with advanced disease at initial presentation.

Conclusion

Malignant thymoma is an extremely rare entity. The etiology of these types of tumours is not known. Standard primary treatment is surgical with en masse resection for invasive tumours. Adjuvant radiation therapy is indicated for stage III and IV malignant thymomas.

References

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