2017

www.jmscr.igmpublication.org Impact Factor 5.84 Index Copernicus Value: 71.58 ISSN (e)-2347-176x ISSN (p) 2455-0450 crossref DOI: _https://dx.doi.org/10.18535/jmscr/v5i12.42



Journal Of Medical Science And Clinical Research An Official Publication Of IGM Publication

Unusual Presentation of Infiltrating Thymoma in Myesthenia Patient: A Case Report and Review of Literature

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Abstract

Myasthenia gravis (MG) is associated with multiple variants of invasive thymomas which have been reported in literature. Unfortunately there is no clear histologic distinction exists between benign and malignant thymomas. These cases if not diagnosed properly may present in later stages with a turbulent course and prognosis. Invasive thymomas were correlated with a higher probability of recurrence. Good outcome can be obtained if early and definitive diagnosis of capsular invasion has been made during histopathological examination (HPE). The consensus is that patients with invasive thymoma, should receive neo-adjuvant therapy for better outcome. We encountered a rare type of invasive thymoma in which thymic cells were infiltrating through the capsule into pericapsular adipose tissue. This type of capsular invasion maybe considered potentially curable, if diagnosed early and can guide clinicians in deciding neo-adjuvant therapy after radical surgery.

Keywords: Invasive thymoma, Myasthenia gravis, neo adjuvent therapy.

Introduction

Invasive thymoma are cytologically bland but locally invasive tumors. They accounts for 20% to 25% of all thymomas. These tumors also occasionally and unpredictably metastasize to adjacent as well as mediastinal organs¹. A large clinico-pathologic series that shows that appropriate staging of these tumors are the best predictor of outcome.²

Unfortunately there is no clear histologic distinction between benign and malignant thymomas exists. The propensity of a thymoma to

be malignant is determined by the invasiveness of the thymoma. Recurrence rate of invasive thymoma is very high³. Invasive thymomas are defined by the Masaoka staging system as above stage II^4 . Staging of thymoma is very crucial in terms of treatment and prognosis⁵. We present unusual case of thymoma with myesthenia gravis which was diagnosed as case of invasive thymoma after histopathological examination which has changed treatment plan. Thus, it is advisable for pathologists to exclude the possibility of invasive thymoma and its variants whenever a thymoma

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specimen comes for hisptopthological examination.

Case report

A man in his late 50s presented with weakness of limb muscles after exertion and recurrent drooping of eye lids since 1 year. He was a professional singer and from last few days, he had difficulty in singing. He was evaluated thoroughly and a diagnosis of myesthenia garvis stage III a (Eye muscle weakness of any severity, moderate weakness of other muscles, Predominantly limb or axial muscles) of American clinical classification of Myesthenia Gravis with thymic hyperplasia was made on the basis of chest CT and antibody against the acetylcholine receptor levels. He was planned for routine thymoma excision surgery. But during surgery unusual fibrosis of posterior thymic wall with adherence to pericardium was noticed. Suspicion of malignancy was arise and thus he underwent radical surgery with removal of adhere pericardium along with thymus and removal of mediastinal lymphnodes. All samples were sent for histopathological examinations to rule out infiltrating thymic carcinoma. On histopathological examination various sections of specimen were studied. After thorough HPE examination diagnosis of invasive thymoma was treatment made and plan was modified accordingly.

Pathologic findings

The resected specimen consisted of a reddish gray, lobulated, rubbery to firm tissue mass that measured $12 \times 10 \times 5$ cm. The outer surface was irregular and focally hemorrhagic. Posterior surface was smooth without any invasion on inspection. Pericardium was adherent to posterior surface of thymic tissue and cannot be differentiated but cardiac surface of pricardium is smooth and without the evidence of invasion. The cut section showed gray white, homogeneous firm to rubbery tissue with focal areas of fibrosis. (Fig. 1)

Histopathological examination Lobules of epitheloid cells with small number of lymphocytes were seen. The epithelial cells are rotated to polygonal with vesicular nuclei and prominent nuclei sharing mild nuclei atypia. Focal areas of cholestrol clefts and gaint cells are seen. The tumor is infiltrating through the capsule into pericapsular adipose tissue. At places cystic areas are also seen.Lympho-vascular invasion is seen without peri-nural infiltration. The tumor was reaching upto the pericardium, however invasion of pericardium was not seen. Rescted lymphnodes shows sinus histocytosis without evidence of metastasis. Thus diagnosis of thymoma histological type B3 (Table 1) Masoka staging IIB was made. (Fig. 2,3)



Fig 1: Cross section of specimen showing fibrotic lesion with cystic areas.



Fig 2: Invasion of capsule by thalamus tumor cells



Fig: 3 H&E staining Polygonal vesicular prominent and mild nuclei atypia along with focal areas of cholestrol clefts and gaint cells are prominent in thymic tumour cells.

Discussion

Thymoma is a unique thymic epithelial neoplasm with indolent growth and rarely presents with local invasiveness and metastases⁶. Thymomaassociated MG (T-MG) is identified as a tumor originating from thymic epithelial cells, most cortical subtype (World being Health Organization typeB)⁷. Because thymoma usually has some morphological similarities with the thymic cortex, they share the ability to propagate the maturation of immature naive T cells into the periphery⁸. Malignant thymomas can invade the vasculature, lymphatics, and adjacent structures within the mediastinum (figure 3). The 15-year survival rate is 12.5% for a person with an invasive thymoma and 47% for a person with a noninvasive thymoma. The single most important factor predicting the outcome of patients with thymomas is evidence of invasion⁹. Thus histologic characteristics, such as microscopic capsular invasion should be assessed in each and every case of thymoma. Because of the welldocumented propensity for late recurrences in invasive thymoma, long-term survival is mainly depend on accurate histopathological examination. A study conducted by the Memorial SloanKettering Cancer Center reported 5-year and 10year survival rates to corresponding stages of thymomas¹⁰. Thymomas are also associated with the development of second malignancies. Early and prompt diagnosis of unusual presentation by quick histopathogical examination allowed patient to receive neo adjuvent chemo and radiotherapy immediately with good postoperative prognosis¹⁰. In this case diagnosis of Type B3 thymoma. (also known as epithelial thymoma, atypical thymoma, squamoid thymoma, and well-differentiated thymic carcinoma) of WHO classification is based on histopathological examination. It accounts for approximately 10% to 14% of all thymomas¹¹. Approximately 46% of cases may be associated with myasthenia gravis. Morphologically, this tumor type is predominantly composed of epithelial cells that have a round or polygonal shape and that exhibit no or mild atypia. The epithelial cells are admixed with a minor component of non neoplastic lymphocytes, which results in a sheet-like growth of neoplastic epithelial cells. The 20-year survival rate (as defined by freedom from tumor death) for this thymoma type is approximately $40\%^{12}$.

Conclusion

We describe a case of a rare variant of thymoma associated with MG, supporting the relationship between this variant of thymoma and MG. Another peculiar feature of the present case is the presence of several foci of capsular and peri lymphovascular invasion which constitutes an infrequent finding in this histological type of thymoma. Considering unusual histo-pathological presentation of this thymoma, clinicians planned adjuvant chemo-radiotherapy after resection in this case. However, the role of adjuvant therapy is still controversial and needs to be supported by the collection of other similar cases, it was decided to provide best available treatment.

In conclusion, this rare type of capsular invasion may be considered potentially cureble, if diagnosed early and can guide clinicians in deciding adjuvant therapy after radical surgery.

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