

# “Stage B2 Thymoma Presenting with Myasthenia Gravis” Successfully Treated with Multimodality Treatment: A Case Report from Tertiary Cancer Centre in North India

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## ABSTRACT

**Introduction:** Thymomas are rare tumors and account for less than 1% of the human cancers. As these tumors are exceedingly rare and having unpredictable natural history multimodality Rx consisting of surgery as mainstay of treatment, adjuvant radiotherapy for local control and chemotherapy is often practiced. Incidence for patients associated with myasthenia Gravis is high in 4<sup>th</sup> decade whereas for patients without myasthenia gravis, the peak age is in the 7th decade or later.

**Case Report:** We present a case of WHO stage B2 Thymoma in a 35-year-old male patient who reported in neurology department in a higher centre with complaints of drooping of eyelids, difficulty in chewing food and generalized weakness. A diagnosis of Myasthenia Gravis was made treatment was started for the same. His CECT Thorax revealed well defined oval homogenously enhancing mass on right side s/o thymoma. Metastatic workup revealed no sign of metastasis.

**Results:** At our institute he underwent total thymectomy which revealed minimally invasive thymoma (B2) with invasion into capsule and peri thymic adipose tissue. Patient was treated with adjuvant radiotherapy 56Gy/28# to the tumor bed and four rounds of chemotherapy based on CAP. Response was assessed with the help of CECT Thorax which revealed no e/o residual disease.

**Conclusion:** Type B2 Thymoma usually present with either encapsulated or partially circumscribed tumors and have tendency to invade mediastinal fat/other organs. However,

there are chances of recurrence of 5-9%. Our patient successfully completed multimodality treatment and is disease free since past 1 year.

**Keywords:** B2 Thymoma, lymphoepithelial, mediastinal neoplasms, WHO Classification

## INTRODUCTION

Thymomas are exceedingly rare tumors. The Surveillance, Epidemiology, and End Results (SEER) project reported that thymoma incidence to be 0.13 per 100,000 person-years.<sup>1</sup> They include thymic epithelial tumors (like thymomas, thymic carcinomas and neuroendocrine epithelial tumors), germ cell tumors, lymphoid/haematopoietic neoplasms and mesenchymal tumors.<sup>2</sup> Myasthenia Gravis is a disorder of abnormal neuromuscular transmission and is associated with thymoma in 15-20% of the cases.<sup>3</sup> Indication of thymectomy for all cases of myasthenia gravis is controversial debate but thymectomy is indicated in all cases with thymomas no matter the stage of myasthenia gravis.<sup>3</sup> Where as resection remains the main stay of the treatment, complete resection is not always possible and relapse is common. Here comes the role of adjuvant radiotherapy to address the local disease. Systemic chemotherapy is a standard for inoperable/recurrent disease.

## CASE REPORT

We present a case of 35-year-old male patient who reported in neurology department in a higher centre with chief complaints of drooping of eyelids, difficulty in chewing food and generalized weakness. A diagnosis of Myasthenia Gravis was made at the same centre and treatment was started for the same. Patient was referred at our cancer centre with diagnosis of Myasthenia gravis with mass in thorax. His CECT Thorax revealed well defined oval homogeneously enhancing mass on right side partly replacing the thymus measuring 2.5x1.3x2.4cm s/o thymoma. There was no mediastinal lymphadenopathy. There was no h/o any comorbidity. He was a tobacco chewer ("Zarda"- local chewable tobacco) non-smoker, did not consume alcohol. General examinations revealed all vitals within normal range. All routine investigations including cardiology clearance (taken for anthracycline based chemotherapy) were within normal range. Metastatic workup revealed no sign of metastasis.

### Treatment

Patient underwent Total Thymectomy. Detailed HPE report was s/o minimally invasive Thymoma (Type B2). Tumor was invading the capsule and peri thymic adipose tissue. Adjuvant radiotherapy and chemotherapy was planned.

Radiotherapy<sup>4</sup> consisted of total dose of 56 Gy/28#/5.5 weeks to the upper anterior mediastinum by AP:PA fields with 2:1 weightage. Radio-opaque clips helped to locate the tumor bed. (Figure 1)

Patient was given 4 cycles of chemotherapy based on cisplatin, doxorubicin, and cyclophosphamide.

Patient tolerated the treatment well with no interruptions due to radiation/chemotherapy induced toxicities.

The response was assessed after 6 weeks of last chemotherapy cycle with the

help of CECT Thorax which revealed no residual disease.

Patient is asymptomatic and disease free after 1 year of two monthly follow up.

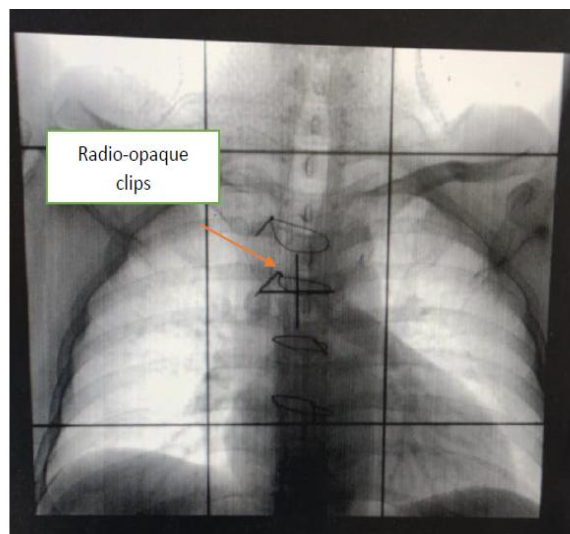


Figure 1: Simulation Images of radiotherapy planning. Radio-opaque clips assisted in ascertaining tumor bed.

## DISCUSSION

Radiographic presence of an anterior mediastinal mass along with myasthenia gravis<sup>6</sup> strongly suggests a clinical diagnosis of Thymic tumor. In this case report patient first reported in department of neurology for his cardinal symptoms of weakness (especially ocular muscles) and easy fatigability of skeletal muscles suggestive of Myasthenia Gravis.

Thymomas are generally characterized by an indolent grown pattern and are locally invasive.

Historically various classification systems have been proposed like Bernatz et al in 1961, Müller-Hermelink and Kirchner in 1985, Suster and Moran in 1999 and finally the current system-the WorldHealth Organization (WHO) classification-is an important step in this direction (Table I) WHO classification, first proposed in 1999 and updated in 2004 recognizes six different types of thymic tumors (A, AB, B1, B2, B3, C).

Masaoka<sup>7</sup> staging system is the surgical staging system of thymomas. (Table: II)

**Table: I Thymoma Classification Systems**

| Bernatz et al. (1961) | Müller-Hermelink and Kirchner (1985) | Suster and Moran (1999)      | WHO (1999) |
|-----------------------|--------------------------------------|------------------------------|------------|
| Spindle cell          | Medullary                            | Thymoma, well differentiated | Type A     |
|                       | Mixed                                |                              | Type AB    |
|                       | Predominantly cortical               |                              | Type B1    |
| Lymphocyte-rich       | Cortical                             |                              | Type B2    |
| Lymphoepithelial      |                                      |                              |            |
| Epithelial-rich       | Well-differentiated thymic carcinoma | Atypical thymoma             | Type B3    |
|                       | High-grade thymic carcinoma          | Thymic carcinoma             | Type C     |

**Table II Masaoka Staging system**

| Stage |   | Description  |
|-------|---|--|
| I     |   | Macroscopically completely encapsulated, with no microscopic capsular invasion       |
| II    | a | Macroscopic invasion into surrounding mediastinal fatty tissue or mediastinal pleura |
|       | b | Microscopic invasion into the capsule  |
| III   |   | Macroscopic invasion into surrounding organs   |
| IV    | a | Pleural or pericardial implants/dissemination  |
|       | b | Lymphogenous or hematogenous metastases  |

Zhengbo Song<sup>9</sup> et al evaluated the outcome of 42 patients with type B2 thymoma who were treated between 1995 and 2010 and indicated that Masaoka stage affects disease-free survival and overall survival of patients with type B2 thymoma. The 5-year disease-free survival and overall survival rates were 62.8% and 84.9%, respectively in their study.

## CONCLUSION

We conclude that Thymoma being a rare tumor: diagnosis, pathological classification as well as adequate treatment remains a challenge for clinicians. Multimodality treatment<sup>8</sup> may be required in B2 thymoma for potential cure and prolonged survival.

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