Original Research Article

Anterior mediastinal mass: A case report of thymoma with Myasthenia gravis

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Abstract

Introduction: Approximately 10-12 % of patients with myasthenia gravis are found to have a thymoma, and 20-20% percent of patients with thymoma may have myasthenia gravis. Thymomatous myasthenia gravis tends to have a difficult clinical course and poor prognosis. **Case presentation:** We report a case of typical presentations of myasthenia gravis associated with anterior mediastenal mass. Patient were diagnosed at a young age 21 yrs and presented with a turbulent course of myasthenia gravis and thymoma, but obtained good outcome after aggressive surgical treatment. **Conclusions:** Although thymomatous myasthenia gravis tends to have a difficult clinical course and poor prognosis, early and aggressive treatment along with multidisciplinary management may improve the outcome of these patients.
Key Word: Anterior mediastinal mass.

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INTRODUCTION

The most common neoplasm arising from thymic epithelial cells is Thymoma.[1] Incidence – 15-20 %. Thymomas are rare in the first 20 years of life. Its histological classification was proposed by the World Health Organization (WHO) in 2004.[2] According to this classification, thymoma can be categorized into 6 subtypes.

Table 1: Who Classification

Type	Histologic Description
A	Medullary Thymoma
AB	Mixed Thymoma
B1	Predominently Cortical Thymoma
B2	Cortical Thymoma
B3	Well- differentiated Thymic carcinoma
C	Thymic Carcinoma

In addition to these basic subtypes, other variants, such as micronodular thymoma with lymphoid stroma[3] and metaplastic thymoma[4] are well described. Other rare variants include microscopic thymoma[5], thymolipoma[6], lipofibroadenoma[7], and sclerosing thymoma.[8, 9]

Table 1. Thymoma staging system of Masaoka.

Stage	Description
I	Macroscopically completely encapsulated. Microscopically without capsular invasion
II	Macroscopic invasion into surrounding fatty tissue or mediastinal pleura. Microscopic invasion into capsule
III	Macroscopic invasion into neighboring organs (pericardium, lung, great vessels)
IVa	Pleural or pericardial dissemination
IVb	Lymphogenous or hematogenous metastasis.

Thymoma with myasthenia gravis a rare presentation. First case since 2008 at our institute.

CLINICAL SUMMARY

A 21-year-old, right-handed, male noticed the inability to perform certain daily activities which was associated with fatigability and decreased stamina for exercise beginning at the age of 19. Facial and proximal muscle weakness with difficulty in lifting his arms above his

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head and difficulty in climbing stairs. He was admitted to othropedic hospital for back ache and had an extensive workup, including brain and spine images as well as serum autoimmune studies, and was incidentaly found to have mediastenal mass in MRI scan. He was given one cycle of intravenous immunoglobulin (IVIG),which slightly improved his symptoms. Inspite of IVIG his condition deteriorated, and complained of difficulty in chewing, swallowing, associated with increase in weakness.He was then reffered to our Hospital. On presentation he manifested with classical myasthenic symptoms of extraocular weakness, bulbar and proximal upper and lower limb weakness with breathing difficulty. AChR binding antibody was elevated at 133.06 and blocking antibody was positive at 43. (AChR- Antibodies- 13.5, MUSK Antibody-0.008) IVIG, and pyridostigmine were initiated.A chest computed tomography (CT) scan showed a large anterior mediastinal heterogeneous mass with solid and cystic components measuring 7cm × 6cm × 5.2cm with invasion of the Left pleura and causing compression over Arch of aorta and left lung.

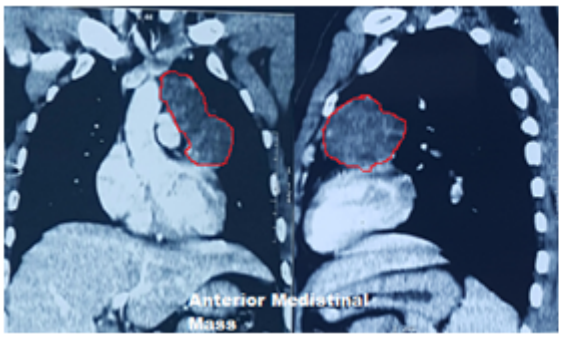

Figure 1: AP and Lateral view

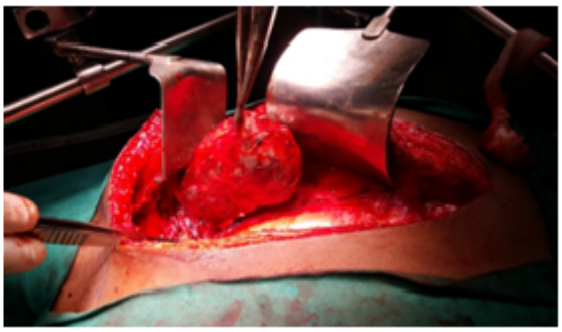

Figure 2: anterior mediastinal mass (intraoperative)

CT guided biopsy performed and showed Small round cell tumor. Pleural fluid cytology - Round cells. He had a complicated clinical course with numerous myasthenia crises requiring repeated admissions for weakness and difficulty in breathing but never needed endotracheal intubation for ventilator support. He later underwent a median sternotomy and 10x8x8 cm encapsulated mass in antero superior mediastinum to the left of midline, abutting arch of aorta and left lung, adherant to pleura was found (Figure-2). Left pleura opened while dissecting the mass - as pleura was adherant. Wide excision of mass done - R0 resection performed. The final pathology showed HPE/IHC- Morphology and immunohistochemistry support the diagnosis of thymoma, type B3.Microscopic invasion through the capsule seen (Masaoka stage IIA). Inked resected margin free. No vascular invasion seen. Immunohistochemistry: Immunohistochemistry s/o epithelial cells express CK, CD5(focally), EMA(diffusely), CK5/6, and CK19.Negative for CD3, CD20, CK7. Assessment scan performed showing No residual tumour. Serial chest imaging showed no evidence of recurrent tumor. He is currently 4 months post thymectomy and has significantly improved to an MGFA minimal manifestation status (MMS). However patient needed admission for generalised weakness with significant improvement in myasthenia symptoms.He is currently maintained on a low dose of both prednisone and pyridostigmine without IVIG.

Pathological findings: The surgically resected specimen revealed encapsulated grey white to grey brown tissue mass measuring 88x55x35mm. Outer surface is bosselated, capsulated. On cut surface, multiloculated cystic spaces are seen which are spongy and have honey-comb appearance. Few cystic spaces contain grey brown, soft to firm material. Greenish fluid exuded from the cyst.

DISCUSSION

The incidence of Myasthenia gravis in the general population is thought to be 0.5/100,000 to 3/100,000, and approximately 1% of all patients with Maysthenia gravis are children.[3] The disease is characterized by easy fatigability and fluctuating strength in skeletal muscles. Patients weaken rapidly with exercise and generally worsen as the day progresses. The most common muscles affected in children are those supplied by the cranial nerves. The defect underlying the disease is thought to be a result of an autoimmune mechanism that causes a decrease in the number of available acetylcholine receptors on the postsynaptic membrane. This is manifested by the presence of measurable antibodies to acetylcholine receptorsY2 as demonstrated in our patient.[4] The disease is counteracted by the use of anticholinesterase drugs. As many as 25% of thymomas are not detected by chest radiographs but are identified by CT scanning. CT remains an essential part of tumor staging, providing information about tumor location and size and extent of involvement of, or invasion into,

surrounding structures.[5] The natural course of MG in children is not well described, although several recent reviews would suggest a nearly normal life expectancy, with approximately 80% alive at 40 years of age. Respiratory failure is a common cause of death in MG and was a serious problem in our patient.[9,10] Several factors may precipitate respiratory compromise, including infections, surgery, emotional stress, decrease in dose of anticholinergic medication, or use of other medications such as aminoglycosides and muscle relaxants.[11,12] In summary, the association of MG and thymoma, although rare, does occur in the pediatric population. We report a case of Thymoma with myasthenia gravis in a 21 year old male.[2]

CONCLUSIONS

There is a known and established relationship between thymoma and myasthenia gravis. Although thymomatous myasthenia gravis tends to have a difficult clinical course and poor prognosis, early and aggressive treatment along with multidisciplinary management may improve the outcome of these patients. Thymectomy has been performed on patients who were refractory to drug therapy. In our institute we found such a rare case thymoma with myasthenia gravis since 2008 and underwent thymectomy which improved patient symptomatically. However patient needed hospitalization for genralised weakness but previous myesthenic symptoms didn't reappear.

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