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Case Report:
Thymoma with Myasthenia Gravis in Adolescent

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Abstract:
Thymomas are exceedingly rare in the first 20 years of life, Thymic lesions comprise approximately 2–3% of all pediatric mediastinal tumors and include thymic cysts, hyperplasia, carcinoma, and thymomas. Fewer than 30 cases in children have been described in the literature. Thymomas in adults are commonly associated with other diseases, the most frequent being myasthenia gravis. However, this association has been rarely reported in childhood. These tumors are typically aggressive, with poor outcomes. We report a case of thymoma associated with myasthenia gravis in a 16-year-old girl and review the literature.
Key Words: Mediastinal mass; Thymoma; Myasthenia Gravis

Case Report:
A 16 year old girl presented with difficulty in breathing, difficulty in swallowing and on and off vomiting since 6 days, she gives history of herpes infection 3 months back. On examination her chest and cardiovascular exam were normal. Her neurological examination reveled neck stiffness, neck flexor weakness and poor reflex, an initial chest radiograph showed an anterior mediastinal mass extending to the right of the ascending aorta. A computed tomographic (CT) scan showed a lobulated 15x12cm non-fat-containing soft tissue mass in the anterior mediastinum. There were no areas of chest wall or pericardial invasion or lymphadenopathy.
Electroneuromyography (ENMG) was normal, repetitive nerve stimulation (RNS) test revealed neuromuscular junction dysfunction, her serum anti acetylcholinestrase receptor antibody level was 9.28nmol/L (Normal range being <0.3nmol/L). Based on radiological and biochemical features patient was diagnosed as thymoma associated with myasthenia gravis and was treated with neostigmine, glycopyrrolate and plasmapheresis 3 hours/day for 7 days followed by an exploratory thoracotomy and a resection of the mediastinal mass were performed. On gross examination it was a well encapsulated mass measuring 14x11x8cms, outer surface was bossulated and smooth and cut surface was gray white solid with zig zag nodules.

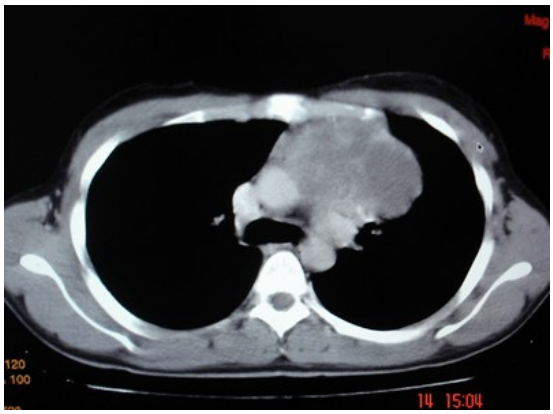

Figure 1: CT scan shows lobulated mass in the anterior mediastinum.

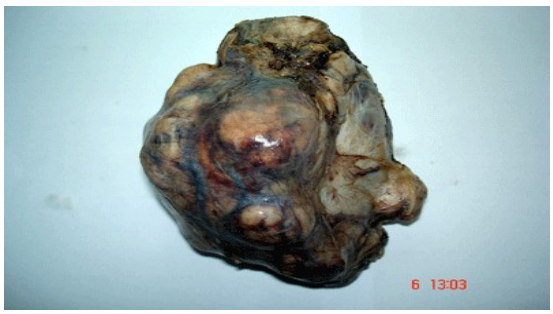

Figure 2: Well encapsulated thymoma with bossulated and smooth surface.

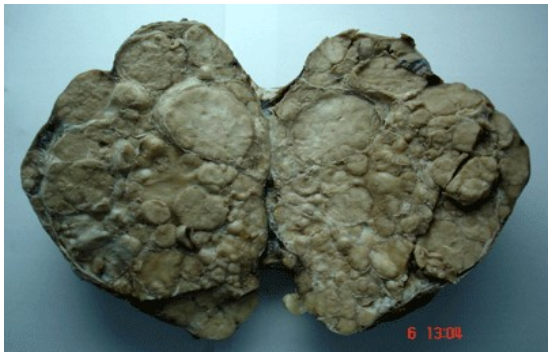

Figure 3: Cut surface of thymoma with zig zag gray white nodules.

Microscopy showed well encapsulated tumor, tumor cells were arranged in sharply defined, angular lobules with motting and trabeculation (caused by epithelial-lymphocyte), epithelial cells were spindly with fine chromatin with occasional mitosis admixed with lymphocytes. There was no evidence of capsular invasion.

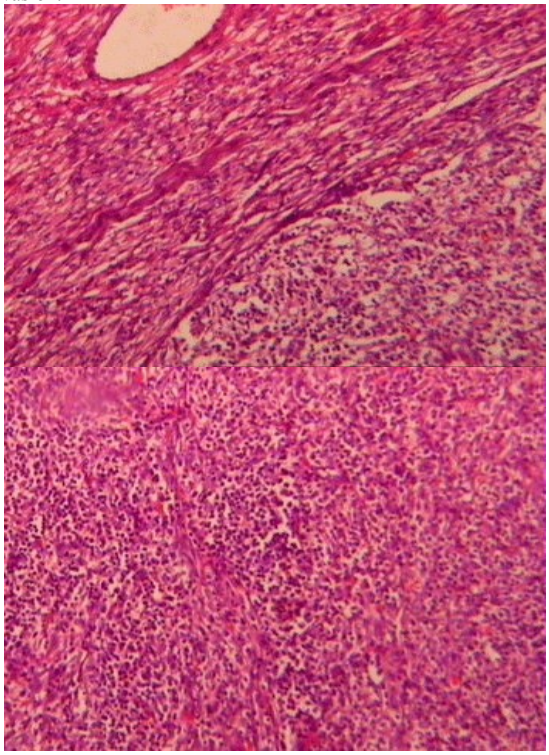

Figure 4,5: Microphotograph 40x Hematoxylin and eosin shows thymoma comprising of admixture of epithelial cells and lymphocytes.

Based on the pathological findings a diagnosis of Thymoma (World Health Organization classification Type AB and Masaoka classification Stage I) was made. Postoperatively patient was continued with treatment for myasthenia gravis and was discharged on 12th post operative day.

Discussion:

The association of thymoma and MG is well described in adults, occurring in anywhere from 10% to 59% of patients in adults. However, it has been stated that this association rarely occur in children. Most mediastinal tumors in the pediatric population are either neurogenic in origin (33%) or lymphomas (41%).[1] Primary thymic lesions (such as thymic cysts, thymolipomas, and thymic hyperplasia) represent only 2.5% of mediastinal tumors [2–4], while thymomas comprise about 1%. Presenting signs are from mass effect in the mediastinum, causing shortness of breath, superior vena cava syndrome, or nonspecific complaints of discomfort.[1,2]

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 World Health Organization recently developed a classification system (Table 1), although most reports follow the general principles outlined by Masaoka et al (Table 2).[6]

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

Table 2: Masoka Classification	
Stage I	Encapsulated tumor with no gross or microscopic invasion
Stage II	Macroscopic invasion into the mediastinal fat or pleura or microscopic invasion into the capsule
Stage III	Invasion of the pericardium, great vessels, or lung
Stage IVa	Pleural or pericardial dissemination
Stage IVb	Lymphogenous or Hematogenous metastases

Thymoma is sensitive both to chemotherapy and radiation, complete surgical resection is the most in a patient with myasthenia gravis or another autoimmune disease, one might consider a complete excision initially, rather than a biopsy, in order to decrease any potential seeding of surrounding tissues.[7,8] The relationship between thymoma and MG remains unclear. Treatment in juvenile MG includes anticholinesterase drugs and prednisone. Thymectomy has been performed on patients who were refractory to drug therapy.[7] 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 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 16-year-old girl with this association.

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