Case report:

Rare case presentation of thymoma

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Abstract:
Thymoma is a tumor originating from the epithelial cells of the thymus. Thymoma is an uncommon tumor, best known for its association with the neuromuscular disorder myasthenia gravis.[1] Thymoma is found in 15% of patients with myasthenia gravis.[2] Once diagnosed, thymomas may be removed surgically. In the rare case of a malignant tumor, chemotherapy may be used. We present a female of 43 years who presented with complaints mainly of giddiness along with occasional cough, chest discomfort. She was obese, non hypertensive, no cervical or other lymphadenopathy was noted. There was little engorgement of neck veins. Haematological investigations and routine urine analysis were normal. There was a small abnormal shadow on the chest X-ray PA view which prompted us to investigate her further. HRCT Thorax was done which indicated the possibility of Anterior Mediastinal Tumour mostly Thymoma. CT guided needle biopsy was done which confirmed our diagnosis. Patient was referred to the higher center where further treatment was available.

Introduction
Thymoma is a tumor originating from the epithelial cells of the thymus. Thymoma is an uncommon tumor, best known for its association with the neuromuscular disorder myasthenia gravis.[1] Thymoma is found in 15% of patients with myasthenia gravis.[2] Once diagnosed, thymomas may be removed surgically. In the rare case of a malignant tumor, chemotherapy may be used.

Illustrative Case
We present a female of 43 years who presented with complaints mainly of giddiness along with occasional cough, chest discomfort. She was obese, non hypertensive, no cervical or other lymphadenopathy was noted. There was little engorgement of neck veins. Haematological investigations and routine urine analysis were normal. There was a small abnormal shadow on the chest X-ray PA view which prompted us to investigate her further. HRCT Thorax was done which indicated the possibility of Anterior Mediastinal Tumour mostly Thymoma. CT guided needle biopsy was done which confirmed our diagnosis. Patient was referred to the higher center where further treatment was available.

Discussion:
Thymomas are rare tumours, with an annual incidence of 0.15 cases per 100,000 person-years, based on data from the US National Cancer Institute Surveillance, Epidemiology and End Results Program. [3] Men and women are affected equally. [4] [5] [6] [7] Thymomas have been reported in infants and also in the very old. The mean age at diagnosis is about 50 years, with a peak broad range between 35 and 70 years. The prevalence of myasthenia gravis in the thymoma population is...
about 30%; 10% to 15% of patients with myasthenia gravis have an associated thymoma. [4] These patients tend to present at a slightly younger age. Thymic carcinomas occur less frequently than thymomas but there are no reliable incidence data. [4] [7] The age distribution is broad, ranging from early childhood to adulthood, with a mean age of 46 years. Most patients present with advanced-stage thymic carcinoma. Myasthenia gravis and other parathymic syndromes are only rarely associated with thymic carcinoma.

Thymic carcinoid (neuro-endocrine) tumours are an extremely rare histological subtype, with only about 200 cases reported. [7] Again, all age groups may be affected. The male-to-female ratio is 3:1. About 30% of patients with thymic carcinoid tumours have associated Cushing's syndrome. Myasthenia gravis and other parathymic syndromes are not associated with carcinoid tumours. The carcinoid syndrome has only rarely been reported in relation to thymic carcinoid tumours. Ninety percent of thymomas and thymic carcinomas occur in the anterior mediastinum. [8] They are the most common malignancies of the anterior mediastinum. [9]

Pathophysiology

Large or invasive thymomas may compress or invade mediastinal or chest wall structures, causing chest pain or cough. Invasive thymomas can involve the phrenic nerve, causing marked dyspnoea, particularly when supine. Rarely, superior vena cava (SVC) syndrome occurs due to compression or invasion of the SVC by the tumour or as a result of a tumour thrombus.

Clinical Features –Thymoma commonly presents as myasthenia gravis or muscle weakness. One-third to one-half of all persons with thymoma have no symptoms at all, and the mass is identified on a chest X-ray or CT/CAT scan performed for an unrelated problem. [1] A third of all people with a thymoma have symptoms caused by compression of the surrounding organs by an expansive mass. These problems may take the form of superior vena cava syndrome, dysphagia (difficulty swallowing), cough, or chest pain. [1] One-third of patients have their tumors discovered because they have an associated autoimmune disorder, the most common of those conditions is myasthenia gravis (MG); 10–15% of patients with MG have a thymoma and, conversely, 30–45% of patients with thymomas have MG. Additional associated autoimmune conditions include pure red cell aplasia and Good's syndrome (thymoma with combined immunodeficiency and hypogammaglobulinemia).

Other reported disease associations are with acute pericarditis, Addison's disease, agranulocytosis, alopecia areata, ulcerative colitis, Cushing's disease, hemolytic anemia, limbic encephalopathy, myocardiitis, nephrotic syndrome, panhypopituitarism, pernicious anemia, polymyositis, rheumatoid arthritis, sarcoidosis, scleroderma, sensorimotor radiculopathy, stiff person syndrome, systemic lupus erythematosus and thyroiditis. [1]
CT scan of the chest revealing a large necrotic mass in the left anterior mediastinum (indicated by the red line). Histology later established the diagnosis of a thymoma.

Another axial slice of a CT scan of the chest showing a small thymoma anterior to the heart (marked with the red line).

When a thymoma is suspected, a CT/CAT scan is generally performed to estimate the size and extent of the tumor, and the lesion is sampled with a CT-guided needle biopsy.

**Treatment**

Surgery is the mainstay of treatment for thymoma. If the tumor is apparently invasive and large, preoperative (neoadjuvant) chemotherapy and/or radiotherapy may be used to decrease the size and improve resectability, before surgery is attempted. When the tumor is an early stage, no further therapy is necessary. Removal of the thymus in adults does not appear to induce immune deficiency. In children, however, postoperative immunity may be abnormal and vaccinations for several infectious agents are recommended. Invasive thymomas may require additional treatment with radiotherapy and chemotherapy (cyclophosphamide, doxorubicin and cisplatin).[1]

**Prognosis and Survival**

Although the oncologic prognosis of thymoma is reported to be more favorable in patients with myasthenia gravis than in patients without myasthenia gravis,[8,10] data are conflicting as to whether the presence of myasthenia gravis is an independent predictor of better outcome. Patients with myasthenia gravis are diagnosed with earlier stage disease and more often undergo complete surgical resection.[10] Treatment with thymectomy may not significantly improve the course of thymoma-associated myasthenia gravis.[11,12]

Thymoma has been associated with an increased risk for second malignancies. In a review of the SEER database of thymoma cases in the United States...
between 1973 and 1998, 849 cases were identified (overall incidence 0.15 per 100,000 person-years).[3] In this study, there was an excess risk of non-Hodgkin lymphoma and soft tissue sarcomas.

References