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Mediastinal Hamartoma Giving Impression of Thymoma in Myasthenia Gravis Patient

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Abstract The mediastinum is a rare localization, as it has been reported in a few cases in the literature for hamartoma, which is a benign tumor. The treatment is surgical resection when an image consistent with anterior mediastinum is observed. In addition, thymoma resection may provide remission in Myasthenia Gravis as it is frequently seen with Myasthenia Gravis and Thymoma. In this case, we submitted our case in which we have resected the anterior mediastinal lesion by applying biportal extended VATS thymectomy in the patient of Myasthenia Gravis with pre-diagnosis of thymoma. Remarkably, the pathology of our case was reported as Hamartoma, and the postoperative six-month follow-up has also indicated regression of Myasthenia Gravis disease.

Keywords: *hamartoma, myasthenia gravis, VATS, thymectomy*

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1. Introduction

Hamartoma, which is usually diagnosed as a solitary pulmonary nodule, is the most common benign tumor of lung [1]. It may contain mixed abnormal tissues such as respiratory epithelium, cartilage tissue, fibrous elements, fat, connective tissue and smooth muscle [2]. Mediastinal hamartoma is very rare [1,2].

The common anterior mediastinal masses are thymoma, lymphoma, germ cell tumor, thymic cysts, parathyroid adenomas and thyroid [1,2]. Myasthenia gravis is seen in 30% of thymoma patients and thymoma is present in 15% of patients with myasthenia gravis [3]. Myasthenia gravis (MG) is an autoimmune disease that causes receptor signal loss of antibodies to Acetylcholine Receptors (AchR) in the neuromuscular junction, leading to clinically weak neuromuscular conduction disorders such as muscle weakness and fatigue [3]. It was reported that Thymectomy is effective in the treatment of myasthenia gravis [4].

2. Case Presentation

The 68-year-old male patient was diagnosed with myasthenia gravis and had complaints of speech disorder, double vision, and weakness four years ago. He was followed up with mechanical ventilator due to tracheostomy in the intensive care unit for four months with the diagnosis of dyspnoea and general condition disorder due to myasthenia gravis. The patient had received intravenous immunoglobulin (IVIG) once a month, 60 mg pyridostigmine bromide (mestinon) four times a day, and 10 mg of Prednisol once a day. In the computed thorax tomography of the patient; the case was evaluated together with the Neurology Department in order to achieve regression in myasthenia gravis symptoms and thymectomy was recommended as soft tissue lesion suggesting 32x17 mm thymoma with containing anterior mediastinum calcifications in anterior mediasten was reported (Figure 1). After IVIG treatment for five days, a bipolaral videothoracoscopic thymectomy was planned.

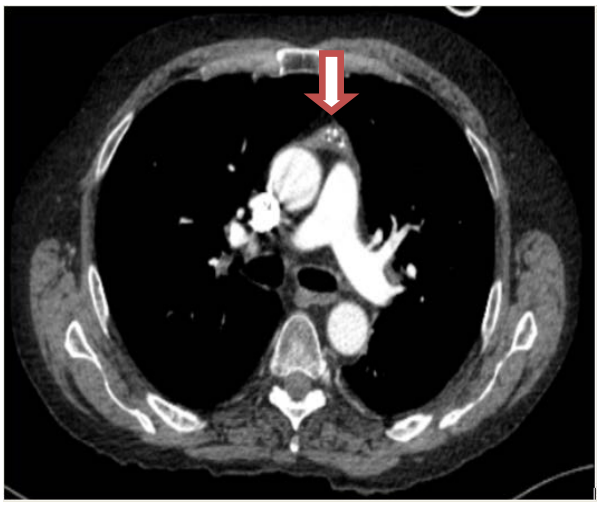

Figure 1. Chest CT shows a 32x17 mm lesion with calcifications at anterior mediastinum

The patient in a supine position with 30° upward angles on the right was applied with incisions of two cm from the

5th right intercostal space mid-axillary line and 1 cm the 7th intercostal space anterior axillary line and thoracoports were placed. A 10 mm and 30 degrees thoracoscope was used for imaging. The lesion in the Thymic tissue was observed adjacent to the aortic arch and bilateral tissues were removed bilaterally between two Phrenic nerves, superior to the Thyroid and the diaphragm in the inferior region. One 24F thorax drain from the lower port was placed in the right thorax.

The patient was followed in the intensive care unit for 24 hours and for 24 hours more in the service in order to prevent side effects of myasthenia gravis and the patient was dehospitalized after removing the thorax drain in the 2nd day after the operation.

The piece's histopathology was reported as "The thymectomy material in the size of 10x7x7 cm, weighing 34 g, is hamartomatoma, consisting of vascular and lipomatous components, no thymus was detected in the removed material." (Figure 2).

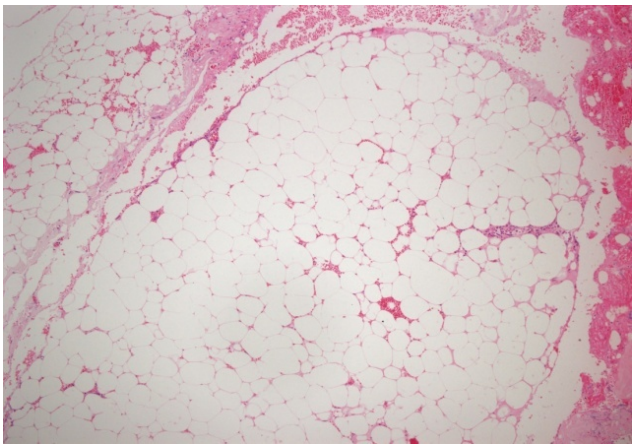

Figure 2. Hamartoma, consisting of vascular and lipomatous components (HEx100)

The patient had expanded PA AC graph in the 4th-month follow-up. Prednisol treatment was not needed in the postoperative period with IVIG treatment in the neurology department and was reduced to 120 mg daily. The patient's mechanical ventilator support requirement decreased to three hours per day. The patient has been followed up for six months without any other problem.

3. Discussion

The lesions observed in the anterior mediastinum are thymic lesions (thymoma, carcinoma, hyperplasia, cysts, timolipoma), germ cell tumors (teratoma, seminoma, embryonal cell cancer, choriocarcinoma) lymphoma, thyroid related lesions, and parathyroid lesions [1,2].

Hamartom is a malformation seen in the form of unorganized tissue mass in abnormal order and quantity in a specific area (epithelium, cartilage tissue, fibrous elements, fat, connective tissue and smooth muscle, etc.) [1,2]. Hamartoma is the most common benign lesion of the lung with 70% of all benign lung tumors [1,2,3]. However, the mediastinum is not an expected localization for hamartoma [1,2]. Hamartomas are radiologically seen as flat, lobulated, well-circumscribed, peripheral lesions with dimensions ranging from 1 to 3 cm in the lower areas of the lung and with increasing diameter of 3-5 mm per year [1,2]. In thorax CT, 5-50% of the cases have calcification in the form of popcorn, and 50% of them have fat tissue. Without dominance of gender, it is mostly observed at 40-50 years of age. Treatment in the lung parenchyma is made with excision of the hamartoma.

Myasthenia gravis is an autoimmune disease with abnormal neuromuscular transmission caused by antibodies to muscle acetylcholine receptors or muscle-specific tyrosine kinase antibodies in the postsynaptic membrane. There are two forms as "ocular MG" where the weakness is limited to the eyelids and extraocular muscles and "generalized MG" where bulbar, extremity, respiratory muscles are affected. Thymus gland abnormalities in patients with MG are seen as thymoma in 10-15% of cases and lymphoid follicular hyperplasia in 70% of cases [3,4,5]. Thymoma, which consists of thymic lesions and contains epithelial cells and reactive lymphocytes, is most frequently seen in the anterior mediastinum. The mean age of diagnosis of thymoma cases is 50 years. In patients with MG, the most sensitive method for thymoma is thorax CT and it is usually seen as located above the right ventricular outflow tract and properly localized or lobulated in front of the ascending aorta or sometimes in the distal mediastinum or at the cardiophrenic angle. In addition, it can exhibit homogeneous density enhancement, as well as cystic components, spot or curvilinear or round calcifications. MG treatment can include symptomatic, immunosuppressive/immunomodulatory agents and thymectomy protocols in patients with thymoma MG. It has been reported that the thymectomy applied to the patients of myasthenia gravis without thymoma is beneficial, and good results are obtained in all generalized MG patients in the first years of the disease regardless of whether they have thymoma [5]. According to a multi-centered and randomized study, it was revealed that the patients with myasthenia gravis who underwent thymectomy had better clinic results, less immunosuppressive needs and less need for hospitalization at the end of three years [5].

Our case had been followed for generalized myasthenia gravis for four years and had generalized weakness, Ptosis in the right eye and respiratory distress. As a result of the recurrent lung infections in the last year, the long-term mechanical ventilator was connected by opening tracheostomy with the continuous requirement of the mechanical ventilator. The lesion at the anterior mediastinal localization containing calcifications detected in Thorax CT was interpreted in the favor of thymoma as the patient also had myasthenia gravis. Biportal VATS extended thymectomy, which is the minimally invasive method, was performed and the patient, who did not have postoperative complications, was discharged on the 2nd day. However, the histopathological result was surprisingly found as hamartomatous lesions composed of vascular and lipomatous components without thymus tissue. In the postoperative 4th month of the patient, who did not have thymus tissues in the resection materials, the need for mechanical ventilator decreased from 24 hours to three hours, the Prednol treatment was stopped and the Pyridostigmine treatment decreased from 240 mg to

120 mg a day, which means that there is remission in myasthenia gravis symptoms. The case is a rare case due to the fact that hamartomatous lesion is seen in the anterior mediastinum as well as the hamartomatous lesion causes regression of myasthenia gravis symptoms in the resection.

As a result, anterior mediastinal located hamartoma is rarely seen. The resection applied in the patient with myasthenia gravis may have a role in the treatment of myasthenia gravis, even though the normal tissue components of the thymus are not seen in the lesion. Further studies should be done for the mechanisms of this effect and the series of cases should be examined rather than individual case reports.

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