

CASE REPORT

A Giant Thymoma Resection with Mini-Sternotomy in a Patient with Ankylosing Spondylitis: A Case Report

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ABSTRACT

Although thymoma is the most common primary tumor in anterior mediastinum, it accounts for less than 1% of tumors in adults. Majority of the patients are asymptomatic and identified in imaging methods by chance. Thymoma is frequently associated with paraneoplastic syndromes and myasthenia gravis is observed in nearly 40% of the thymoma patients. Dyspnea, dysphagia, or superior vena cava syndrome refer to symptoms associated with tumor size and are observed in 40% of all thymoma patients. Successful treatment of thymomas, be they invasive or non-invasive, depends on complete resection if possible. Nefrotic syndrome is generally linked to long-term steroid therapy in patients who had undergone surgical excision for thymoma. In this article; a 42 year old male patient with ankylosing spondylitis who was operated for mediastinal thymoma, is presented. Following a successful mini sternotomy, total excision of the tumor was made and the patient was safely discharged from the hospital.

Key Words: Thymoma; Ankylosing spondylitis; Nefrotic syndrome.

Ankilozan spondilitli hastada mini-sternotomi ile dev timoma rezeksiyonu: Olgu Sunumu

Timoma ön mediastende en sık görülen primer tümörü olmasına rağmen, erişkinlerde tümörlerin %1'den azını oluşturur. Hastaların büyük çoğunluğu asemptomatik olup yapılan görüntüleme yöntemlerinde tesadüfen saptanırlar. Timoma paraneoplastik sendromlarla sık birliktelik gösterir yaklaşık olarak tüm timomalı hastaların %40'ında myasthenia gravis izlenir]. Dispne, disfaji veya vena cava superior sendromu tümörün büyüklüğüyle ilgili semptomlar olup tüm timomalı hastaların %40'ında görülür Timomaların başarılı tedavisi (invaziv ve non-invaziv) eğer mümkünse komplet rezeksiyona bağlıdır. Timoma nedeniyle cerrahi eksizyon uygulanan hastalarda nefrotik sendrom, uzun süreli steroid kullanımına bağlıdır. Bu yazıda, mediastinal timoma nedeniyle opere edilen ankilozan spondilitli 42 yaşında erkek bir olgu sunulmuştur. Mini sternotomi ile tümör total olarak eksize edilmiş ve hasta taburcu edilmiştir.

Anahtar Kelimeler: Timoma, ankilozan spondilit, nefrotik sendrom.

INTRODUCTION

Although thymoma is the most common primary tumor in anterior mediastinum, it accounts for less than 1% of tumors in adults (1,2). Dyspnea, dysphagia, or superior vena cava syndrome refer to symptoms associated with tumor size and are observed in 40% of all thymoma patients (3). Complete resection alone is capable of treating most of the cases with mediastinal mass and is the method that should be considered as the primary treatment of life-threatening tumors compressing large vascular structures and large airways such as trachea and main bronchi (4,5). The success of thymoma treatment to a great extent depends on complete resection (6). The first transsternal thymectomy was performed by Alfred Blalock in 1939 and since then the standard surgical approach has been the median sternotomy (6,7).

CASE REPORT

A 42-year-old male patient with complains of dyspnea on exertion, chest pain and dry coughs was admitted to our polyclinic. The patient had a history of ankylosing spondylitis and had been receiving steroid treatment for a long time. He was evaluated for dyspnea at the cardiology clinic and no cardiac pathology was detected. Due to his complains of coughs and chest pain, the patient was referred to our thoracic surgery clinic on his request.

The physical examination revealed no significant finding. The PA chest x-ray demonstrated no other finding than mediastinal enlargement (Figure 1). The blood tests yielded no abnormal finding. A non-contrast thoracic computed tomography (CT) was performed to investigate the mediastinal enlargement. The non-contrast thoracic CT revealed a mass lesion of 10x5 cm in the homogeneous internal structure with lobular contour projecting through the right paracardiac region in the

anterior mediastinum and compressing the neighboring vascular structures or trachea and bronchi (Figure 2). This view was considered to be significant in consideration of thymoma by the radiologist reporting the CT.

A PET/CT was scheduled for the patient by the authors (Figure 3). The case was reported as a mass formation filling up the retrosternal area and right prevascular space in the anterior mediastinum, stretching till the diaphragmatic level, having an axial diameter of nearly 42x69 mm at the thickest part and a length of nearly up to 95 mm on the craniocaudal axis, with a lobular contour, seemingly closely related to pericardial and other mediastinal structures and characterized by increased FDG uptake (SUDmaks=6.1). Moreover, ankylosis in the intervertebral disc spaces in columnae vertebralis, bilateral sacroiliac joints, and bilateral first sternocostal joints substantiated the patient's history of ankylosing spondylitis. A CT-guided transthoracic fine-needle aspiration biopsy (TFNAB) was administered to the patient by the authors. Based on the result of the pathological analysis, a tissue biopsy was recommended to the patient who was observed to have epithelial cell clusters with a small number of atypias between lymphocytes.

Following the conduction of the required preoperative examinations, a cardiovascular surgery specialist was referred to as the mass contacts the innominate vein in particular. Taking the median sternotomy into account, a total resection of the mass was planned.

The mass in the mediastinum was totally resected by employing mini-sternotomy under general anesthesia. The innominate vein was crossed with the mediastinum and so entangled by the mass during the course where it flows to the superior vena cava as not to allow for exploration. Therefore, the innominate vein

was tied by a cardiovascular surgeon and excised together with the mass. Then, the proximal and distal ends were tied and the transfixation sutures were placed.

The intubated patient was monitored in the general intensive care unit after the operation and then extubated within hours. On the second postoperative day at the hospital, an acute nephrotic syndrome occurred in the patient. The development of nephrotic syndrome in the patient operated on because of thymoma was associated with his long-term steroid use for the treatment of ankylosing spondylitis and his present condition. In the related literature, there are studies reporting the development of nephrotic syndrome in patients with thymoma and having used immunosuppressive drug for a long time.

The patient who was intubated on the fourth day after the operation were discharged without an issue on the eighth day. The patient who was observed to develop pleural effusion occasionally was monitored with an anti-inflammatory and therapeutic thoracentesis treatment and then referred to the oncology clinic (Figure 4).

In the report of the material sent for pathological analyses, no rupture was revealed on the capsule with a diameter of 11.5 cm at the thickest part and no infiltration was observed in the adipose tissue, and it was stated that it was congruent with the thymic neoplasia. However, it was found to be congruent with invasive thymoma in consideration of the results concerning the material sent for analysis consultation to a senior center for the inspection of invasive thymoma or thymic carcinoma. Moreover, the report also mentioned innominate vein excised together with the mass as a tissue with a size of 4x3.5x2 cm, which was construed to be the vascular structure in the sent material.



Figure 1. The patient's preoperative PA chest x-ray



Figure 2. The mediastinal mass (arrow) in the patient's thoracic CT



Figure 3. The Patient's PET/CT imaging



Figure 4. The patient's postoperative PA chest x-ray. Pleural effusion observable on the right.

DISCUSSION

Thymus is a lymphoid organ playing a critical role in the maturation of lymphocytes and cellular immunity. Embryologically it originates from the third and fourth pharyngeal pouches. During the embryogenic migration, thymic tissue fragments or accessory lobes erroneously locate to the cervical region (4%) or the middle mediastinum (1,8-10). Thymoma is present typically in the fourth and fifth decades of life and exhibits no gender predilection (11). It is annually observed in 0.15 out of 100000 cases (2). 90% and 6% of thymoma cases occur in the anterior-superior and superior mediastinum, respectively, while the remain rates are observable in inferior mediastinum. In the series reported by Cohn, 2 of 70 thymoma cases were identified in anterior-superior mediastinum (12). Majority of the patients are asymptomatic and identified in imaging methods by chance. Thymoma is frequently associated with paraneoplastic syndromes and myasthenia gravis is observed in nearly 40% of the thymoma patients (13,14). Clinical symptoms may vary across patients suffering from thymoma. The associated symptoms are persistent cough, dyspnea and other upper airway findings (7). Large masses generally grow out of the mediastinum compartment they originate from towards the other compartments and then keep spreading towards only one of or both hemithoraces (4). By growing larger, they produce the symptoms through invasion or by compressing the adjacent vascular structures, large airways, and esophagus (5). Half the thymoma patients occur asymptotically and detected incidentally by radiographic imaging methods (11). Following the identification of the mediastinal mass on the chest radiography, a scanning is conducted with computed tomography. Computed tomography offer some information about the character of tumor and peripheral tissue invasion. Computed tomography with intravenous contrast is the preferred method to evaluate the vascular invasion and cystic components of a tumor (6). Dynamic magnetic resonance imaging (MRI) is a method for staging and differential diagnosis. Positron emission tomography is employed in tumor scanning and differential diagnosis of invasive and non-invasive thymoma. In addition to these diagnostic methods, a diagnostic-focused transthoracic fine-needle

aspiration biopsy (TFNAB) was administered to the patient. Successful treatment of thymomas, if it is invasive or non-invasive, depends on complete resection if possible. Regardless of the diagnosis in the preoperative period of large mediastinal tumors, radiotherapy can be employed to ameliorate or remove serious compression symptoms. Complete resection is the most important determinant in long-term survival, yet radiotherapy or chemotherapy or both combined ameliorate prognosis and survival (10). Although thymoma responds to radiotherapy and chemotherapy, surgical resection is the mainstay of treatment (7). Non-operable patients should be re-evaluated for surgical treatment after induction chemotherapy. Thymoma is classified by its histopathological and clinical properties. According to the available literature, adjuvant radiochemotherapy has not proven fruitful for completely resected stage one thymoma. Multimodality treatment including induction chemotherapy, surgery, and adjuvant radiotherapy were revealed beneficial in the treatment of primary unresectable thymic tumors.

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