Diagnosis and Management of True Thymic Hyperplasia; Description with Cases in Two Sisters

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True Thymic Hyperplasia / Gerçek Timik Hiperplazi

Özet

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Timus; Mediastinal Tümörler; Genetik

Abstract
Thymic hyperplasia is an extremely rare neoplasm of thymus which is localized in the anterior mediastinum. Generally it occurs secondary to the different diseases. Some of them occurs idiopathically. Until today there are more than 50 cases reported and most of them are children between the ages of 1 and 17. A few cases are adults. But until today there is no reported case with the same brothers and sisters. Therefore we want to point on whether thymic hyperplasia can be hereditary or not.

Keywords
Thymus; Mediastinal Neoplasms; Genetics
Introduction
Thymic hyperplasia is an extremely rare neoplasm which is localized in the anterior mediastinum [1,2]. Etiology and prognosis of thymic hyperplasia is not well defined. Fewer than 50 cases have been reported until today. Most of the patients are infants and children between the ages of 1 and 17 [2-5]. True thymic hyperplasia is rarely observed in adults. It occurs twice as often in boys than girls [6,7]. In our report our patients are two sisters and both of them submitted with the same symptoms such as cough, respiratory distress, shortness of breath and often pulmonary infections. And both of their computed tomography (CT) findings were huge masses that were filling the mediastinum in large areas. After a surgery for total excision of masses, the histopathological examinations were adjusted with idiopathic true massive thymic hyperplasia.

Case Presentation
Case 1
A 4 years old girl had submitted to a pediatrics clinic with the complaints of cough, respiratory distress, shortness of breath and 39 degree persistant fever. All the laborotory examinations were normal except lymphcytosis in the blood analysis. According to the posteroanterior chest X-ray of her first submit, nearly whole of the left lung was consolidated, so she was diagnosed with pneumonia (Figure 1:A). Medical treatment had started but despite treatment clinical and radiological improvement had not provided. Despite all the researches her disease could not be diagnosed and due to increasing respiratory distress she was referred to our thoracic surgery clinic for the purpose of both diagnosis and treatment.

In the physical examination in our clinic, left lung sound was characterized with rhoncus and rales. So we planned a thorax CT for diagnosis. CT showed a huge mass was filling the anterior mediastinum and huge part of left hemithorax in a solid form (Figure 1:B). For a further analysis abdominal ultrasonography (USG) and cranial CT were seen and both of them were normal. Fine needle aspiration biopsy was suggested for diagnosis but her parents were not agreed because patients respiratory distress had increased and would lose time waiting for pathology results. Surgery was performed for both diagnosis and treatment as advised, especially to relief the mediastinal compression. Left posterolateral thoracotomy was preferred because the mass was generally in the left side. The approach to the mass for total excision with left thoracotomy was easier than median sternotomy. During the operation, we saw that the mass had filled nearly whole of left hemithorax and adhered to the arround tissues, and the left lung was consolidated because of compression. The mass was in the dimension of 16x14x9cm. Mediastinal mass composed entirely normal thymic tissue. The mass was excised totally.

The histopathological examinations showed that the mass contained well developed cortical areas, thymic medulla and Hassal’s corpuscules. Thymic cortex was divided in to the lobules by septas. The septa extends to the corticomedullary junction (Figure 2:A). Hassal’s corpuscles expressed high molecular weight cytookeratin cortex contained CD1 a positive T cells. CD 20 positive B cell component was largely confind to the thymic medulla (Figure 2:B). B cells were surrounded with Hassals corpuscules.

There was no problem on her postoperative control posteroanterior chest X-ray (Figure 1:C).

Case 2
Our first patient’s sister submitted to a different hospital with cough, respiratory distress, 39,5 degree fear and shortness of breath, almost the same complaints with her elder sister when she was 6 months old in 2007. Her posteroanterior chest X-ray showed consolidation on bilateral upper lobes, right lobe and lingula (Figure 3:A). Bilateral lung sound was correlated with rales and rhoncus. First diagnose was pneumonia. So, medical treatment had started but despite treatment clinical and radiological improvement had not provided. So she had referred to our hospital. We thought that all of her complaints could be secondary to a mediastinal mass such as her elder sister. And also we took care whether that mass was hereditary or not. So we immediately planned a thorax CT (Figure 3:B). CT showed a huge mass that was filling the both lung parancymal distances begining from anterosuperior compartment of mediastinum and surrounding the hearth. Abdominal USG and peripheral blood analysis were normal. We immediately planned surgery, our choise was median sternotomy that time. Because the mass was filling both hemithorax and it was easier to aproach to the both side masses. Surgery was performed and the mass

Figure 1. Preoperative posteroanterior chest X-ray showed nearly whole of the left lung was consolidated(A). Preoperative thorax CT scan showed a huge mass that filling the anterior mediastinum and a great part of left hemithorax in a solid form(B). Postoperative posteroanterior chest X-ray(C).

Figure 2. Histopathological studies: Normal thymic tissue which contains well developed cortical areas, thymic medulla and Hassal’s corpuscles. Thymic cortex was divided in to the lobules by septas. The septa extends to the corticomedullary junction (A). Hassal’s corpuscles expressed high molecular weight cytookeratin cortex contained CD1 a positive T cells. CD 20 positive B cell component was largely confined to the thymic medulla (B).

Figure 3. Preoperative posteroanterior chest X-ray showed consolidation on bilateral upper lobes, right lobe and lingula (A). Preoperative CT scan showed a huge mass that is filling the both lung parancymal distances begining from anterosuperior compartment of mediastinum and surrounding the hearth (B). Postoperative posteroanterior chest X-ray (C).


excised totally. There was no problem on her postoperative control posteroanterior chest X-ray graph (Figure 3:C). The excised mass was in the dimension of 14x7x5 cm and in the weight of 160 grams. Its surface was grey-white colored and lobulated (Figure 4). All the histopathological study findings were adjusted with Idiopathic True Massive Thymic Hyperplasia, and all of lymph nodes were reactive. During postoperative controls, there were no complications. All these results directed us to another baby of this family whom is a boy and was born from another mother. We looked for such a mediastinal mass. But he was safe. Our patients were safe in their postoperative controls and all of their preoperative complaints were cured.

**Discussion**

Thymic hyperplasia is a rare cause of anterior mediastinal mass in children [1-3]. Etiology and prognosis of thymic hyperplasia is not well defined [2-5]. It is separated in two categories such as lymphoid or follicular hyperplasia that characterised with activated germinal centers and lymph follicles. The other is True thymic hyperplasia which has a normal thymic architecture and normal germinal centers and lymph follicles expected for age and occurs in two forms as rebound thymic hyperplasia and idiopathic true massive thymic hyperplasia [6,7]. Rebound hyperplasia occurs after especially graves disease, Cushing Syndrome, burns, after steroid therapy, association with endocrine abnormalities, testicular tumors, sarcoidosis, lymphoma, and Beckwith Wiedeman Syndrome [8]. Idiopathic true thymic hyperplasia has a well defined clinicopathological profile prevalence in children or young patients, absance of associated autoimmune disease and often presence of cough, respiratory distress, shortness of breath, peripheral blood lymphocytosis. All of these patients have normal immun systems. CT generally shows a mass located in anterior mediastinum. There have been cases reported about rebound massive thymic hyperplasia generally secondary to the non hodgkin lymphoma, hodgkin lymphoma and after chemotherapy. And also there are a few reported cases of idiopathic true massive thymic hyperplasia especially in children and young patients. [8]. Treatment is surgical removal of the thymic mass. Linegar and coworkers suggested median sternotomy, clamshell incision or single-sided posterolateral thoracotomy as surgical approach.

When we examine the literature mentioned with multiple cases, we did not detect familial intimacy or brotherhood in any of them. However, that is strange that our cases are two sisters and they have the same complaints, both of them were operated and both of their histopathological study results were idiopathic true massive thymic hyperplasia form. We want to point on whether it can be hereditary or not.

**Competing interests**

The authors declare that they have no competing interests.

**References**

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