Case Report / *Olgu Sunumu*

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Type B3 Thymoma in a Twelve-Year-Old Girl

On İki Yaşında Bir Kızda B3 Tipi Timoma

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Abstract

Thymic epithelial tumors consist of thymomas, thymic carcinoids, and thymic carcinomas. Most cases occur over the age of 20 years, and 70%of cases in patients over 40 years of age. Thymomas are extremely uncommon in children, accounting for less than 1%of childhood mediastinal tumours compared with 30%in adults and, because of their rarity, may be difficult to diagnose. In this paper, we present a 12-year-old girl who presented with symptoms of pneumonia such as cough, fever and chest pain. Her chest x-ray showed expansion of the mediastinum and opacity in the right hemithorax. An anterior mediastinal mass was determined by chest computed tomography. Histological and immunochemical examination of the surgically excised mass showed type B3 thymoma. The case is presented for its rarity in children. *(Haseki Tıp Bülteni 2013; 51: 200-02)*

Key Words: Childhood, mediastinal mass, thymoma

Özet

Timik epitelyal tümörler; timomalar, timik karsinoidler ve timik karsinomaları kapsamaktadır. Hemen hemen tamamı 20 yaş üstü olan olguların %70'i 40 yaş üstüdür. Mediastinal tümorler içinde erişkinlerdeki %30'luk oran ile karşılaştırıldığında çocuklarda %1 oranındaki görülme sıklığı tanı koymayı güçleştirir. Biz burada öksürük, ateş ve göğüs ağrısı gibi pnömoni bulguları ile gelen 12 yaşında bir kız çocuğunu sunduk. Akciğer grafisinde mediastinal genişleme ve sağda opasite görülen hastanın bilgisayarlı tomografisinde kitle saptandı. Cerrahi olarak çıkarılan kitlenin histolojik ve immünohistokimyasal incelemeleri B3 timoma olduğunu gösterdi. Olgu çocuklarda timomanın nadir görülmesi nedeni ile sunulmuştur. *(The Medical Bulletin of Haseki 2013; 51: 200-02)*

Anahtar Kelimeler: Çocukluk çağı, mediastinal kitle, timoma

Introduction

Thymomas are neoplasms of the anterior mediastinum. The peak incidence of thymoma is usually in the fifth and sixth decades of life (1). These tumors behave more aggressively in children than in adults. It has a much more rapid course and a poor prognosis. Patients usually present with respiratory symptoms, superior vena cava syndrome, or paraneoplastic syndrome including myasthenia gravis, pure red cell aplasia (2). Herein, we report the case of thymoma in a child who presented with signs of pneumonia.

Case Report

A 12-year-old girl who presented with chest pain, fever and cough was admitted to our hospital. She had been suffering from these complaints for the last three days. The patient was diagnosed with pneumonia at another health center and treated for three days with antibiotics, but her symptoms had not improved. The patient was an otherwise healthy child. Her past medical history was unremarkable. She had no dysphonia, dysphagia or weight loss.

On physical examination, she had a temperature of 37.7°C and respiratory rate of 24/minute. Auscultation of the chest demonstrated decreased breath sounds at the right lung base, with inspiratory rales Other physical examination findings were normal. Laboratory findings were: Hb - 12.2 g/dL; Hct - 38.6%; white blood cell (WBC) - 15200/mm^3; platelet - 297000/mm^3; erythrocyte sedimentation rate (ERS) - 53 mm/hr, and C-reactive protein (CRP) levels - 12.4 mg/dL (N:0-3). Her urine analysis, serum biochemical tests, blood gases, serum levels of tumor markers (beta-human chorionic gonadotrophin and alpha-fetoprotein) and serum lactate dehydrogenase were all normal. Chest X-ray revealed mediastinal widening and opacity in the right anterior hemitorax (Figure 1). Echocardiography was unremarkable. She was started

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on antibiotics. Clinical response to medical therapy was not satisfactory. Thus, a computerized tomography (CT) scan of the thorax was performed and it showed a right anterior mediastinal mass measuring 10x11 cm in diameter, which appeared to be heterogenous with solid and cystic components. The differential diagnosis on CT was a mature teratoma and thymoma. Total excision of the tumoral mass was performed by a median sternotomy. The mass and surrounding fatty tissue were resected together. The tumor was a smooth, irregular lobulated, ovoid mass measuring 13x7x4.5 cm, weighing 200 grams. It contained an irregular pseudocystic area measuring 6x3x2 cm. Capsular, neighboring pericardial and pleural invasions were seen. Immunhistochemically, the neoplastic cells were positive for EMA at the epithelial component. CD5 was positive at lymphocytes associated tumor. Ki-67 score was high. Based on histopathological findings, the tumor was classified as a type B3 (cortical) thymoma according to the World Health Organization (WHO) classification and she had Masaoka stage IVa. The postoperative course was uneventful. She was referred to the pediatric oncology unit to begin a chemotherapy protocol.

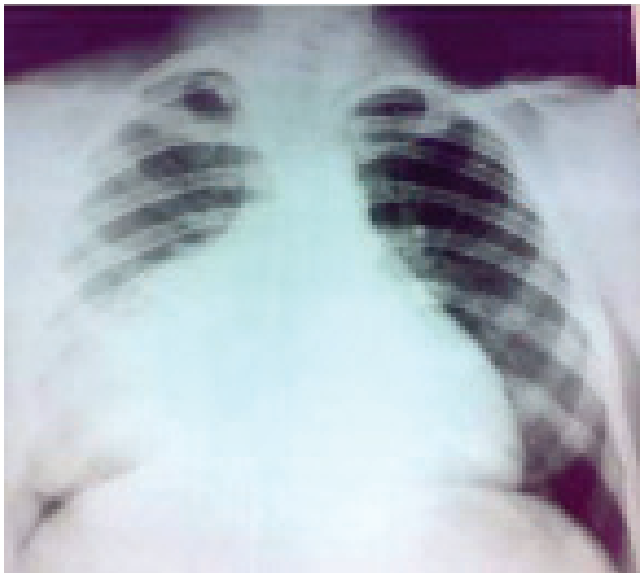

Figure 1. Chest X-ray showing mediastinal widening and an opacity in the right anterior hemitorax

Table 1. Thymoma staging according to Masaoka et al. (6)	
Stage I	Totally encapsulated
Stage II	Capsular invasion and/or invasion into surrounding fat or pleura
Stage III	Invasion into organs (pericardium, lung, great vessels)
Stage IV- A	P leural or pericardial implants
Stage IV-B	Haematogenous metastases

Discussion

In children, the mediastinum is the site of 1%of all tumors; the most common ones are lymphomas (41%), neurogenic tumors (33%), and germ cell tumors (19%-25%), whereas thymomas are exceedingly rare (1%) (3). Approximately 32 cases have been reported in the pediatric literature in the past 30 year (2). Takeda et al. (4) reported 130 cases of primary mediastinal tumors in children and there were only 5 pediatric patients (4%) with thymoma. Type B3 thymoma is relatively rare (6%-17%of all thymomas) and occurs chiefly in adults (mean: 41-47 years) (5).

In 1999, the WHO classified thymic epithelial tumors (TET) histologically into types A, AB, B1, B2, B3, and type C, which are all thymic carcinomas. The 2004 update further accommodates identified genetic alterations in TETs (5). Staging of thymomas is most commonly based on the Masoka classification (Table 1) (6). Type A, type AB and type B1 thymomas are encapsulated and benign tumors. However, B3 thymomas and thymic carcinomas (type C) tend to be clinically malignant.

Thymomas are usually detected incidentally on routine chest x-ray, however, they may also be symptomatic, resulting in cough, chest pain, dyspnea, dysphagia, hoarseness, or recurrent respiratory infection. Superior vena cava obstruction, Horner's syndrome and weight loss are rare manifestations (1). Although development of a thymoma in childhood is rare, children are more likely than adults to have symptoms. Our case presented with chest pain, cough and fever. Firstly, pneumonia was considered.

The parathymic and paraneoplastic syndromes are frequently associated with thymomas in adults, such as myasthenia gravis (44% of cases) (1). Myasthenia gravis and other paraneoplastic syndromes are rarely associated with thymoma in children. Only ten cases of myasthenia gravis have been reported in the pediatric literature (7).

Chest radiographic manifestations of thymoma range from subtle mediastinal contour abnormalities to a large anterior mediastinal mass. As many as 25% of thymomas are not detected by plain radiography. Thus, CT is the main imaging technique for the evaluation of thymomas (8). In our case, plain radiographs showed opacity in the right hemithorax which suggested the presence of a pneumonic infiltration or a mass. CT confirmed the existence of a mass of possible thymic origin.

Treatment options include complete surgical resection, radiotherapy and chemotherapy. The treatment

strategy depends on the clinical stage of the thymoma. Surgical resection is chosen for stage I and II tumors and chemotherapy is preferred in patients with stage-III or -IV tumors. Adjuvant radiotherapy is performed in patients with tumors larger than 5 cm. The tumor has a good prognosis if it is well encapsulated and completely resected. Prognosis for invasive or metastatic disease is poor (9). Careful intraoperative inspection and accurate histopathologic evaluation are very important for correct staging and treatment. In our patient, we observed capsular invasion and local invasion of the tumor into surrounding fatty tissue, pleura and pericard. Therefore, the patient was diagnosed as stage IVa and complete resection was performed. She was referred to the oncology unit at another hospital for chemotherapy, however, later, she was lost to follow-up.

In conclusion, thymoma is very rare in children. Imaging and clinical findings may potentially be misinterpreted as pneumonia and other respiratory diseases. B3 thymomas behave aggressively, and the prognosis is poor. However, correct and early diagnosis increases the chance of favorable treatment.

References

1. Aghaji MA, Uzuegbunam C. Invasive thymoma and myasthenia gravis in a three-and-a-half-year-old boy: case report and literature review. Cent Afr J Med 1990;36:263-6.
2. Liang X, Lovell MA, Capocelli KE, et al. Thymoma in children: report of 2 cases and review of the literature. Pediatr Dev Pathol 2010;13:202-8.
3. Chaturvedi M, Kalgutkar AD, Khandekar JV. Extensive ossification of thymoma in a child. Ann Thorac Surg 2013;95:139-41.
4. Takeda S, Miyoshi S, Akashi A, et al. Clinical spectrum of primary mediastinal tumors: a comparison of adult and pediatric populations at a single Japanese institution. J Surg Oncol 2003;83:24-30.
5. Park MS, Chung KY, Kim KD, et al. Prognosis of thymic epithelial tumors according to the new World Health Organization histologic classification. Ann Thorac Surg 2004;78:992-8.
6. Masaoka A, Monden Y, Nakahara K, Tanioka T. Follow-up study of thymomas with special reference to their clinical stages. Cancer 1981;48:2485-92.
7. Nikolic DM, Nikolic AV, Lavrnic DV, Medjo BP, Ivanovski PI. Childhood-onset myasthenia gravis with thymoma. Pediatr Neurol 2012;46:329-31.
8. Rothstein DH, Voss SD, Isakoff M, Puder M. Thymoma in a child: case report and review of the literature.Pediatr Surg Int 2005;21:548-51.
9. Iorio R, Evoli A, Lauriola L, Batocchi AP. A B3 type-thymoma in a 7-year-old child with myasthenia gravis. J Thorac Oncol 2012;7:937-8.