CHEST PAIN DUE TO A GIANT THYMOMA IN AN ADOLESCENT BOY

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Abstract

Chest pain in children is a common referral for emergency examination, although it is believed that the underlying condition is not a serious event. The authors present the case of a 17 years old male with complaints of chest pain with a very recent onset, constant, experienced after physical effort. All medical history and physical examination findings were normal, the poster-anterior chest X-ray revealed a heterogenous opacity in the middle third of the right hemithorax confirmed on computerized tomography as a large solid anterior mediastinal mass with a diameter of 7.5/10.3 cm. By thorax surgery the mass was identified in the anterior and superior mediastinum and total resection was achieved for the tumor mass and thymus. Pathology examination showed poor small and medium epithelial cells proliferation and the tumor was classified as thymoma type 1B. Mediastinum is a rare location of space occupying processes in children. Also, there is a high rate of asymptomatic lesions in this area, considering that half of them are incidentally discovered for various chest X ray examinations. Many causes of chest pain in children are benign; nonetheless, some serious events do exist sometimes and pediatricians must pay attention to identify and manage those cases.

Key words: thymoma, chest pain, children.

INTRODUCTION

Chest pain in children is a common referral to any Emergency Department, although it is widely believed that the underlying condition is not a serious or life threatening event as it is in adults (1, 2). Major causes of chest pain in pediatric population are benign idiopathic, musculoskeletal, gastrointestinal, pulmonary or psychogenic, while cardiac issues are quite rare, as many studies have shown (3, 4). Even so, chest pain in children, especially adolescents, has a dramatic perception in the population, considering that in one study, almost 70% of patients seek for medical advice because they were worried about

having heart attacks and 44% of them changed their behavior because of it (5). Pediatricians often face parental concerns and need to adopt a more invasive attitude in order to reveal the cause. Acquired abnormalities such as thoracic lesions might be in incidental radiological reveal; out of these, thymomas are rare and slow-growing neoplasms, occurring mainly in adult age. In children, primary thymic lesions are seen in no more than 1% of all mediastinal masses (6). About one third of this pathology is asymptomatic and recognized only by incidental radiology examination (7). It occurs with a slight male predominance and therapeutic attitude needs further prospective data in order to develop a unitary guideline (8). European Cooperative Study Group on Pediatric Rare Tumors (EXPeRT) coordinates the activities of five different national rare tumour groups in Europe including France, Germany, Italy, Poland and the United Kingdom and reported only 36 cases of thymic tumours treated between 2000 and 2012 in paediatric patients (9).

OBJECTIVE

We present the case of a 17 years old male adolescent presented with complains of chest pain with a very recent onset, constant, located in the anterior thorax.

SUBJECT AND METHODS

A 17-year-old boy was addressed to our “Grigore Alexandrescu” Emergency Children Hospital for chest pain located in the right anterior hemi thorax going to cervical zone and homolateral shoulder and arm, experienced after physical effort, sharp, constant pressure over the right chest with the intensity rating of 8 out of 10.

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RESULTS

There are no worthy data regarding the patient past medical history of chest pain or syncope. Family history was negative for cardiovascular disorders, like acute coronary syndromes at young ages or sudden cardiac death. He declared he has a daily sport activity, practicing force gym for 3 hours a day. Also, patient denied any substance abuse. All physical examination findings were normal, with a normal S1S2, no murmur, gallop, rub or distended jugular veins; chest wall tenderness was absent, so as hepatomegaly. Neurologic examination was within normal ranges, so as complete blood count and biochemistry panel.

The initial poster-anterior chest X-ray revealed large mediastinum due to a heterogenous opacity in the middle third of the right hemithorax, closely to the heart but without lung infiltration, having no mass effect in the upper abdomen, no pleural effusion or pulmonary opacities (Fig. 1).

Echocardiography was unremarkable. Computerized tomography (CT) with contrast enhanced of the thorax showed a large solid anterior mediastinal mass with a diameter of 7.5/10.3 cm that abutted the great vessels and had posterior displaced and compressed the superior vena cava (Fig. 2).

The patient was afterwards referred to a thorax

Figure 1. Poster-anterior chest X-ray revealing large mediastinum due to a heterogeneous opacity in the middle third of the right hemithorax, closely to the heart but without lung infiltration, having no mass effect in the upper abdomen, no pleural effusion or pulmonary opacities.

surgery department. The protocol included surgical approach through full sternotomy. The tumoral mass was identified in the anterior and superior mediastinum, on intimate contact with the left mediastinal pleura and thymus edges. Total resection was achieved for both the tumor mass and thymus entirely, using a wide dissection between the two phrenic nerves, diaphragm and brachiocephalic vein. No intraoperation complication was experienced.

The tumor was macroscopically of ovular shape, measuring 15/12/5.5 cm, with smooth outer surface. The cut surface was tan with lobulated architecture, separated by fibrous septae, grey, with areas of hemorrhagic necrosis and cystic degeneration, and had an intact capsule of 2-10 mm. Pathology examination showed preserved thymic architecture, with granulomatous inflammation, lymphoid tissue hyperplasia, surrounded by necrotic lymphoid tissue. Immunohistochemically staining revealed: positive 'TdT' in normal tissue; negative 'ER'; positive 'L26/CD20' in normal thymus and in the rare remaining lymphocytes; negative 'CD30'; positive 'CD3' in the hyperplastic thymic areas, and in small lymphocytes; positive 'Ck19' in the epithelial cells of normal thymus, negative in the necrotic areas; positive 'AE1-AE3' in epithelial normal cells and negative in the necrotic zones (Fig. 3).

Based on these findings the tumor was classified as lymphocyte-predominant thymoma (type B1 thymoma), according to the World Health Organization (WHO) classification.

Post operatory, the patient developed right pleural effusion as shown in Figure 4, which required drainage for 5 days. The recovery was fully accomplished, the patient was discharged with no sequels from the surgery unit and he was referred to the oncology service, but he did not complete his follow-up visits, so we were not able to perform tests for myasthenia gravis.

DISCUSSION

There is a limitation of current literature regarding epidemiology of CP in children due to many reasons such as limited details, lack of follow-up procedures, small number for characterizing rare events and no evidence based guidelines. Many causes are benign; nonetheless, some serious events do exist sometimes and pediatricians must pay attention in order to identify and properly manage those cases. As some studies affirm, only about 0.3-0.6% of all

Figure 2. Pre-operative computerized tomography showing large solid anterior mediastinal mass with a diameter of 7.5/10.3 cm that abutted the great vessels and had posterior displaced and compressed the superior vena cava.

Figure 3. Histologic findings: poor small and medium epithelial cells proliferation (A), with vesicular nuclei, small nucleolus, surrounded by mature or immature lymphoid cells (B), medullary proliferation, Hessel corpuscles (C).

emergency department visits are counted by CP in children (10), having a median age for presentation of 12-13 years old (11, 12). As seen in our presented case,

several studies report that CP duration is less than 1 day (12, 13). The role of an emergency pediatrician is to rapidly assess any potential severe outcome

Figure 4. Post operative right pleural effusion, showing blunting of the costophrenic angle, blunting of the cardiophrenic angle, with mediastinal shift away from the effusion; pleural drainage for 5 days improved the evolution.

based on skilled history, clinical examination and proper investigation. Thus, the differential diagnosis is extensive and should be quickly performed by trained physicians in their attempt to identify specific etiology (Table 1).

As it is obvious in our presented case, the patients had no additional or worrying cardio-vascular and pulmonary symptoms or neurological deficits, so he could easily skip additional evaluation or referral.

On the other hand, mediastinum is a rare

location of space occupying processes in children. Also, there is a high rate of asymptomatic lesions in this area, considering that nearly half of them are incidentally discovered for various chest X ray examinations (15). Of all mediastinal lesions, thymomas represent almost 20- 30% of these masses in adults but only 1% in children. A high degree of susceptibility is risen when compressive symptoms appear like respiratory symptoms or superior vena cava syndrome, although this has a rare incidence. In

Table 1. Differential diagnosis of chest pain in children (13)

Cardiovascular	Respiratory
Arrhythmia	Asthma
Coronary artery disease (anomalous coronary arteries, acute Kawasaki disease [coronary arteritis], premature atherosclerosis [eg, dyslipidemia])	Pneumonia
Coronary artery vasospasm (toxicologic ingestion [cocaine, marijuana])	Pneumothorax/pneumomediastinum
Structural (hypertrophic cardiomyopathy, valvular stenosis [pulmonary, aortic], mitral valve prolapse)	Pulmonary embolism
Myocarditis	Pleuritis/pleural effusion (eg, systemic lupus erythematosus)
Pericarditis	Pleurodynia (coxsackievirus)
Endocarditis	Chronic cough
Congenital absence of pericardium	Airway foreign body
Aortic aneurysm or dissection (Marfan, Turner, Noonan syndromes)	
Abdominal and gastrointestinal	Musculoskeletal and chest wall
Esophagitis (gastroesophageal reflux disease, eosinophilic esophagitis, bulimia, pill esophagitis)	Chest wall strain (exercise, overuse injury, forceful coughing)
Esophageal foreign body	Skeletal (chest wall or thoracic spine) anomaly
Esophageal spasm/dysmotility	Trauma (contusion/rib fracture)
Gastritis	Costochondritis/Tietze syndrome
Hiatal hernia	Slipping rib
Referred pain from abdominal trauma (Kehr sign)	Precordial catch (Texidor twinge)
Cholecystitis	Breast tenderness
	Cutaneous (eg, herpes zoster)
Neurologic	Psychiatric
Migraine	Anxiety
Spinal nerve root compression	Panic
Slipping rib	Somatoform disorder (eg, conversion)
Precordial catch (Texidor twinge)	Depression, Emotional distress
Hematologic and oncologic	Chest wall, thoracic, or mediastinal tumor
Sickle cell disease	

Table 2. Masaoka Staging System of Thymomas and Corresponding Therapy (17)

Type	Histologic Description	Disease-Free Survival at 10 years,* %
A	Medullary thymoma	100
AB	Mixed thymoma	100
B1	Predominantly cortical thymoma	83
B2	Cortical thymoma	83
B3	Well-differentiated thymic carcinoma	35
C	Thymic carcinoma	28

*Series of 100 thymomas resected in Japan between 1973 and 2001 using WHO classification

adult population, a large number of entities associate thymomas, including myasthenia gravis and other autoimmune diseases, as pure red cell aplasia (16). However, this association is rarely described in childhood; even so, the patient should be investigated for myasthenia gravis on further evaluations.

Classically, a radiological enlarged mediastinum rises the susceptibility of a thymus pathologic mass. Sometimes, pleural effusion is seen in relation to bigger tumors. Anyway, precise diagnosis and anatomic reconstruction is possible only with a CT examination as the examination of choice in these situations (16). Thus, the suspected radiographic diagnosis was accurately sustained, allowing the

description of the thymic mass, the relationship with great vessels, dimensions and infiltration degree.

As other case report mentions, there are a limited number of cases and surgical experience with thymoma in pediatric patients, the diagnosis and treatment are very challenging for this age group (17). The prognosis is excellent in patients like ours, having an encapsulated tumor making possible total resection. Histologic findings enabled to classify the tumor according to Masaoka Staging System of Thymomas and Corresponding Therapy and World Health Organization (WHO) Pathologic Classification as seen in Tables 2 and 3 (18, 19). Future evolution might take into account that 3-27% of thymoma patients develop

Table 3. Masaoka Staging System of Thymomas and Corresponding Therapy (18)

Stage	Definition	Treatment I
I	Encapsulated tumor with no gross or microscopic invasion	Complete surgical excision
II	Macroscopic invasion into the mediastinal fat or pleura or microscopic invasion into the capsule	Complete surgical excision and postoperative radiotherapy to decrease the incidence of local recurrence
III	Invasion of the pericardium, great vessels, or lung	Complete surgical excision and postoperative radiotherapy to decrease the incidence of local recurrence
IV A	Pleural or pericardial metastatic spread	Surgical debulking, radiotherapy, and chemotherapy
IV B	Pleural or pericardial metastatic spread	Surgical debulking, radiotherapy, and chemotherapy

the second primary malignancies (20).

The peculiar interest of this case resembles in the surprising finding of thymoma in otherwise healthy adolescent patient that could easily be misdiagnosed.

In conclusion, chest pain is a frequent symptom seen in children and adolescents addressing to Emergency Departments. Although most cases of CP have a benign etiology in pediatric population, some of those resemble in serious organic lesions that need to be discovered. We present a rare etiology for CP revealing an encapsulated thymoma in a sport practicing adolescent boy. Due to lack of evidence based guidelines for evaluation, one should have in mind a large spectrum of rare and frequent causes of CP in children in order to proceed further investigation. The importance of radiology is once again fully considered not misdiagnosing these patients.

Conflict of interest

The authors declare that they have no conflict of interest concerning this article.

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