Case Report / *Olgu Sunumu*

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A Cause of Recurrent Pleural Effusion is Familial Mediterranean Fever

Tekrarlayan Plevral Sıvının Bir Nedeni Ailevi Akdeniz Ateşi

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Abstract

A 16-year-old woman was admitted to our hospital due to recurrent fever, chest pain, and dyspnea. She had an exudative pleural effusion and responded to antibiotic therapy with partial radiographic resolution. She had recurrence of her symptoms one weeks after and five weeks after the completion of therapy. Pleural biopsy by video-assisted thoracoscopy revealed chronic nonspecific pleuritis. The patient disclosed that mother offspring had FMF. Gene analysis showed she was heterozygous carrier of M680I (G/C) mutation. Treatment with colchicine led to resolution of her symptoms and of the pleural effusion. Turkey has a high prevalence of FMF. Pulmonologists should consider FMF in the differential diagnosis of patients with recurrent pleural effusions.

Keywords: Chest pain, pleural effusion, familial Mediterranian fever

Öz

On altı yaşında kız hasta tekrarlayan ateş, sol yan ağrısı, nefes darlığı yakınmalarıyla başvurdu. Arka-ön akciğer grafisinde solda plevral efüzyon ile uyumlu görünüm saptandı. Torasentez ile alınan plevral sıvı örnekleri eksüda özelliğinde olup hastaya antibiyoterapi uygulandı. Kısmi klinik ve radyolojik iyileşme ile taburcu edildi. Bir hafta sonra ve yine tedaviden beş hafta sonra aynı şikayetlerle tekrar kliniğimize müracaat etti. Hastaya Video Assisted Torakoskopi uygulandı. Biyopsi sonucu kronik nonspesifik plörit olarak geldi. Anamnez derinleştirildiğinde annede ailevi Akdeniz ateşi (AAA) olduğu ve hastanın M680I (G/C) heterojen taşıyıcısı olduğu öğrenildi. Romatoloji bölümüyle konsülte edilerek hastaya oral kolşisin başlandı. Bu tedaviden sonra hastanın kliniği düzeldi, plevral sıvısı geriledi. AAA tanısı olan hastalarının %95'inde ana yakınma abdominal ağrıdır. Abdominal ağrı olmaksızın plevral ağrı nadirdir. Plevral sıvı daha da nadirdir. Plevral efüzyonların etyolojisini araştırırken Akdeniz bölgesinde yer alan ülkemizde, AAA'nın da plevral sıvı nedeni olabileceğini aklımızda tutmamız gerekmektedir.

Anahtar Terimler: Göğüs ağrısı, plevral sıvı, ailevi Akdeniz ateşi

Introduction

Pleural effusion is one of the most common clinical problems characterized by fluid accumulation in the pleural space that the chest diseases and thoracic surgery departments encounter. The frequency of pleural effusion is about 280 thousands per year in Turkey (1). By the chest diseases department, thoracentesis is primarily applied to the patients to reach the differential diagnosis (2,3). The most common causes are heart failure, pneumonia and malignancies (4).

Familial Mediterranean fever (FMF) is a febrile illness that occurs in the form of attacks in the Mediterranean. Fever in the form of short-term attacks is accompanied by inflammation of the serous membranes such as peritoneum, synovial

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membrane, tunica vaginalis and pleura. The first finding usually occurs before the age of 20 and in the foreground as peritoneal symptoms (5,6). Application with pleural effusion as the first manifestation is a rare condition. The material obtained is generally exudate and shows polymorphonuclear cell dominance (6).

Here, we wanted to report our case to remind that AAA is among the causes of pleural effusion in Mediterranean countries, including our country.

Case Report

A 16-year-old female patient consulted to the emergency department with fever, left side pain and shortness of breath. She had recurrent fever and chest pain for the last 3-4 years. Complaints relieved in 4-5 days and abdominal pain did not accompany the attacks. The attacks were not associated with the menstrual cycle.

Physical examination body temperature; 38.9°C, a reduction in breath sounds and maturation were obtained from the lower zone of the left hemithorax. Other system examinations were normal. The left sinus was closed in the posterior anterior chest radiography. Thorax USG revealed a right 17 mm and left 34 mm effusion.

In laboratory examination, acute phase reactants; sedimentation rate was 75 mm/h, white sphere was 13.500/mm^3 and C-reactive protein was found to be 201 mg/L. Urinary findings were normal. Her electrocardiography was in normal sinus rhythm. D-dimer was negative. Left thoracentesis was performed, thorax tube was inserted and serous fluid was removed. The fluid was exudate and had polymorphonuclear cell dominance. PPD was 10 mm.

Patient; after pleuropneumonia was considered, other causes of pleural fluid were excluded, antibiotic treatment was started (Cefotaxime IV). She was discharged after partial clinical and radiological improvement but she was admitted to our clinic with severe pain on her left side at the end of one week and 5 weeks. Bronchoscopy was performed to the patient pursuant to the posterior anterior chest X-Ray and computed thorax tomography, complying with fluid on the left (Figure 1). Both endobronchial systems were normal in bronchoscopy. Lavage was taken from the left lower lobe; Lymphocytes, polymorphonuclear leukocytes, macrophages and bronchial epithelial cells were detected in the lavage fluid.

It was learned that her mother had a diagnosis of AAA when the patient's history was deepened. The gene mutation of the patient was investigated and it was learned that she was a M608I (G/C) heterogeneous carrier. Following this, the patient was consulted to the rheumatology department and antibiotherapy was completed and the oral colchicine

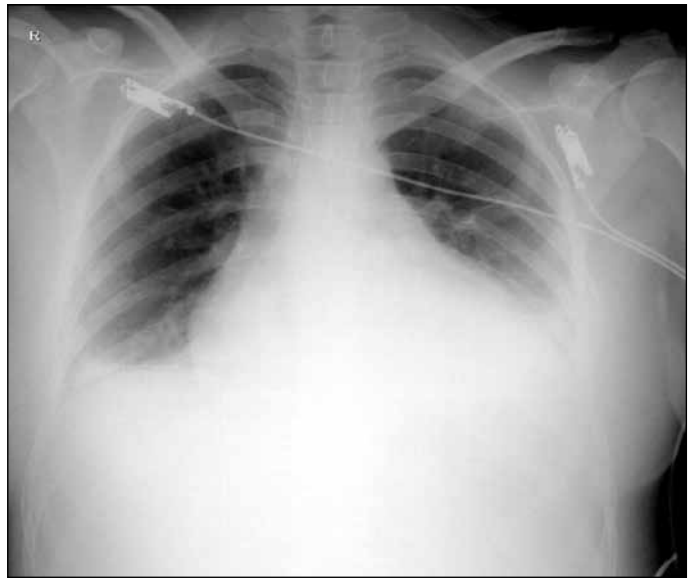

Figure 1. Opacity in the chest radiography, consistent with pleural fluid on the left extending to the middle zone.

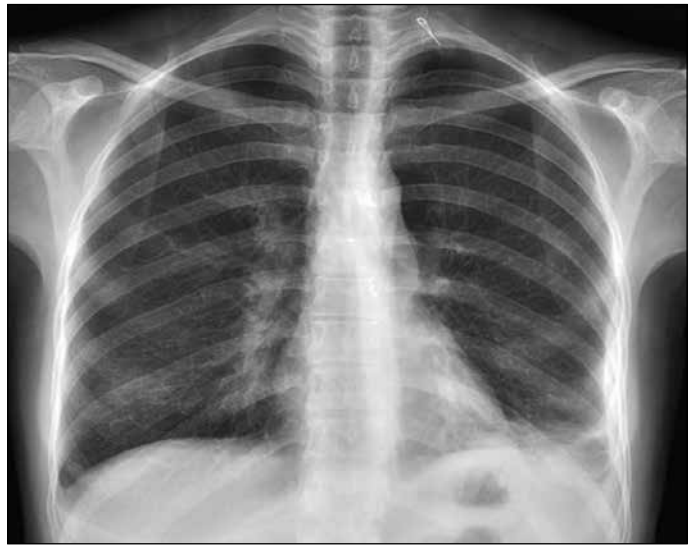

Figure 2. Fluid on the left side in the chest radiograph was significantly reduced after colchicine treatment.

was started (0.5 mg 3 x 1). The patient responded to colchicine treatment. The patient was discharged providing consulting for ambulatory control later on. Outpatient radiographs showed a complete disappearance of fluid after discharge (Figure 2).

Discussion

Pleural effusions which sometimes have to be diagnosed by chest diseases and thoracic surgery together constitute one of the most common clinical problems. About 280 thousands of patients with pleural effusion are seen per year in Turkey, and diagnostic tests are applied (1). Although most pa-

tients with pleural fluid have to undergo thoracentesis, if there is fluid less than 10 mm in the lateral decubitus radiography, or if the patient with known heart failure has bilateral fluid, there is no hastiness for thoracentesis and response to treatment can be expected. In light of light criteria, pleural fluid taken by thoracentesis is classically classified as exudate and transudate. Transudate fluids are often thought to be caused by a systematic failure in the production and distribution of pleural fluid. Exudate fluids are thought to be caused by disorders in the pleural surface and capillary vessels. The most important reason for this distinction is to be able to select the transudate fluids which do not necessiate further examination. Examples of such diseases are heart failure, carcinoma failure and cirrhosis (2,3). If we consider the causes of pleural effusions in general, fluids based on heart failure, pneumonia and malignancy are the most common (4). It should be noted that in Turkey, the tuberculosis is often among these diseases. In this case, the etiology of exudate fluid is attempted to be clarified by the chest diseases. The appearance, cytology, cell counting, microbiological and biochemical studies of the fluid are performed. The diagnostic algorithm also includes pleural biopsy and VATS / thoracoscopy.

AAA is a febrile disease that occurs mostly in the Mediterranean race. Fever in the form of short-term attacks is accompanied by inflammation of the serous membranes such as peritoneum, synovial membrane, tunica vaginalis and pleura. The first finding usually occurs before the age of 20 and in the foreground as peritoneal symptoms (5,6). Fever is the indispensable criterion of diagnosis. Generally it is over 38 and it takes 1-3 days to start and disappear. In 90% of patients with AAA, the main complaint accompanying the fever is abdominal pain. Pain occurs in 6-12 hours and its severity begins to decrease in 24-48 hours. Perito-neal irritation findings and acute phase reactants increase. Sometimes diarrhea accompanies this scene. It may also occur with the findings of arthralgia, arthritis, myalgia and pericarditis. Scrotal attack is also rarely seen (7). Application with pleurisy as a first manifestation is a rare condition. It is more rare to find pleural findings such as chest pain and fluid development without abdominal pain (8-11). The material obtained is generally exudate and has a polymorphonuclear cell dominance. Rarely eosinophil dominance can be also observed (6). With the appropriate treatment regimen, the liquid completely disappears. Amyloidosis developing in the later stages of the disease determines the prognosis of the disease. Since this condition can lead to renal failure, treatment with colchicine, sulfasalazine and sometimes methotrexate should be initiated regardless of the manifestation of the disease (7,12).

The autosomal recessive disease is frequently associated with MEFV gene mutations, and it is known that M694V, M680I, V726A, E148Q mutations are most commonly found in the Turkish population. It is also known that symptoms may occur in both homozygous and heterozygote cases (7,9,13). In our case, M680I mutation was heterozygous. Early diagnosis and treatment in patients with AAA prevents the development of renal amyloidosis. AAA should be considered among differential diagnosis in patients with chest pain and fever-inducing pleural effusions, especially in the absence of response to routine nonspecific treatment and in areas where the disease is common. The diagnosis should be confirmed with both the gene mutation analysis and the response to drugs such as colchicine.

Conclusion

Pleural effusion is one of the most common clinical problems characterized by fluid accumulation in the pleural space that the chest diseases and thoracic surgery departments encounter. AAA should be considered among differential diagnosis in patients with chest pain and fever-inducing pleural effusions, especially in the absence of response to routine nonspecific treatment in Turkey, a Mediterranean country. Early diagnosis and colchicine treatment may prevent further renal failure.

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