A CASE REPORT OF FMF WITH ISOLATED PLEURITIS

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SUMMARY: Familial Mediterranean Fever (FMF) is characterised by recurrent fever episodes and polyserositis. Pleura is a very rare site of the serosal membrane inflammation. We report a case of FMF with isolated pleuritis and fever.

Key Words: Periodic Disease, Pleurisy.

INTRODUCTION

Familial Mediterranean Fever (FMF) is a chronic genetic autosomal recessive disease, which is characterized by recurrent polyserositis confined to certain ethnic groups (1-4). Clinical findings of the disease are diverse and peritoneal involvement dominates (95%). Pleural attacks are seen in 40%, and synovial inflammation in 78% of the patients with FMF (2). The incidence of serosal inflammation varies in the literature. An isolated involvement of the pleura is unusual, and usually seen together with other serous membrane inflammation (usually peritoneum) (2, 5). The disease is characterized by recurrent fever episodes, and not always associated with peritonitis, pleuritis and/or synovitis. Exudative or hemorrhagic pericarditis, erisipelaelike lesions of the skin, orchitis, and myalgia may also be observed (2).

In this case report, we describe the clinical findings of FMF which was initially recognized by peritonitis and fever, but thereafter continued by only pleuritis and fever without any other serosal inflammation.

CASE REPORT

A 21-year-old woman was diagnosed as FMF with the signs and symptoms of recurrent peritonitis and fever in 1993. She was taking 1g colchicine per day for three years, but she had stopped colchicine by herself twelve months ago. Three months after the drug was stopped, she had a chest pain located at the right hemithorax. The pain was associated with fever and dyspnea. The symptoms continued for three days and resolved spontaneously. Then, the same clinical symptoms repeated for two times during nine months. She was given clarithromycin at the last attack. The clinical picture was the same as described above, which consisted of right chest pain, high grade fever and malaise. Physical examination revealed decreased breath sounds at the base of right hemithorax and dullness of the right costophrenic sinus.

Laboratory findings were as follows: Hemoglobin 12.9g/dl, white blood cell count 22300/L, platelet count 325000/L, ESR 54 mm/h, fibrinogen level 258 mg/dl (normal:200-400 mg/dl), CRP 16 mg/dl (normal: 6 mg/dl). Blood
chemistry and urinalysis were normal. In 24h urine collection analysis, creatinine clearance was 88.2 ml/min and, proteinuria was 16.8 mg/d. Anti-nuclear antibody and anti-ds-DNA were negative. Chest X-ray revealed mild pleural effusion in the right costophrenic sinus (Fig. 1). Lung ventilation-perfusion scan revealed nonspecific findings.

After these laboratory investigations, the patient was started 3 g colchicine per day. After two days of therapy, clinical signs and symptoms regressed. On the fourth day of therapy, the pleural effusion on the chest X-ray disappeared (Fig. 2).

**DISCUSSION**

FMF is a recurrent disease associated with fever, peritonitis, pleuritis, and/or synovitis. It is prevalent in certain ethnic groups, like non-Askhenazi Jews, Arabian, Turkish, or Armenian descent. Recently, a gene is described that encodes susceptibility for FMF. This gene is called “MEF”. In Israel, short arm of the 16th chromosome is defined to carry this gene in non-Askhenazic Jewish families. Afterwards in Armenian population, this gene was also isolated (4). Many studies supporting the genetic inheritance of FMF have been reported. Shohat and colleagues have shown that monozygotic twins with FMF showed the same clinical progress. In dizygotic twins, only one of the twins usually displays the clinical features of FMF. Also in monozygotic twins, affected organ systems are the same, but in dizygotic twins affected localizations are different (5). In this twin study, pleuritis is determined together with fever, peritonitis and synovitis. Only one of the monozygotic twins showed isolated pleural involvement and fever.

In our case, the patient initially had peritonitis and fever, but afterwards, the clinical picture changed and she had pleuritis and fever. Pleural involvement in FMF is very frequently associated with other serous membrane inflammation. The initial attack is characterized by pleuritic chest pain and fever in fewer than 10% of patients, but approximately 40% have an attack of febrile pleurisy during the course of their disease (6).

Isolated pleural inflammation and fever in FMF may lead physicians to search for infectious factors and treatment with antibiotics or to investigate non-infectious factors. In this case, pleural attack of FMF was misdiagnosed and an empirical antibiotherapy was given. Colchicine has been used for FMF attacks since 1972, and 90% of attacks are being prevented (7-9). Before the advent of colchicine in the treatment of FMF, a great number of FMF patients had progressed to secondary amyloidosis and end-stage renal failure. Colchicine is still being used for both the prevention and attenuation of acute attacks. In our case, both clinical symptoms and radiologic signs disappeared after 4 days of colchicine therapy.

It is noted that FMF is a relative frequent disease in the Turkish population, and isolated pleural attacks should be kept in mind in the differential
diagnosis of selected patients. Since we were aware of the patient’s disease, we were able to predict that the patient was having a pleural attack. As a result; fever and pleural effusion with pain might be the first manifestations of FMF in patients with no history of the disease.

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