

Acute Recurrent Pericarditis as the Inaugural Manifestation of Familial Mediterranean Fever

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ABSTRACT

Familial Mediterranean fever (FMF) is an inherited autosomal recessive disorder commonly found among individuals of Mediterranean or Middle Eastern descent and caused by Mediterranean Fever gene (MEFV) mutations on chromosome 16. It is the most frequent periodic febrile syndrome among the autoinflammatory syndromes. Typical febrile attacks include serosal inflammation, but although FMF is characterized by recurrent polyserositis, there is a little documentation of pericardial disease among patients.

We report the case of a patient who presented recurrent pericarditis. Each episode occurred with a high fever and elevation of the C-reactive protein level. Treatment with colchicine was effective. The patient was found to be heterozygous for the V726A mutation.

LEARNING POINTS

- The aetiology of recurrent pericarditis is still largely unknown.
- The occurrence of pericarditis as a manifestation of familial Mediterranean fever is controversial.

KEYWORDS

Pericarditis, familial Mediterranean fever

CASE DESCRIPTION

A 26-year-old Turkish man was admitted because of recurrent chest pain. He was a 10-pack-year smoker but denied any alcohol use. His medical history before 2016 was unremarkable.

In September 2016, he was admitted to a cardiology unit due to chest pain and a high fever of 38.5°C. Laboratory results revealed marked elevation of C-reactive protein (CRP) levels (70 mg/dl). Echocardiography disclosed minimal pericardial effusion. He was diagnosed with acute pericarditis. He was given aspirin and colchicine. One month after discharge, treatment was discontinued by patient himself without giving any reason.

In May 2018, he was admitted to our unit for chest pain and fever of 38.5°C. On general examination, he seemed to be acutely ill. He complained of severe chest pain which had begun 24 hours prior to admission. The temperature was 38.5°C, his pulse rate was 100/min and his blood pressure was 109/65. He did not exhibit abdominal pain, myalgia, lower limb oedema or skin eruption. Once again, CRP was increased and pericardial effusion was minimal. ECG was abnormal, documenting ST depressions. No pleural effusion was detected on CT of the chest. Infectious diseases, namely tuberculosis, malignancy, and viral and autoimmune diseases were excluded by clinical and laboratory analysis.

The repeated episodes of pericarditis together with marked elevation of the CRP level led us to suspect the presence of autoinflammatory disease. However, no family history of autoinflammatory disease (familial Mediterranean fever, tumour necrosis factor receptor-associated periodic syndrome, hyper-IgD syndrome) could be found. Nevertheless, the MEFV gene was assessed in order to confirm the diagnosis and the patient was found to be heterozygous for the V726A mutation. Although this mutation is rare in patients with familial Mediterranean fever (FMF) and its penetrance is low, it is considered to be a disease-causing mutation.

A differential diagnosis was performed in order to rule out the possibility of other autoinflammatory diseases. Hyper-IgD syndrome (HIDS) is characterized by periodic fevers, cervical lymphadenopathy, erythematous macules, abdominal pain, vomiting and arthralgia with persistent inflammation. Tumour necrosis factor receptor-associated periodic syndrome (TRAPS) is characterized by periodic fevers, conjunctivitis, erythematous skin lesions, myalgia, arthralgia and abdominal pain, with fever being long term. Based on the patient's features, the diagnoses of HIDS and TRAPS were ruled out clinically. Behcet's disease was also ruled out based on the absence of oral ulcers, genital ulcers, erythema nodosum and uveitis.

As in the first episode, treatment with colchicine and aspirin was rapidly effective.

At discharge, the patient was prescribed long-term colchicine. Two years later, no disease recurrence has been detected.

DISCUSSION

Familial Mediterranean fever is an autosomal recessive disease, most common among Sephardi Jews, Arabs, Armenians, Turks and other peoples of the Mediterranean basin. FMF is characterized by recurrent episodes of febrile serositis, manifesting mainly as abdominal, chest and joint pain. The attacks may also involve the skin, testes and muscles, or present as fever alone.

Recurrent pericarditis is a state of repetitive inflammation of the pericardium with intervals of remission. Recurrence of pericarditis was defined on the basis of 'pericardial' pain and one or more of the following signs: fever, pericardial friction rub, ECG changes, echocardiographic evidence of pericardial effusion, and an elevated white blood cell count, CRP level or ESR^[1].

The aetiology of recurrent pericarditis is still largely unknown, but most causes are presumed to be immune mediated. Several diseases are frequently associated with such manifestations and include systemic lupus erythematosus, FMF and TRAPS.

The occurrence of pericarditis as a manifestation of FMF is controversial. Although Sohar et al. found no cases of pericarditis in their report on 470 FMF patients^[2], many studies have found that the incidence of pericarditis in FMF patients is higher than in the general population. In a retrospective study by Kees et al. that included 4000 FMF patients over a 20-year period, the prevalence of pericarditis was reported to be 0.7%^[3]. The Turkish FMF Study Group reported that the incidence of pericarditis was 1.4% in a cohort study^[4]. In contrast, Dabestani reported a much higher prevalence (27%) of pericarditis in patients with FMF^[5]. Pericardial involvement in FMF usually occurs late in the course of the disease.

In our case, acute recurrent pericarditis was the sole manifestation of FMF. Similar situations are rarely reported^[6-8].

However, idiopathic recurrent pericarditis remains the most frequently observed clinical condition and the aetiology of this disease still needs to be elucidated^[1,2].

Since genetic diagnosis for FMF is available, we suggest that mutation analysis for FMF should be considered in patients with idiopathic recurrent pericarditis, especially those of Mediterranean origin.

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