Pericardial involvement in familial Mediterranean fever

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Summary: Two patients with familial Mediterranean fever showed the classic features of pericardial involvement and one of them (Case 2) had pericardial effusion detected by echocardiography. These and previously published cases show that familial Mediterranean fever should be considered as a cause of pericarditis and/or pericardial effusion.

Introduction

Familial Mediterranean fever (FMF) is an inherited disease of unknown aetiology. Its characteristics are recurrent, short episodes of fever, peritonitis, pleuritis or synovitis. Systemic amyloidosis with subsequent renal failure and death develops in some patients. FMF is usually seen in Jews, Armenians, Arabs and Turks. Although it primarily affects serous membranes, there have been only a few case reports and an echocardiographic study on pericardial disease in nonuraemic patients with this disease.1–8 We report two patients with FMF who had pericarditis during their attacks.

Case reports

Case 1

A 26 year old man was admitted because of recurrent chest pain frequently associated with abdominal pain for the last 3 months. His past history revealed episodes of abdominal pain and operations on three occasions with diagnosis of appendicitis and intestinal obstruction. On physical examination there was only a pericardial friction rub heard at the left lower sternal border. Laboratory findings showed normal values except for an elevated sedimentation rate (38 mm/h). The electrocardiogram and chest X-ray were normal, but the echocardiogram and chest X-ray taken one month before the admission in a private surgery demonstrated typical ST-T changes and mild cardiomegaly consequently. Abdominal ultrasonography revealed mild hepatosplenomegaly and enlargement of both kidneys. M-mode and two-dimensional echocardiography showed normal findings. The pericardial friction rub disappeared 2 days later. He came back to the hospital 2 months later with another attack of chest and abdominal pain. A pericardial friction rub was heard again. Laboratory examination showed elevated sedimentation rate (46 mm/h), white cell count was \(10.7 \times 10^9/l\), urinalysis and renal function tests were normal. Antinuclear antibodies, anti-DNA and LE cells were negative. Electrocardiogram and echocardiogram were normal. He was put on colchicine therapy (2 tablets daily) and has had no other attacks in 2 years' follow-up.

Case 2

A 32 year old woman was referred to the Cardiac Research Center because of chest pain and dyspnoea. She gave a 27-year history of episodes of abdominal pain diagnosed as FMF. She was on colchicine therapy, but she had not taken the tablets lately. On physical examination she looked very ill and had a temperature of 37.5°C. There was a left pleural friction rub. No pericardial friction rub was heard. Chest X-ray showed a blunted left costo-phrenic angle, a left lower infiltration and a slightly enlarged heart. Two days later a pericardial friction rub was present. Electrocardiogram showed typical ST-T changes indicating pericarditis. M-mode and two-dimensional echocardiography demonstrated a small pericardial effusion. Laboratory examination revealed an elevated sedimentation rate (127 mm/h), white cell count \(15.2 \times 10^9/l\). All the other routine findings, including urinalysis and renal function tests, were normal. Antinuclear antibodies, anti-DNA and LE

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cells were negative. She improved in two days with bed rest only. The pericardial friction rub disappeared, the pericardial effusion was no longer present and the chest X-ray was normal. She had another attack 15 days later. A pericardial friction rub was again present and echocardiography showed a pericardial effusion. She was given colchicine and she has remained free of further attacks of chest and abdominal pain in 12 months' follow-up.

Discussion

Familial Mediterranean fever is a disease characterized by recurrent polyserositis, but there are only a few reports of pericarditis in this disease. All these cases were reviewed by Dabestani et al. and only two of them were considered as having the real features of FMF. In all these cases only electrocardiograms and clinical findings were used to diagnose pericarditis. The study by Dabestani et al. is the only one in which echocardiography was used to detect pericardial involvement. According to this study, 2 patients out of 4 with chest pain and 6 patients out of 22 without a history of chest pain showed pericardial disease at the time of the study. It was concluded that pericardial involvement increased with the duration of the illness.

Although we could not exclude the possibility that our patients could have had an intercurrent viral pericarditis, the differential diagnosis of our patients was based on their past histories, the lack of prodromal symptoms, clinical and laboratory examinations, the natural courses of the attacks and the good results obtained with colchicine therapy. They showed pericarditis at the time of their attacks and both of them had pericardial friction rub and electrocardiographic changes typical of acute pericarditis. Chest X-ray series suggested the presence of pericardial effusion one month before admission in Case 1, but there was no effusion on admission. In his second attack, clinical and electrocardiographic findings were typical for pericarditis, but again pericardial effusion was not detected. In Case 2, we demonstrated pericardial effusion by echo during both her attacks.

In conclusion, physicians should bear in mind that FMF is a cause of pericarditis and pericardial effusion.

References

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