Recurrent Pericarditis as a Manifestation of Familial Mediterranean Fever

To the Editor:

We read with great interest the article by Adler et al regarding colchicine treatment for recurrent pericarditis.\(^1\) The authors described 51 patients from Israel, Spain, and the United States. The pericarditis was idiopathic in 33 patients and secondary in 18. Recurrences of pericarditis did not respond to conventional therapy consisting of nonsteroidal anti-inflammatory drugs, corticosteroids, pericardiocentesis, or some combination thereof and could only be controlled after the initiation of colchicine treatment. The authors concluded that colchicine was effective and safe for the long-term prevention of recurrent pericarditis, especially in idiopathic cases.

Colchicine therapy has been shown to be an effective method for eliminating attacks of familial Mediterranean fever (FMF), which is an autosomal recessive disorder characterized by recurrent and self-limited attacks of fever accompanied by inflammation of peritoneum, synovium, and pleura.\(^2\) Pericardial involvement is a rare but well-known feature of the disease.\(^2\) Tauber et al\(^1\) reported 2 patients who had recurrent pericarditis as an initial sole manifestation of FMF. Although pericardial inflammation has been considered rare, an incidence of pericardial involvement of 27% has been reported when echocardiography is used for the diagnosis.\(^4\) We also have 2 patients from Turkey (8- and 11-year-old girls) who had recurrent pericarditis as an initial feature of FMF. They had no further pericardial attacks during 6 and 56 months of follow-up after initiation of regular colchicine therapy (unpublished data).

FMF is a disease that primarily affects Jews, Turks, Armenians, and Arabs.\(^2\) Although Adler et al\(^1\) did not mention their patients’ ethnic backgrounds, Israel and Spain are countries in which North African Jews make up a considerable part of the population. It is also noteworthy that a substantial number (35.8%) of their patients experienced relapses after discontinuation of colchicine. These findings suggest that at least some of the patients with idiopathic pericarditis in this report might have FMF. The diagnosis of FMF was based on clinical findings, ethnicity, and response to colchicine. In atypical cases, however, FMF is difficult to diagnose.\(^2\) Cloning of the recently identified FMF gene (MEFV) allows a new and reliable diagnostic test for FMF. The 4 missense mutations were reported to be present in 85% of FMF carrier chromosomes.\(^5\) Because patients with recurrent pericarditis are admitted mainly to cardiology clinics, it is quite possible to miss the diagnosis of FMF as a cause of recurrent pericarditis. Thus, cardiologists, especially those working in the countries where FMF is prevalent, should be aware that recurrent pericarditis might be a part of the clinical spectrum of this disease. Since the genetic diagnosis of FMF is now possible, we suggest that mutation analysis for FMF should be considered in patients with idiopathic recurrent pericarditis, especially those of Mediterranean origin. It might provide useful data concerning not only the incidence of recurrent pericarditis in FMF but also the incidence of FMF as a cause of idiopathic recurrent pericarditis.

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Response

We are delighted with the interest of Tutar et al in our study on colchicine treatment for recurrent pericarditis.\(^1\) Indeed, the most troublesome complication of acute pericarditis is recurrent episodes of pericardial inflammation, which occur in 15% to 32% of cases. The cause of the disease is usually unknown.\(^3\) We reported the results of the largest international multicenter study on 51 patients with recurrent pericarditis who did not respond to conventional therapy and were successfully treated with colchicine to prevent further relapses. The duration of follow-up was 10 years.\(^1,3\)

Tutar et al remind us that pericardial involvement is a rare but well-known feature of familial Mediterranean fever (FMF), but that its incidence is up to 27% according to echocardiographic studies. Thus, they raise the possibility that some of our patients with idiopathic recurrent pericarditis suffer from FMF, since most of them are of Mediterranean origin. It is indeed well-known that FMF patients may present with episodes of pericarditis. However, affected patients always present with other manifestations of FMF within a short period of time (usually in a few months).\(^4\) In our study,\(^1,3\) the mean follow-up of patients with recurrent pericarditis with unknown etiology was 42.3±35.2 months (range, 6 to 134 months). None of them demonstrated any other possible manifestation of FMF during this period. Moreover, one of our patients, a 12-year-old boy, suffered 6 recurrent episodes of idiopathic pericarditis within 3 years, all of them with an excellent response to colchicine treatment after a failure of conventional anti-inflammatory therapy. Although this child did not demonstrate any other manifestation of FMF, cloning for the known FMF gene (MEFV) mutation was carried out, with negative results.\(^5\)

In conclusion, we generally accept that genetic studies of FMF should be carried out in patients with recurrent episodes of pericarditis of unknown etiology, especially patients of Mediterranean origin.
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