

Familial Mediterranean fever in Iranian children. First report from Iran

Sirs,

Familial Mediterranean fever (FMF) is the most common form of the hereditary periodic fever syndromes, which has autosomal recessive pattern and presents with self-limited periodic fever and serositis in its classic form (1). It usually occurred in populations from Mediterranean descent (Armenian, Arab, Jewish, Greek, Turkish and Italian populations) (2). Recently it has been reported from northwest of Iran with high frequency (3). On the other hand, on the basis of a recent genotype study in Iranian FMF patients (4, 5), we expect some difference in clinical presentation and course of the disease in this region and patients. The most frequent mutation are M694V (28%), followed by V726A (9%), E148Q (7%), M680I (7%), and M694I (1%) mutations in one study (5), and in other study of the 30 cases with full criteria, 9(30%) were positive for M694V mutation (2 homozygote, 6 simple heterozygote, and one compound heterozygote), 4 (13.3%) for M680I mutation (one homozygote, 2 simple heterozygote, and one compound heterozygote) and one (3.3%) for V726I mutation. All of the controls were negative for the three mutations (4). These results indicate some differences in the frequency of individual mutations (6). The high frequency of E148Q in Azeri Turks compared with Mediterranean ethnic groups is rather interesting (7). Forty-eight patients aged between 2 and 18 years old who fulfilled Tel-Hashomer criteria were included in this study.

Abdominal pain was the main symptoms in these patients, but the most common symptoms were systemic one, like fever (98%), although it was only in 12.5% main symptom, and then followed by GI (89.6%) symptoms. In more than 88.8% of patients severity of pain was more than 8. In most patients the duration of each crisis was 1-3 days. Most of the patients had hospital admission for diagnostic work up (43%) and some of them (22.9%) had erroneously had previous surgical operations. Five patients had positive family history of FMF (4 in paternal and one in maternal family). The parents of patients were first-degree relatives in 29.2% and in 54.2% delay in diagnosis was more than 3 years. All took colchicine (0.5 - 1.5mg) as a first choice of therapy, and more than 87.5% had good response to colchicine, 19% of patients were FTT, although most of them had nutritional problems. Early recognition of FMF is optimal. We have more

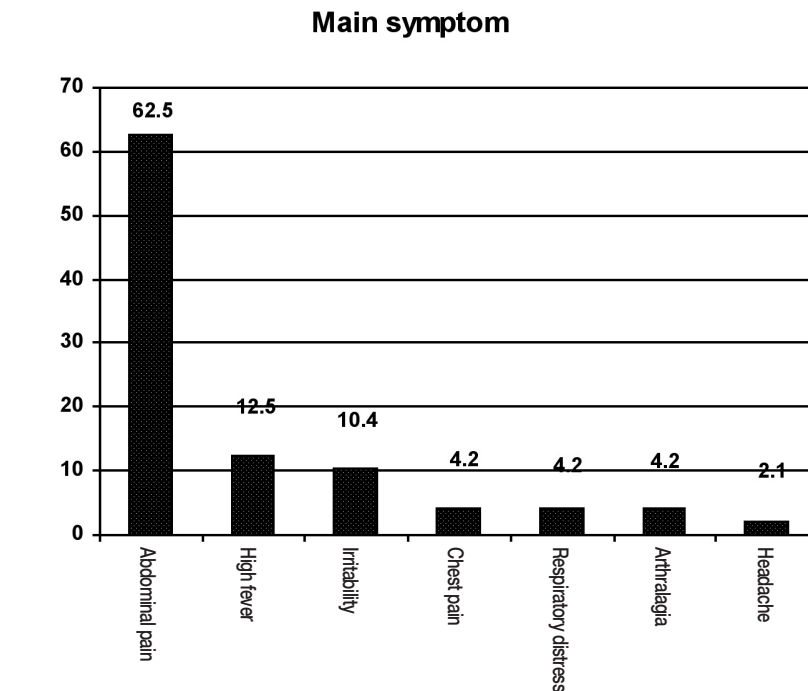


Fig. 1. Main symptoms in patients.

than 3 years delayed in diagnosis in most patients. This delay could be due to the low index of suspicion for the diagnosis. 54% in this study were females with F/M ratio of 1.17:1. Recurrent generalised abdominal pain was the most common feature and the incidence and duration of pain was similar to that reported by others. (8) In more than 20% because of its severity, patients underwent surgical operation; it seems this rate is high; undoubtedly low index of suspicious could be the causes. Chest pain (20%) was followed by abdominal pain in our patients (Fig. 1.) The frequency of arthritis is variable among different ethnic groups: in our group it was 4.2%, while it has been reported higher in Jews, followed by Armenians, and less commonly in Turks (9). The pattern of arthritis was monoarthritis of large joints as the most common presentation. Classic skin feature, erysipeloid erythema, is described in 7 to 40% of patients (8), but it was not seen in our patients. One patient had renal disease complicating FMF with nephritic findings; he was not on colchicine prophylaxis.

Because this is the first report from Iranian children, it provides a clinical profile of FMF in this area and allows comparison with other studies.

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