Familial Mediterranean fever patients may have unmet needs for treatment of erysipelas-like erythema

Sirs,

Familial Mediterranean fever (FMF) is an autosomal recessive disease with recurrent polyserositis (1). Erysipelas-like erythema (ELE) is the pathognomonic skin manifestation of FMF which may be triggered by physical effort and subside spontaneously within 48 to 72 hours of bed rest (2). Even if colchicine might decrease the mean frequency of all kinds of the cutaneous lesions including ELE (3), as far as I concern, there is no study that evaluated treatment efficacy in ELE.

While I was examining FMF patients in my Rheumatology Outpatient Clinic, I realised that several FMF patients were suffering from ELE, even though their polyserositis symptoms were in remission. Therefore I have applied a short questionnaire to the patients with ELE who were admitted within the last six months. This questionnaire includes age and gender of the patients; disease duration; main FMF symptoms; colchicine's efficacy in the main FMF symptoms and ELE which was rated by each patient within a 0–100 point scale (0 equals to no response, 100 equals to complete response), maximum colchicine dosage; co-occurrence of the main FMF symptom with ELE and precipitating causes of ELE. Furthermore, treatment efficacy scores divided into three categories (<50 inadequate response, 50 to 75 moderate response, >75 adequate response). This questionnaire was approved by the Local Research Ethics Committee and carried out in compliance with the Helsinki Declaration. All the patients gave written informed consent. The results are presented as mean±standard deviation.

Table I. Treatment responses of 49 FMF patients with ELE.

Treatment Response	Main FMF symptoms[a] n (%)	ELE[b] symptom n (%)
Inadequate response[c]	4 (8.2)	24 (49.0)
Moderate response[d]	29 (59.2)	18 (36.7)
Adequate response[e]	16 (32.7)	7 (14.3)

[a]Main FMF symptoms: Abdominal pain, fever and fever/abdominal pain together;
[b]ELE: Erysipelas-like erythema;
[c]Inadequate response: Treatment efficacy score <50;
[d]Moderate response : Treatment efficacy score 50 to 75;
[e]Adequate response: Treatment efficacy score >75.

In the duration of six months, I have examined 228 FMF patients who met the Tel-Hashomer Criteria (4). Only 49 (21.4%) of them had at least one attack of ELE in their life. This frequency was compatible with the disease characteristics of Turkish FMF patients (5). Furthermore, 36 (73.5%) of the patients were woman. Mean age of the patients was 33.89±10.52 years and mean disease duration was 7.81±5.96 years. All patients have been taking colchicine and two of them were on anakinra in addition to colchicine. Mean maximum colchicine dosage was 1.57±0.48 gr. Moreover, 34 (69.4%) of the patients' main symptom was abdominal pain. More than two out of three patients noticed at least one precipitating cause of ELE including staying standing up for a long time, long distance journey and excessive physical activity whereas only two (4.1%) of the patients reported co-occurrence of their ELE attacks with serositis. While with FMF treatment, only four (8.2%) of the patients had inadequate response for serositis, the number of patients having inadequate response to ELE was 24 (49.0%) (Table I). Furthermore, for both of the patients who were on anakinra and colchicine together, the treatment efficacy in ELE got better from inadequate to adequate response.

Even though, the data were obtained from a simple questionnaire with limited number of patients, it may show that even if colchicine successfully treats polyserositis symptoms, there might still be an unmet need for the treatment of ELE. Additional studies that evaluate treatment options for ELE may be essential to meet the requirements of the patients.

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References

1. BEN-CHETRIT E, LEVY M: Familial Mediterranean fever. *Lancet* 1988; 351: 659-64.
2. BARZILAI A, LANGEVITZ P, GOLDBERG I *et al*.: Erysipelas-like erythema of familial Mediterranean fever: clinicopathologic correlation. *J Am Acad Dermatol* 2000; 42: 791-5.
3. MAJEED HA, QUABAZARD Z, HIJAZI Z, FARWANA S, HARSHANI F: The cutaneous manifestations in children with familial Mediterranean fever (recurrent hereditary polyserositis). A six-year study. *Q J Med* 1990; 75: 607-16.
4. LIVNEH A, LANGEVITZ P, ZEMER D *et al*.: (1997) Criteria for the diagnosis of familial Mediterranean fever. *Arthritis Rheum* 1997; 40: 1879-85.
5. TUNCA M, AKAR S, ONEN F *et al*.: Familial Mediterranean fever (FMF) in Turkey: results of a nationwide multicenter study. *Medicine* 2005; 84: 1-11.