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IL-18 serum concentration is continuously elevated in typical familial Mediterranean fever with M694I mutation and can distinguish atypical type

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Objectives

Familial Mediterranean fever (FMF) can be classified into typical and incomplete/atypical types based on clinical findings and gene analysis, although biomarkers that distinguish typical from atypical FMF have not been unclear.

Methods

We here investigated the serum cytokine profiles of IL-1β, IL-6, IL-8, TNF-α, IFN-γ, and IL-18 in FMF compared with those in Kawasaki disease.

Results

IL-1β, IL-6, IL-8, TNF-α, and IFN-γ were not increased in either type of FMF in the remission state and in controls, and IL-6 was elevated during attack periods among patients. Serum IL-18 levels were significantly higher in typical FMF patients with M694I *MEFV* mutation in remission than in controls at the same level as flared Kawasaki disease, which further increased during attack periods. In contrast, IL-18 levels in atypical FMF with P369S-R408Q mutation or in typical FMF without M694I mutation was not increased, in either disease states.

Conclusion

Thus, serum IL-18 levels at attack increase more than in remission, and that are an excellent marker to distinguish between the two types of FMF.

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