Abstract

Introduction: Periodic fever syndromes are clinical entities classified as autoinflammatory diseases. The most of the periodic fever syndromes have genetic predisposition (monogenic periodic fever syndromes). PFAPA (*Periodic Fever, Aphtous stomatitis, Pharyngitis a Adenitis*) syndrome is an idiopathic disease with unknown aetiology.

Results: In our study, we described the largest clinical series of patients with PFAPA syndrome from a single center. The laboratory results have confirmed uncomplicated course of PFAPA syndrome. In our measurements we observed significantly higher levels of serum cytokines (IL-1β and IFN-γ) during episodes of fever in PFAPA patients compared to the control group. Our measurements showed increased numbers of plasma cells in the peripheral blood of PFAPA patients. We have found increased levels of naïve CD4 and CD8 T cells and approximately 2-fold higher proportion of CD8 T cells in tonsils of PFAPA patients. Significant differences were also present at levels of IFN-γ, IL-1β, IL-6 and TNF-α in stimulated supernatants compared to supernatants from unstimulated peripheral blood from patients with PFAPA syndrome. Measurements of bacterial profile showed individual microbial profile in patients.

Conclusion: Removal of the tonsillar tissue with the potential bacterial/viral/other trigger leads to disappearance of recurrent episodes of fever. Patient with PFAPA syndrome is probably immunologically immature individual susceptible to external factors.

Keywords: periodic fever, innate immunity, inflammation