

PFAPA syndrome study at Imam Khomeini Hospital Pediatrics Rheumatology Clinic in Ardabil from 2003 to 2007.

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Abstract

Background: PFAPA (periodic fever, aphthous stomatitis, pharyngitis, adenitis) syndrome is a new syndrome of periodic fevers that has been introduced in the past two decades. This syndrome is a common cause of periodic fevers in children, but often is not diagnosed by physicians. Furthermore, the choice of therapy is still unknown. Therefore, in this study 25 patients with PFAPA syndrome have been evaluated for demographic descriptions, presentation, clinical course, therapeutic responses and outcome of disease.

Methods: This study is a case series one that has been done on the base of patient's Files, interview, physical examination and filling the questionnaire for 25 patient who have been diagnosed with this syndrome over a 4-year period at Imam Khomeini Hospital Pediatrics Rheumatology Clinic in Ardabil.

Results: In this study, 72% of cases were male. The syndrome began at a mean age of 27.5 ± 13 months old. Twelve percent of patients had a family history of periodic fever. Episodes of fever occurred at the mean intervals of 30.2 ± 16 days and lasted a mean of 3.8 ± 1.3 days. Ninety two percent of patients had fever above 39°C (axillary). Fever and malaise were in 100% of patients, pharyngitis in 84%, chills in 76%, cervical adenopathy in 60%, nausea in 56%, aphthous stomatitis and vomiting in 52%, cough and abdominal pain in 48%, headache in 36%, Rash and conjunctivitis in 16% and arthralgia in 12%. All patients responded to prednisolone therapy. Intermittent use of prednisolone resulted in complete resolution of the episodes in 64% of patients and in the rest of cases the episodes recurred at the mean intervals of 12 weeks that were controlled with a single dose of prednisolone. Long – term sequel did not occurred.

Conclusions: clinical features of patients varied in different studies. Therefore, to identify the patients, the completion of Diagnostic criteria is a priority. Treatment with prednisolone is highly effective which can also be used as a diagnostic criterion in patients that may have PFAPA syndrome.

Key words: Periodic fever, PFAPA syndrome, Prednisolone.