Pediatric Rheumatology

Poster presentation

Efficacy of tonsillectomy in a family with a PFAPA-like phenotype

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Background

PFAPA syndrome is one possible cause of periodic fever in childhood.

The etiopathogenesis is unknown. The diagnosis is clinical and differential to exclude other similar diseases. Laboratory findings show only non-specific acute inflammatory response. The disease usually appears in early childhood. The most effective therapy is one-two low doses of oral corticosteroid, which however doesn't prevent recurrences. Effectiveness of tonsillectomy is still debated. Family history of the disorder is usually negative and it's unclear whether there is a genetic defect.

Case report

Female, age 2.5 years, clinical manifestations and family history suggestive of PFAPA syndrome.

The girl had a history of high fever with pharyngitis and cervical lymphoadenitis, almost every 15 days from the age of 6 months. Fever disappeared after steroid therapy (betamethasone). During the febrile episodes, acute phase reactants were increased. Between the episodes, the girl was well with a normal growth. The main infectious diseases and the most common causes of monogenic fevers were excluded. The patient's family tree shows that almost all the members of her mother's family had a similar clinical history in childhood, and in all the cases clinical manifestations disappeared after adenotonsillectomy.

Our patient was treated with adenotonsillectomy in October'07 and, to date, she no longer had fever or other PFAPA symptoms.

Conclusion

Although the mechanisms underlying this syndrome are unknown, tonsillectomy can be offered as an effective intervention for PFAPA syndrome. Even though PFAPA syndrome has no documented genetic basis, this family history is very interesting and should be further studied.