Diagnosis Process of PFAPA Syndrome in a Case Who Applied to the Pediatric Emergency Department with the Complaint of Fever and Later Found High CRP

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Abstract

Physical examination revealed aphthous stomatitis and pharyngitis in the patient who applied to us with the complaint of fever. It was learned that the patient’s complaints were constantly recurring. Upon this situation, a preliminary diagnosis of PFAPA syndrome was considered and all complaints regressed after treatment.

Keywords: CRP; Fever; PFAPA syndrome

Introduction

When fever is observed in children, infectious diseases come to mind first. In addition, some diseases with recurrent fever attacks and healthy periods between attacks have also been defined under the name of periodic fever syndromes. PFAPA syndrome, which is characterized by periodic fever, aphthous stomatitis, pharyngitis and adenitis, is a rare periodic fever in childhood with an unknown incidence. The characteristic feature of the syndrome is high fever, which is seen in all patients, recurs regularly every 3-6 weeks, lasts 3-6 days, starts suddenly and can reach 41°C. Although the clinical picture of the syndrome is well defined, the absence of a disease-specific laboratory finding makes it difficult to diagnose. Treatment options for the disease include steroids, cimetidine and tonsillectomy. Antibiotics, acetaminophen and nonsteroidal anti-inflammatory drugs are ineffective in treatment [1-6].

Case Report

A two-year-old girl applied to the Pediatric Emergency Service with fever. In his history, it was learned that his fever recurred at intervals of two weeks. On physical examination, fever was 40 degrees, aphthous stomatitis and pharyngitis were present. Other system examinations were normal. In the examinations, hemoglobin: 10.1 mg/dl, leukocytes: 12.3 thousand and CRP: 237 mg/l were found. Other examinations were normal. The patient, who was thought to have PFAPA syndrome, was admitted to the pediatric service for further follow-up and treatment. Antibiotherapy was not started after consent was obtained from the family. It was stated to the family that a single dose of methylprednisolone treatment could be tried. After methylprednisolone IV was administered at 1 mg/kg, all of the patient’s complaints completely regressed. It was observed that the CRP value decreased to 177 in the blood control taken at the 6th hour of the patient, and the CRP value decreased to the normal limits in the blood control taken on the 3rd day. The patient was followed up with the diagnosis of PFAPA. He was referred to the Department of Medical Genetics to be examined for Familial Mediterranean Fever.

Brief Discussion and Conclusion

PFAPA syndrome should be considered in the differential diagnosis of recurrent upper respiratory tract infections and recurrent febrile patients. In our case, a dramatic response was obtained to methylprednisolone. It should be kept in mind that high acute phase reactant levels can be detected in PFAPA syndrome. When the diagnosis of PFAPA syndrome is made, unnecessary antibiotic use will be prevented. This case is presented for the purpose of creating physician awareness and contributing to the literature.
References