Fedorenko Olha, Mykhailova Oleksandra , Gaben Moatasem

CLINICAL CASE OF A RARE DISEASE IN CHILDREN'S PRACTICE

Department of Pediatrics No.1 with Neonatology
Kharkiv National Medical University
Kharkiv, Ukraine
Scientific advisor: Doctor of medicine,prof. Riga Helen, c. med. s., assoc. prof. Uryvaieva Marina

Introduction: Marshall Syndrome (PFAPA syndrome) is characterized by Periodic Fever, Aphthous Stomatitis, Pharyngitis, Adenitis. It typically starts in early childhood (between ages 2 years and 5 years) and tends to be more common among males. Although genetic causes have not been determined, this syndrome tends to be grouped with hereditary fever syndromes. PFAPA syndrome is an immune mediated disease characterized by a cytokine dysfunction; moreover, the strong familial clustering suggests a potential genetic origin of the syndrome. Its treatment depends on early diagnosis, proper conservative or surgical treatment, this disease can be acquainted by different specialists of many branches of medicine: pediatricians, family physicians and otolaryngologists. Currently there is 374 of PFAPA syndrome registered cases in literature, Prevalence 2.3 per 10,000 children.

Objective: Analysis of the clinic of anamnestic data and differential diagnosis of rare pathology in a child with recurrent chronic tonsilitis.

Materials and methods: To demonstrate a rare pathology, we present you with our own clinical case, which has been discovered in children's polyclinic №2 in Kharkiv. The parents of a 4 years old complaining about a rise of temperature up to febrile figures within three days, signs of intoxication and refusal of the child to eat due to the presence of a rash on the mucous membranes of the oral cavity. Three days later she fell ill, the doctor examined her and prescribed antipyretics and antiviral therapy, the treatment was without any effect. From anamnesis it is

known that similar episodes of increase temperature and rash on the mucous membrane of oral cavity also plaque on tonsils was disturbing her during the last year almost monthly with an interval up to three weeks. Each time hyperthermia treatment was the primary objective in addition to prescribing antibacterial agents.

The girl was examined with the exception of: persistent of viral infection (herpes virus), primary immunodeficiency and diffuse disease of connective tissue.

She was examined by an otolaryngologist as a result tonsillectomy was recommended after permission of child's cardio rheumatologist. From the anamnesis of life it is known that the girl is susceptible for undergoing tonsillectomy because all of the children in the paternal line suffer from chronic tonsillitis and most of them underwent tonsillectomy. At the object of the study draw attention to the signs of intoxication: crying, pale skin, inadequate contact, decrease physical development for her age, when examining the oral cavity - plaques on the tonsils (otolaryngologist diagnosed her with chronic tonsillitis), signs of stomatitis on the mucous membrane and the presence of enlarged submandibular and anterior cervical lymph nodes.

Organ systems are without alterations. The girl underwent clinical, laboratory and instrumental methods to rule out exacerbation of chronic viral infections (herpes simplex virus, cytomegalovirus infection by polymerase chain reaction), atypical infections, coronavirus disease and diffuse connective tissue diseases.

According to instrumental methods of examination: electrocardiography and ultrasonography – no deviations where observed.

Results: Given the course of the disease, the results of clinical examination and research, differential diagnosis, as well as the severity of the reduction of hyperthermic syndrome, Marshall syndrome (PFAPA) was suggested. Due to this syndrome the course of treatment has shifted and glucocorticosteroid was administered once, after which the temperature immediately returned to normal and did not rise again.

Conclusion: for the treatment of hyperthermic syndrome with bacterial infection and the presence of stomatitis in children otolaryngologist should consulted. Before performing tonsillectomy, consider the possible presence of Marshall's syndrome (PFAPA) and carry out the treatment with corticosteroids.