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AUTOIMMUNITY, INFLAMMATION AND DYSREGULATION

PFAPA SYNDROME IN UKRAINE

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Background:

PFAPA-syndrome is most common periodic fever in pediatric population. Pathogenic pathways are still unknown and treatment is not completely standardized. Objective of this study was to describe clinical picture of periodic fever, aphthous stomatitis, pharyngitis and cervical adenitis in Ukrainian children.

Methods:

Our study was retrospective. The diagnosis of PFAPA-syndrome was established according to the Feder's criteria. We collected data from 40 children with PFAPA-syndrome and analyzed clinical picture, laboratory and demographic parameters. All cases were confirmed in reference center of pediatric immunology in Kyiv.

Results:

We observed delay in diagnosis for 1-1.5 years in more than 50% of children. All of them had pharyngitis, 80% cervical adenitis, 40% aphthous stomatitis and 15% of children three symptoms together. Additional symptoms such headache, abdominal pain, rash, vomiting, arthralgia, diarrhea were in 25% of children. 3 patients had disease manifestation in age older than 5 years. Elevated ESR, CRP, leukocytosis were observed in 80% of children. 37 patients originated from Ukrainian families, one - Turkish family, one - Syrian family and one Armenian. We observed shortness of symptoms free intervals in those on steroid therapy in more than 50% of children. Tonsillectomy - in two patients.

Conclusion:

We describe the largest group of PFAPA patients presented in Ukraine. We showed no significant differences in clinical picture between Ukrainian cohort and cohorts from other countries. We confirm that PFAPA-syndrome may have broad spectrum of other symptoms that not always accord with traditional criteria. Low awareness of Ukrainian pediatricians about PFAPA-syndrome leads to significant delay in diagnosis.