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Tonsillectomy in a patient with PFAPA and 7p22 deletion syndrome

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Syndrome is a rare cause of regular, repeated episodes of fever, sore throat and swollen neck glands in children although the exact etiology of PFAPA syndrome is unknown; one hypothesis suggests that the presence of infections and aberrant cytokine regulation are successive steps in a single ethiopathogenic process. The management of PFAPA is now considered to need the cooperation of ear, nose and throat (ENT) specialist. The role of tonsillectomy in literature is controversial. Some studies reported high success rates with tonsillectomy, further investigations are needed with larger numbers of patients. However, it is uncertain whether adenoidectomy combined with tonsillectomy adds any additional benefit to tonsillectomy alone. A 2-year Caucasian/Azerbaijan girl demonstrated repeated fever episodes with high levels (90-200mg/L) of C-reactive protein (CRP) since 6 months. In neonatal period she was observed because of microcephalus, slight developmental delay and growth retardation, muscle hypotonus and dysmorphic phenotype (broad forehead, hypertelorism, micrognathia and retrognathia, fluffy eyebrows, long and tight eyelashes, long filtrum, narrow lips). She was consulted by genetic and diagnosed with 7p22 microdeletions. During a period of 10 month she was hospitalized 6 times with the high fever, cervical/adenitis and sore throat Abdomen ultrasound, chest X-ray, EKG and EHHOKG were normal. Blood test for ANA, HIV, EBV, Borreliosis, tuberculosis, urine test and urine culture revealed no pathology. Cervical ultrasound showed increased lymphoid nodules with normal structure. There were no episodes of neutropenia. Procalcitonin level and blood culture were repeatedly negatives. Brain MRI with spectroscopy was performed to exclude intracranial pathology because of congenital problems. The patient was under ENT supervision to exclude the presence of otitis media during the high fever episodes. Adenoid hypertrophy was considered.

PFAPA was diagnosed by a pediatrician (repeated episodes of fever with aphthous pharyngitis, cervical/adenitis and high CRP levels, absence of neutropenia). Sequencing of genes was performed to exclude MEFV, MVK, TNFRSF1A, IL1RN and other gene abnormalities, using Illumina TruSight One expanded panel (6700 genes). No monogenic fever syndrome was revealed.

Prednisolone treatment 1mg/kg per os was used twice with excellent but temporary effect. Adenoidectomy was performed. The tissue of adenoids and tonsils has all signs of chronic inflammation. After the surgical treatment in a period of 7 months the patient was ill 4 times with no high fever (gastroenteritis, conjunctivitis, rhinopharyngitis and varicella with acute otitis media) and just once needed antibiotic treatment.

Biography

Andrei Antonov has completed his Medical School in 2001 from S. M. Kirov Military Medical Academy, Saint-Petersburg, Russia and then postgraduate studies in ENT from Tartu State University, Estonia. Currently he works as Head of ENT department in Tallinn Children Hospital and also in Tartu University ENT Clinic and North Estonia Medical Centre, Tallinn.

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