Surgical outcomes after tonsillectomy in children with Periodic Fever, Aphthous stomatitis, Pharyngitis and Adenitis syndrome (PFAPA)

Dytrych, P., Katra, R., Jurověč M., Skřivan J.

Department of ENT, 2nd Faculty of Medicine, Charles University in Prague and Motol University Hospital

Objectives
PFAPA (Periodic Fever, Aphthous stomatitis, Pharyngitis, Adenitis) syndrome is a benign inflammatory disease of childhood characterized by recurrent febrile episodes associated with aphthous stomatitis, pharyngitis and cervical adenitis. The pathogenetic background of the disease is unknown. The most effective non surgical treatment is episodic administration of corticosteroid. A positive effect of tonsillectomy was also described.

Purpose
To asses the efficacy of tonsillectomy in children with PFAPA and to evaluate histologic and cultivation findings in PFAPA tonsils.

Methods
14 patients with PFAPA syndrome underwent adeno/tonsillectomy using cold steel technique (between May 2011 and May 2016) (Fig.1). In order to exclude other causes of recurrent fevers all patients underwent general pediatric and ENT assessments (Tab. 1).

Diagnostic algorithm
The case history
Laboratory evaluation:
- erythrocyte sedimentation rate, full blood count with differential, liver and kidney function tests, anti streptolysin O.
- nose and throat culture
- serum immunoglobulin (Ig) levels (namely IgG, IgA, IgD), herpetic virus serology and mevalonic acid in urine collected during the febrile episode

Results
The median age at diagnosis was 39 months (range 14–78). The median duration of fever flare was 4,5 days (range 2–7) with a median interval between attacks of 4 weeks (range 2–7). Tonsillectomy was performed at a median age of 66 months (range 36–96). Pharyngitis was observed in all 14 patients (100%), aphthous stomatitis in 5 patients (35%) and cervical adenitis in 6 patients (43%) during PFAPA flares. Arthralgia was observed in 3 patients (21%), abdominal pain in 2 patient (14%) and vomiting in 3 patients (21%) (Tab.2). 12 of 14 of the PFAPA patients (86%) had complete remission of the symptoms for at least 9 months after surgery. The PFAPA tonsil cultivations were negative for bacteria in all patients. The histologic examination showed features of chronic tonsillar inflammation in all PFAPA patients.

Fig.1

Conclusion
Tonsillectomy is an effective treatment for PFAPA syndrome.