



POSTER PRESENTATION

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Periodic fevers associated with celiac disease and marked increase of NK cells

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Case report

A five year old boy was referred to our department for a history of repeated fevers. Family history was uneventful, however his father suffered from repeated episodes of tonsillitis in childhood.

Our patient presented with a history of repeated fevers with a clinical signs of tonsillitis since he was 2.5 years old. Fever attacks recur approximately every 6 weeks and are associated with lymphadenitis, sometimes accompanied by aphthous stomatitis. His inflammatory markers were elevated in a time of fever episode, but did not reach very high values typical for periodic fever syndromes. Chronic fatigue and progressive anemia also formed a persistent part of his clinical presentation.

Immunological investigation revealed significant decrease of T lymphocytes (CD3+ cells between 25-40%, normal values above 60%, absolute number between $0.5-0.9 \times 10^9/l$, normal values above $1 \times 10^9/l$), and a surprising increase of NK cells reaching over 50% of lymphocyte population. Further detailed search for underlying cause led to an unexpected diagnosis of celiac disease. Appropriate diet was introduced and led promptly to a decrease of celiac disease associated antibodies and to an improvement of anemia and fatigue.

His fever attacks, however, did not resolve, and his highly pathological findings of low T cells and extremely high NK cells were still detectable, not responding to antiinflammatory and corticosteroid treatment. PET scan was performed and showed an accumulation of activity in a pharyngeal arch and tonsil area. Tonsilectomy was performed at his 6 years of age and led to a prompt improvement in his clinical presentation, with a complete resolution of febrile episodes. His abnormal laboratory findings in cellular immune parameters very slowly improved within the next 6 months, with an increase of

CD3+ T cells and drop of NK cells to their normal values.

Conclusion

Here we present an unusual combination of periodic fevers with celiac disease, accompanied by peculiar and highly significant changes in cellular immunity, with low T cells and high NK cells. Combination of celiac diet and tonsillectomy led to a resolution of all clinical problems. Slow and gradual improvement of deep immune disbalance in lymphocyte subpopulations took further 6 months after tonsillectomy before it reached almost normal values. Association of celiac disease and periodic fevers is extremely rarely mentioned in a literature, but might be considered in some cases. Aphthous stomatitis is a common feature of both celiac disease and PFAPA syndrome.

Consent to publish

Written informed consent for publication of their clinical details was obtained from the patient/parent/guardian/relative of the patient.

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