A CASE OF FAMILIAL MEDITERRANEAN FEVER WITH VASCULAR INVOLVEMENT

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Introduction. Familial Mediterranean fever (FMF) is the most frequent autoinflammatory disease caused by mutations of MEFV gene encoding pyrin. **Aim**. To describe the evolution of FMF with a particular vascular involvement. **Materials and methods**. A case a patient of Druze descent. **Results.** A 17 y.o. male patient presents with pain and hemorrhagic rash of the left lower limb. The patient is considered to be sick from the age of 11: after playing football the child started developing pain in the left leg (soft tissue) and abdominal pain, fever (38-39°C), malaise for 1 month. The leg was inflamed for 3 months. X-ray showed bony abscess, for which the patient was given Amoxicillin for 3 months. After 5 months, the opposite leg got involved. In 2 weeks petechia developed on the lower limbs. The rash started on ankles and progressed upwards. Consequently, the child was diagnosed with small vessel vasculitis (IgA). The child was hospitalized and given corticosteroids 60 mg for 3 months. Genetic testing for FMF showed 2 mutations (FMF-V726A and FMF-E148Q). The child developed obesity, growth retardation from corticosteroid use. He continued having abdominal pain and fever twice a month while he was on colchicine 0.5 mg daily. The patient stopped having FMF attacks once he started canakinumab (a human anti-IL-1β monoclonal antibody neutralizing IL-1β signaling – therefore preventing the acute inflammatory response following aberrant inflammasome assembly in pyrin mutations). **Conclusions.** FMF patients may develop vasculitis, IgA deposit being the most frequent one. FMF is a potentially debilitating disease with a poor response to conventionally used colchicine and corticosteroids. High doses of corticosteroids should be avoided in pediatric population. Biologic therapy should be prompted in severe cases as early as possible in the course of disease. **Keywords:** Familial Mediterranean Fever, MEFV gene, vasculitis.