Pediatric Rheumatology

Poster presentation

Anakinra for secondary amyloidosis in an adolescent with FMF and Behcet Disease

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Familial Mediterranean Fever (FMF) is associated with mutation in the gene coding for pyrin which lead to accentuated innate immune responses involving the IL1 and probably Th1 pathways. We present a teenager who had FMF and Behçet disease and developed secondary amyloidosis. We hypothesized that anti-IL1 treatment would be beneficial for both controlling the disease activity and maintaining renal function.

A 17 year old girl with FMF (M694V/M694V) and Behcet disease unresponsive to colchicine (2 mg/day), methotrexate and indomethacine therapy developed proteinuria during her therapy of eight years. She has arthritis, fever and erythema nodosum as well as aphtous lesions twice a month. She suffered a poor qualitiy of life and often unable to attend school. Acute phase reactants, ESR 89 mm/hour, CRP 6.4 mg/dl were high independent of her attacks. 10 months ago she developed proteinuria of 300 mg/dl proteinuria with dipstick and 1.8 gr/day proteinuria was found. Her renal function tests were normal.The renal biopsy confirmed the diagnosis of secondary amyloidosis.

Anakinra was started at 1 mg/kg/day subcutaneously without stopping colchicine treatment with a special permission from the Ministry of Health. Izoniasid was also started because of the marked tuberculin reaction. In the following 6 months the patient was free of clinical symptoms. Acute phase reactants decreased. The level of proteinuria did not increase and renal functions remain stable.

We suggest that secondary amyloidosis due to autoiflammatory diseases is a new indication for the use of anti-IL1 treatment.