

Relapsing polychondritis in a patient with familial Mediterranean fever and amyloidosis

Sirs,
 Familial Mediterranean fever (FMF), a common autoinflammatory disease in eastern Mediterranean populations, is characterized by self-limited episodes of fever, serosal and synovial inflammation (such as peritonitis, pleuritis, and arthritis), as well as myalgia and erysipelas-like erythema (1, 2). Its most severe complication, amyloidosis, develops in 10-15% of patients (1). Relapsing polychondritis (RP) is a rare multi-systemic disease of unknown etiology that is characterized by recurrent inflammation and subsequent progressive destruction of the cartilaginous structures and connective tissue. The clinical features are auricular chondritis, saddle nose deformity, laryngotracheal/bronchial chondritis, scleritis/episcleritis and oligo/polyarthritis. RP can be associated with diverse forms of autoimmune or myelodysplastic diseases (3, 4). We report here a male patient with a 16-year history of FMF who presented with relapsing polychondritis and nephrotic syndrome due to amyloidosis.

A 34-year-old man was admitted to our hospital with a 6-month history of recurrent, painful, erythematous swelling – first of the left ear, and then the right ear, that spared the earlobes. The patient had undergone an emergency appendectomy at the age of 8, but did not experience any episodic pain until the age of 18. Since then he had been suffering from recurrent attacks of fever, abdominal pain, and peripheral arthritis once a month and lasting from 1 to 3 days. He had also a strong family history of FMF. He was diagnosed with FMF and started on colchicine (1.5 mg/day) 8 years ago. While using colchicine (1.5 mg) regularly he was still experiencing 2 or 3 attacks per year, the last episode being 5 months ago. Six months before his admission, he noticed a painful red swelling in the upper part of the pinna of the left ear. The inflammation lasted for 2 months, and then disappeared after he received several injections of dexamethasone, but reappeared 2 months later, this time on the right side.

His physical examination, including a laryngeal examination, was unremarkable except for the red, swollen and tender right ear (Fig. 1). An exudative and hemorrhagic lesion was observed on the left eye fundus. Pulmonary function tests were normal, and no abnormalities with the cartilaginous structures in the thorax appeared on CT. He had a high acute phase response with slight anemia, leukocytosis and thrombocytosis. His daily urinary protein excretion was 7 g and creatinine clearance was 41 ml/min. He was homozygous for the M694V mutation. AA amyloid deposition was demonstrated in the rectum and kidney biopsies. After one week of oral prednisolone therapy (1 mg/kg) the patient's ear symptoms disappeared. He was discharged on colchicine (1 mg/day)



Fig. 1. Auricular chondritis.

and prednisolone (50 mg/day), which was tapered to 10 mg in 4 months; infliximab was given for 2 courses later on. During outpatient follow-up, he experienced 4 abdominal attacks, but there was no recurrence of auricular symptoms. On the 11th month, maintenance hemodialysis was started due to a deterioration in renal function.

Our FMF patient developed RP, which was diagnosed at the same time as secondary amyloidosis. The latter presumably must have been present long before the first episode of chondritis. RP has shown a rather mild clinical course in our patient, probably due to treatment with both corticosteroids and colchicine (4). It was limited to auricula and manifested in the form of just one episode in either ear. It did not resemble the pseudo-erysipela that is commonly seen in FMF, since inflammation spared the ear lobes. Moreover, he had also retinal hemorrhage, supporting the diagnosis of RP. On the other hand, chronic renal failure due to secondary amyloidosis followed a rapid course and progressed to end-stage renal disease within 1 year of the diagnosis.

A literature search (PubMed) disclosed several other conditions that may co-exist with FMF (Table I) (5-16). The list is quite similar to one published in a recent survey by the Turkish FMF Study Group (2). At the same time, auricular chondritis could well be a rare manifestation of FMF, similar to

Table I. Reported conditions associated with familial Mediterranean fever (ref.).

Polyarthritis nodosa (5)
Henoch Schönlein purpura (5)
Multiple sclerosis (6)
Seronegative spondyloarthropathies (7)
Inflammatory bowel diseases (8)
Uveitis, episcleritis (9, 10)
Aseptic meningitis (11)
Immunoglobulin A nephropathy (12)
Behçet's syndrome (13)
Mesothelioma (14)
Optic neuritis (15)
Systemic lupus erythematosus (16)

uveitis or episcleritis (9, 10). However, the longer duration of the episodes and the good response to corticosteroids in our patient militate against this hypothesis. Finally, coincidence is obviously a possibility.

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