Scleritis and sudden hearing loss associated with familial Mediterranean fever

Sirs,

Familial Mediterranean fever (FMF) is an autosomal recessive disease caused by mutations in MEFV gene. Typical manifestations of the disease include recurrent attacks of fever, serositis and arthritis. Eye involvement is rare in FMF, and to our knowledge, sudden sensoryneural hearing loss (SNHL) has not been reported so far.

A 43-year-old man presented with redness and pain in his right eye in September 2007. He had had a 6-year history of recurrent episcleritis attacks. He was diagnosed as having FMF when he was 5 years old, with M694V mutation in both alleles. He had been taking colchicine 1 mg/day and sometimes developed pain and swelling over the ankles after walking long distances. Physical examination was normal except for redness in the right eye. Complete blood count, liver function tests, serum electrolytes and urine analysis were normal. ESR was 13 mm/h and CRP was 10 mg/L (normal <6). Diagnosis of scleritis was made by an ophthalmologist, and methylprednisolone 1mg/kg/d, and topical dexamethasone was commenced. Scleritis resolved rapidly. He had not experienced recurrence of scleritis until now.

In November 2008 he was admitted to the ENT clinic with sudden hearing loss. External auditory canals and tympanic membranes were normal on physical examination. CRP was 24mg/L. Odiometry showed SNHL in the right ear (Fig. 1a). Methylprednisolone 80mg/d was initiated with the diagnoses of sudden SNHL. He rapidly improved and control odiometry was normal (Fig. 1b). He later developed three more sudden SNHL attacks in the same ear, which were treated with corticosteroid and immunosuppressive agents (methotrexate or azathioprine). Magnetic resonance imaging (MRI) of temporal bones and thoracic MR angiography were normal. Antinuclear antibodies, anti neutrophile cytoplasmic antibodies, anti cytomegalovirus Ig M, anti herpes simplex virus Ig M, anti Ebstein-Barr virus EA, anti EBV VCA Ig M and anti EBV EBNA Ig M antibodies were negative. He developed the last attack while taking azathioprine 75mg/d for three months. The azathioprine dose was increased to 150mg/d, and methylprednisolone 60mg/d was initiated. Hearing loss ameliorated in several days. Colchicine dose was increased to 1.5 mg/day. He has been receiving azathioprine 150mg/d and colchicine 1.5mg/d, with no sudden SNHL attack or arthritis for the last 7 months.

Yazici and Pazarli first documented an FMF patient complicated by uveitis and episcleritis (1). Scharf *et al*. reported two adult patients with inactive FMF and episcleritis (2). They concluded that episcleritis was a manifestation of FMF despite a seemingly inactive disease. Hirsh *et al*. reported bilateral panuveitis in a male patient with FMF (3). Increasing the colchicine dose resulted in complete resolution of the panuveitis. Akman *et al*. described panuveitis and episcleritis in two siblings with FMF (4). Episcleritis responded well to NSAIDs and topical corticosteroids in the younger sibling.

Our patient had high serum CRP levels and experienced bilateral ankle swelling when walking long distances, which show the activity of the disease despite the lack of fever or abdominal attacks. After augmentation of colchicine to 1.5mg/d, his ankle pain and swelling resolved.

Cogan's syndrome (CS) should be included in the differential diagnosis of patients presenting with ocular and audiovestibular symptoms (5, 6). Our patient had scleritis and SNHL, but the interval between onset of eye involvement and SNHL was about 6 years, which was not consistent with either typical or atypical CS.

Fong *et al*. showed vasculitis in 75% of scleral biopsy specimens of 30 patients with scleritis(7). Occurence of sudden SNHL in some types of vasculitis (8, 9) also suggests a vasculitic etiopathogenesis for sudden SNHL. Because some vasculitides have been reported to occur more frequently in FMF than in the normal population (10), it is conceivable that scleritis and SNHL might be a vasculitic manifestations of FMF in our patient.

We think that inadequate colchicine dose and ongoing disease activity seem to be responsible for the development of scleritis and sudden SNHL in our patient. Given the probable vasculitic nature of the eye and inner ear involvement in FMF, if the increase of the colchicine dose fails, immunosupressive drugs, such as methotrexate and azathioprine, would be reasonable choices for treatment of these manifestations.

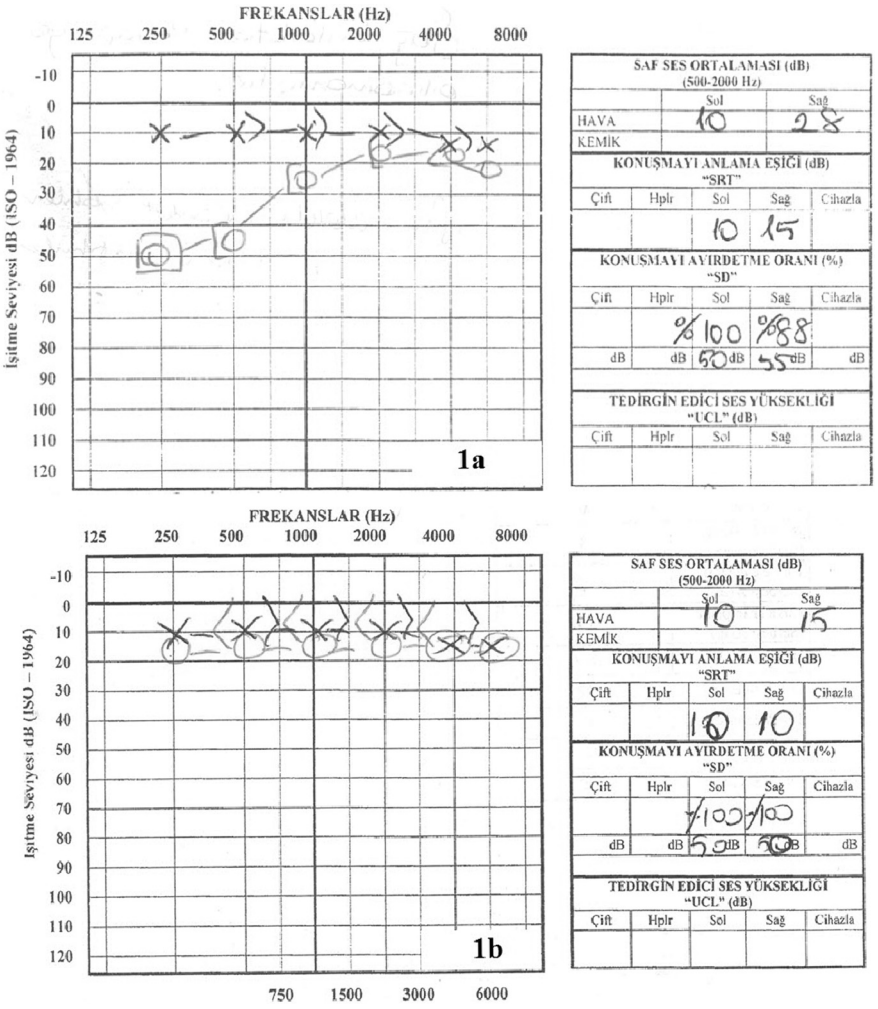

Fig. 1. Odiometry showing sensoryneural hearing loss in right ear (**a**) , and normal repeat odiometry carried out 10 days later (**b**).

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