

Immune mediated sensorineural hearing loss in a myositis patient

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An unusual case of an immune mediated sensorineural hearing loss in a myositis patient is described. A 24-year-old male developed progressive sensorineural hearing loss (SNHL) and subsequently clinical picture compatible to acute myositis. He was ANA positive. The diagnosis of myositis is based on clinical picture, aldolase levels and electromyogram (EMG) findings although, muscle biopsy was not performed due to the patient's refusal. Laboratory investigation for infectious myopathies, sarcoidosis as well as systematic vasculitides, Cogan syndrome or secondary myositis due to connective tissue disorders like systemic lupus or Sjogren syndrome was negative. He received high doses of steroids (1,5 mg/kg BW) and clinical status, CRP, aldolase levels and EMG findings were normalized, except for the SNHL where there was no improvement. During the follow-up period the patient had steady transaminasaemia. Although definite autoimmune liver disease was not confirmed by liver histology, azathioprine 100 mg daily was added, and transaminases declined to normal values. We conclude that, patients with inflammatory muscle disease and hearing disturbance should be considered to and investigated for immune mediated inner ear disease. *Hippokratia* 2005; 9 (3): 132-133

Key words: *immune mediated inner ear disease, sensorineural hearing loss, myositis*

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Immune mediated inner ear disease (IMIED) is a syndrome that is characterized by subacute onset of sensorineural hearing loss, often accompanied by vestibular symptoms. It may develop as a primary disorder, without any organ involvement, or complicates systemic diseases like vasculitides (Wegener's granulomatosis, polyarteritis nodosa and Cogan's syndrome) and connective tissue disorders (mainly SLE and Sjogren's syndrome)¹. Here we describe a patient with IMIED who subsequently developed primary myositis.

Case history

A 24-year-old man was admitted to the Otorhinolaryngology department because of a 70-day history of progressive sensorineural hearing loss (SNHL). His hearing impairment was initially presented unilaterally, accompanied by vertigo and tinnitus. On admission, he had bilateral SNHL confirmed by serial audiograms (Fig. 1). His past medical history was clear, and he had not previously taken ototoxic drugs. Prednisolone 1 mg/Kg BW was administered for 20 days without any response. A few days after the cessation of steroid treatment he manifested a high fever, fatigue, severe proximal extremities myalgias and was transferred to our department. On clinical examination, he had symmetrical involvement of proximal muscles (pain, weakness and difficulty in rising from a squatting position). Laboratory investigation showed a white-cell count of 20,100/mm³ (79% neutrophils), haemoglobin 13.1 g/dl, platelet count 210,000/mm³, alanine aminotranferase 91 IU/l, ESR 15 mm/h,

CRP 17.9 (0-5) mg/l, aldolase 12.5 (< 7) IU/l and normal aspartate aminotranferase, creatine phosphokinase and lactate dehydrogenase values. Renal, liver function tests and electrolytes levels were normal. ANA was positive 1:160 (Hep-2, speckled pattern). Other autoantibodies including rheumatoid factor, anti-dsDNA, anti-mitochondrial, anti-smooth-muscle, anti-extractable nuclear antigens (including anti-Ro, anti-La, anti-Sm, anti-Jo1, anti-RNP), anti-cardiolipin, ANCA were within normal limits. Immunoglobulins (IgG, IgM, IgA) and complement levels were also normal. Serology for hepatitis B virus, hepatitis C virus, HIV, herpes simplex virus, cytomegalovirus, Epstein B virus, Coxsackie virus as well as for Lyme disease and Syphilis (FTA-ABS) were negative. Blood cultures were negative. The chest radiogram was clear. The electromyogram (EMG) was typical for acute myositis showing spontaneous fibrillations, abnormal low amplitude, short duration, polyphasic motor potentials and bizarre high frequency discharges. The MRI-gadolinium scan of the inner ear and posterior fossa was normal. Echocardiography did not reveal any abnormal findings. The ophthalmological examination revealed no evidence of keratitis or retinal disease. The Schirmer test was normal as well. Muscle biopsy was recommended to the patient but he refused. Taken together the clinical, EMG and laboratory findings, in combination with the specific features of hearing loss (which developed over 3 months, and was sensorineural), the diagnosis of primary polymyositis with autoimmune inner ear disease was made. Treatment with high dose of

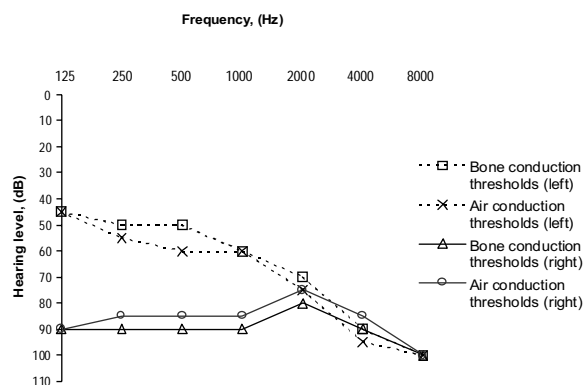


Figure 1. Audiogram of the patient which reveals high-frequency hearing loss.

methylprednisolone was commenced immediately (1.5 mg/Kg BW for 15 day-loading dose, tapered to a 16 mg/day-maintenance dose).

The patient's clinical status, CRP, aldolase levels and EMG findings were normalized, except for the SNHL where there was no improvement. Twenty-five per cent residual left ear hearing ability was finally retained. Immunosuppressives such as azathioprine were not added to steroid treatment because during the follow-up period the patient had steady transaminasaemia. The possible coexistence of myositis with autoimmune involvement of the liver, presented the case for a liver biopsy. Liver histology revealed mild fibrosis and infiltration of inflammatory cells-predominantly lymphocytes-in the portal tracts, sparse inflammatory lesions of hepatic lobules, and steatosis in approximately 60% of hepatocytes. Of note, steatosis in more than 50% of the patients with various types of collagen diseases attributed to dosage of administered corticosteroids is also observed by others². In our patient with seronegativity for viral hepatitis, the clinical, and biochemical data indicated probable autoimmune hepatitis, according to the International AIH group's scoring system (score without histology 11)²⁻⁴. Although definite autoimmune liver disease was not confirmed by liver histology, azathioprine 100 mg daily was added to the patient's treatment and transaminases declined to normal values.

Discussion

The patient was diagnosed as IMIED according to current syndrome definition (time course of hearing loss, bilateral involvement, characteristic SNHL audiogram)¹. On the other hand, proximal muscle weakness and pain, the EMG characteristic findings and the increase in aldolase in combination with ANA positivity lead to the

diagnosis of primary polymyositis (muscle biopsy was not performed due to the patient's refusal). Differential diagnosis from other inflammatory myopathies supported the initial diagnosis⁵. It should be noted that the patient never had skin manifestations or clinical and laboratory findings of other collagen and vascular systemic diseases. Furthermore, infectious myopathies were excluded by serological investigation.

IMIED is described in coexistence with SLE and aortic insufficiency, chronic discoid lupus, iritis and AS and other autoimmune diseases⁶⁻⁹. To the best of our knowledge, it is the first time that IMIED incorporates the initial manifestation of primary polymyositis. Moreover, probable autoimmune liver involvement in this patient comprises an interesting finding and could be considered as a part of the underlying connective tissue disease¹⁰. In conclusion, patients with inflammatory muscle disease or other autoimmune diseases and hearing disturbance should be considered to and investigated for IMIED, so that proper treatment will begin early to prevent irreversible hearing loss.

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