Case report

Sensorineural hearing loss in adult onset Still's disease

HARRY M MARKUSSE, BEREND STOLK, ANDEL G L VAN DER MEY, JOHANNA M DE JONGE-BOK, AND KAREL J HEERING

From the Departments of Rheumatology, Medicine, and Otorhinolaryngology, University Hospital, Leiden and the Departments of Rheumatology and Medicine, Bleuland Hospital, Gouda

SUMMARY A 26 year old man with a nine year history of adult onset Still's disease (AOSD) developed sensorineural hearing loss during an exacerbation of his disease. This complication has not previously been described in association with adult onset Still's disease. He responded favourably to steroid treatment.

Key words: deafness, juvenile arthritis.

A patient with a polycyclic systemic adult onset Still's disease (AOSD) is reported, who developed a sensorineural hearing loss during an exacerbation of his disease. The hearing loss and the systemic activity of AOSD were both controlled by corticosteroid treatment. To the best of our knowledge the association of sensorineural hearing loss with AOSD has not been published before.

Case report

In 1987 a 26 year old man presented with a nine year history of periodic illness which, in 1978, had been evaluated for quotidian spiking fever, evanescent rash, arthralgias, lymphadenopathy, leucocytosis, and liver enzyme abnormalities. At the time negative results were obtained in the following investigations: cultures of blood, stools and urine, Rose-Waaler test, antinuclear antibody, muscle biopsy, bone marrow biopsy, and *x* ray films of the chest, gall bladder, stomach, and sacroiliac joints. A diagnosis of AOSD was made, and salicylate treatment resulted in symptomatic improvement. During the following years splenomegaly was found and several exacerbations were successfully treated with salicylates. A flare up in 1986 responded to indomethacin. Another exacerbation in February 1987, however, failed to improve with indomethacin

150 mg daily. On 20 March 1987 he was admitted to hospital still suffering from quotidian spiking fever and arthritis of the ankles and metacarpophalangeal joints, an evanescent rash of the forearms and hands, and weight loss of 6 kg. A few days after admission he gradually developed hearing loss and tinnitus. Indomethacin treatment was stopped on 22 March 1987, but despite this a pure tone audiogram taken two days later showed a sensorineural hearing loss.

Investigations showed erythrocyte sedimentation rate (ESR) 72–94 mm/1st h, white cell count (WCC) $9.5\text{--}17.2 \times 10^9/\text{l}$, with a normal differential, and slightly raised liver enzymes. Creatinine, calcium, glucose, thyroxine, serum protein immunoelectrophoresis, vitamins B₁, B₆, and B₁₂, and serum folate, and 24 hour urinary calcium excretion were normal.

The following investigations failed to show any abnormality: rheumatoid factor titre, antinuclear antibodies, antibodies to native DNA, angiotensin converting enzyme, cryoglobulins, hepatitis B antigen, and syphilis serology. A lumbar puncture yielded clear, colourless cerebrospinal fluid with low cell counts and no protein; cultures of blood, stool, urine, and cerebrospinal fluid showed no growth. There was no sequential serological evidence for recent infection with chlamydia, varicella zoster virus, herpes simplex virus, echovirus, Cocksackievirus A and B, cytomegalovirus, adenovirus, mumps virus, or Epstein-Barr virus. *x* Rays of the chest and hands, electrocardiography, slit lamp examination

Accepted for publication 24 December 1987.

Correspondence to Dr Harry M Markusse, Staffcentre Rheumatology Building 1, C2-Q, University Hospital, PO Box 9600, 2300 RC Leiden, The Netherlands.

Fig. 1 Pure tone and speech audiograms. Because both ears disclosed the same abnormalities only the audiograms of the right ear are shown.

of the eyes, and pulmonary function tests were normal. Echography of the abdomen showed splenomegaly. Biopsy specimens from the nasal mucosa and the rectal mucosa and needle aspiration of the abdominal subcutaneous fat showed no abnormalities. A computed tomographic scan of the skull base and brain were normal. Audiological examination, including brainstem evoked response audiometry (BERA), showed a (cochlear type) sensorineural hearing loss of 40–50 dB in the speech frequencies, without any evidence of middle ear or retrocochlear localisation of the lesion (Fig. 1A).

The audiometric findings made two days and 19 days after refraining from indomethacin treatment were essentially identical (Fig. 1A). Twenty one days after the withdrawal of indomethacin, prednisone 60 mg daily was started. A prompt response was manifested by a fall in temperature, resolution of the arthritis, and a subjective improvement in the patient's hearing.

The fifth day after starting prednisone treatment there was clear improvement in both pure tone and speech audiometry (Fig. 1B). On the 14th day of treatment the improvement for both ears was even more obvious (Fig. 1C).

After four weeks the prednisone dosage was tapered to 25 mg daily; the ESR was 10 mm/h, WCC $7.5 \times 10^9/l$, and the liver enzymes were normal. There was no longer any fever, rash, or arthritis, the tinnitus gradually disappeared, and further progress in hearing acuity in both ears was found on audiometric control.

Discussion

Adult onset Still's disease is a cyclic systemic inflammatory disorder mostly diagnosed before the age of 35, characterised by high spiking quotidian fever, arthritis, and evanescent rash. Leucocytosis, lymphadenopathy, serositis, hepatosplenomegaly, and hepatic enzyme abnormalities are other features of this disease.^{1–3} In patients with a systemic disease and hearing loss, conditions such as Cogan's syndrome, polyarteritis nodosa, Wegener's granulomatosis, relapsing polychondritis, and systemic lupus erythematosus have to be considered.^{4,5} In this case these diseases were conclusively excluded by the long history, the clinical features, and the comprehensive investigations. This patient fulfilled all the AOSD criteria proposed by Calabro and Londino.⁶ Although the predictive value of these criteria has never been tested, we believe that in this case a diagnosis of AOSD is justified. The cyclic systemic disease activity with only mild articular involvement was characteristic. After nine years he developed hearing loss and tinnitus during another exacerbation manifested by generalised malaise, weight loss, high spiking fever, mild arthritis, leucocytosis, raised hepatic enzyme concentrations, splenomegaly, and an evanescent rash.

Tinnitus, normal otoscopy, pure tone audiometry, speech audiometry, tympanometry, and BERA pointed to cochlear involvement.

Many drugs may cause audiovestibular toxicity, but we could not find any report of indomethacin

related sensorineural hearing loss. The time between the introduction of indomethacin treatment and the onset of cochlear symptoms, the lack of improvement of the hearing after indomethacin was stopped, and the striking amelioration of the cochlear function after starting prednisone treatment virtually exclude indomethacin as a cause of this hearing loss.

Extensive viral and bacteriological studies were unremarkable and a normal computed tomogram of the skull and a normal lumbar puncture made a central nervous system process unlikely.

The simultaneous improvement of this patient's cochlear function and his general condition immediately after the start of prednisone treatment make AOSD the most likely cause of the hearing loss.

References

- 1 Larson E B. Adult Still's disease. Evolution of a clinical syndrome and diagnosis, treatment and follow-up of 19 patients. *Medicine (Baltimore)* 1984; **63**: 82-91.
- 2 Cush J J, Medsger Jr T A, Christy W C, Herbert D C, Cooperstein L A. Adult-onset Still's disease. *Arthritis Rheumatism* 1987; **30**: 186-94.
- 3 Wouters J M G W, van de Putte L B A, Cooperating Centres. Adult-onset Still's disease; clinical and laboratory features, treatment and follow-up of 45 cases. Adult-onset Still's disease. Nijmegen, 1986: 19-39. (Thesis.)
- 4 Haynes B F, Kaiser-Kupfer M I, Mason P, Fauci A S. Cogan syndrome. *Medicine (Baltimore)* 1980; **59**: 426-41.
- 5 Cardarelli D D, Rejowski J E, Corey J P. Sensorineural hearing loss in lupus erythematosus. *Am J Otolaryngol* 1986; **7**: 210-3.
- 6 Calabro J J, Londino Jr A V. Adult onset Still's disease. *Rheumatol* 1986; **13**: 827-8.