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Bilateral sudden deafness and acute acquired toxoplasmosis

MORTEN KATHOLM, M.D.,* NIELS JON JOHNSEN, M.D.,* CHRISTIAN SIIM, M.D.,† LARS WILLUMSEN, M.D.‡
(Denmark)

Abstract
An 18-year-old woman, while suffering from acute acquired toxoplasmosis, experienced sudden deafness and a total loss of vestibular function first in the right ear and three months later also in the left. Following treatment with sulphadiazine and pyrimethamine, hearing was retrieved to such a degree that the patient was enabled to communicate by means of a body-worn hearing aid and lip-reading. Taking the differential diagnostic possibilities into account, we believe that toxoplasmosis was the cause of the severe hearing loss. Since effective treatment seems to be available, we recommend that patients with acute bilateral sensorineural hearing loss of unknown origin are examined for acute toxoplasmosis with a view to instituting chemotherapy.

Introduction

Sudden Sensorineural Hearing Loss (SSHL) may be associated with diseases of the inner ear and cochleo-vestibular nerve such as Menière's disease, perilymph fistula, bacterial meningitis and labyrinthitis, syphilis and acoustic neurinoma, but very often there is no clues to the aetiology. A multiplicity of potential causes seem available; many cases presumably represent the result of a virus infection (Schuknecht and Donovan, 1986) but the possibility that other microrganisms may very occasionally be responsible cannot be excluded.

Toxoplasmosis is an infectious disease caused by a protozoan, *Toxoplasma gondii*. The cat is the principal host and may excrete infectious oocysts. Infection in man occurs after ingestion of contaminated, undercooked meat or by contact with infected animals (Krick and Remington, 1978). The ingested parasites invade the epithelium of the intestine, multiply in the mesenteric lymph nodes and later spread haematogenously to other organs (Krick and Remington, 1978). The acquired infection is common. In Denmark, the prevalence of increased antibody titres among adults is 30–50 per cent (Siim, 1961), and similar figures have been reported from other parts of the world (Krick and Remington, 1978; Aspock, 1985). The most common clinical manifestation is lymphadenopathy which may be local or generalized (Siim, 1961; Krick and Remington, 1978). There may be a slight rise in temperature and general malaise but in most cases the infection is subclinical and remains undiagnosed.

Usually the infection lasts a few months, but in some cases *T. gondii* may survive for years in tissue cysts and in the immunocompromised host the organism may become reactivated (Townsend *et al.*, 1975; Ruskin and Remington, 1976; Krick and Remington, 1978). Affection of the central nervous system may occur but is uncommon in the immunocompetent patient (Townsend *et al.*, 1975; Ruskin and Remington, 1976). Hearing is rarely affected in acquired toxoplasmosis as far as can be judged from the very few reports available on the subject (Wilke, 1961; Grimaud *et al.*, 1961; Partsch and Rieder, 1962; Rost, 1963). In the present study we found that acute, acquired toxoplasmosis presumably was the cause of a severe bilateral hearing loss in a young and otherwise healthy woman. Since this conclusion seemed of consequence for the further management of the patient, we find it of interest to present the case and to discuss the differential diagnostic possibilities.

Case report

An 18-year-old woman presented in the spring of 1980 because of recurrent episodes of otosalpingitis. She had previously been healthy apart from slight hay-fever. There was no predisposition to ear disease or hearing impairment and no history of neuroinfections or head trauma. She was treated with air insufflation (Politzer). The hearing improved though possibly not quite to its former level; moderate tinnitus occurred intermittently in the left ear.

In March 1981, episodes of fluctuating, left-sided hearing impairment occurred in association with gyratory vertigo, nausea, vomiting and aggravation of tinnitus. There was no pain or any sensation of 'fullness'. Otological and neurological examinations showed no abnormality. The audiogram is shown in Figure 1. The stapedial reflex could not be elicited on either side. As hearing thresholds were uncertain in the left ear, electrocochleography was performed indicating a conductive loss of 30 dB HL at 2 kHz in addition to a sensorineural high frequency hearing loss. Bilateral brainstem audiometry showed no signs of retrocochlear involvement and the Hallpike caloric vestibular examination showed normal responses. The conclusion of these tests was a diagnosis of otosclerosis, aggravated by Menière-like attacks. In the ensuing period there was no fluctuation of hearing and no dizziness, whereas tinnitus persisted on the left ear. The general condition of the patient was good.

In October 1981, the patient experienced acute gyratory vertigo with nausea and vomiting accompanied by severe right-sided tinnitus. She had no fever and there was no indication of any concomitant systemic illness. After a few days anacusis developed on the right side. The clinical examination revealed a normal otoscopy, there was a spontaneous nystagmus towards the left, but fistula test was negative. The vestibular function was now extinct on the right side. Electrocochleo-

From: *Departments of Audiology and Otorhinolaryngology, Gentofte University Hospital.
†Department of Otorhinolaryngology, Glostrup University Hospital.
‡Department of Infectious Diseases, University Hospital, Rigshospitalet, Copenhagen, Denmark.
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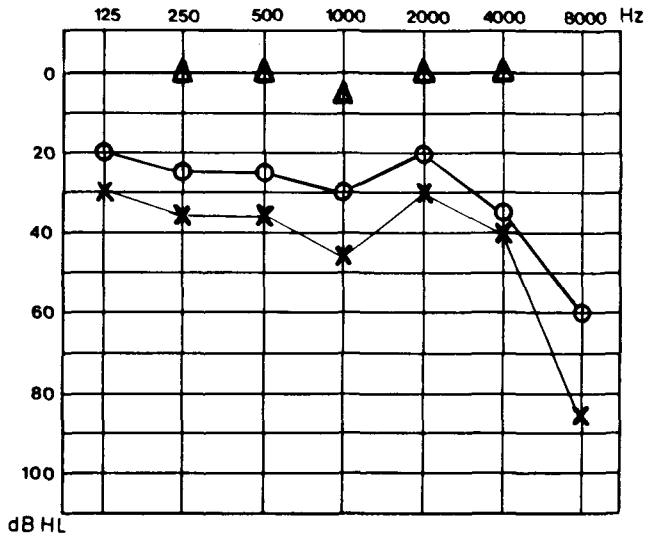

FIG. 1

Audiogram at time of first admission. △ indicates bone conduction level of best hearing ear. Discrimination: 100/100 per cent. Electrocochleography in the left ear showed a 30 dB conductive hearing loss at 2 kHz (see text).

graphy on the right side showed strongly reduced a cochlear microphonic and no identifiable action potential. The audiogram on the left ear was unchanged from previous examinations (Fig. 2) and brainstem audiometry showed no indication of retrocochlear involvement on this side. Neurological examination, EEG and spinal fluid examination were normal; there was no ataxia. Tomography of the temporal bone and CT scanning gave no suspicion of acoustic neuroma or other intracranial processes. The dizziness abated over a period of three weeks, whereas tinnitus and complete deafness persisted in the right ear.

In January 1982, the patient developed anacusis on the left ear within a few days and the vestibular function also became extinct on this side; there were no concomitant symptoms. Treatment with prednisone (20 mg × 4 daily) was instituted.

In the period between the two attacks of anacusis the patient developed a hazelnut-sized swelling on the neck; no other adenopathy or hepato-splenomagaly was found. The lymph node was removed and microscopy revealed changes characteristic of toxoplasmosis, but no indication of malignancy or haematological disorder. The parasite was not demonstrated at microscopy.

Titre analysis for toxoplasmosis showed a Sabin-Feldman-Dye test of 1:6250, IgG indirect fluorescence antibody-test (IgG-IFA) of 1:31250 and a complement binding titre of 1:256. IgM-IFA was positive at 1:10. The patient's parents had negative toxoplasmosis serology.

The steroid treatment which the patient had received for 10 days was stopped immediately and treatment was initiated with sulphadiazine 1 g × 4 daily, pyrimethamine 25 mg daily as well as injections of folinic acid 3 mg × 2 weekly. This regime was followed for six weeks, ending in March 1982. After three weeks of treatment the patient noted return of hearing on the left side and audiometry showed a slight improvement for the low frequencies. The hearing further improved on this side and a small degree of low frequency hearing was also probably retrieved on the right side. Gradually the hearing reached a serviceable level and the patient became able to communicate by means of a body-worn hearing aid and lip-reading (Fig. 3).

During the entire course the cardio-vascular, haematological and coagulatory status of the patient was normal. Our examinations gave no reason to suspect any neurological nor granulomatous disease, including tuberculosis and sarcoidosis. The ESR was not raised and rheumatological tests were within normal limits; the patient had normal blood sugar values. WR, TPI, Paul-Bunell, *Mycoplasma pneumonia* titre and cold agglutinins were not increased. Gammaglobulins were slightly increased but no M-components were demonstrated. Mantoux was negative; antinuclear antibodies could not be demonstrated, and chemotactic tests of granulocytes as well as lymphocytic transformation with various mitogens and antigens were normal. Cytomegalovirus complement binding titre was slightly positive (1:16) but parotitis, *Varicella zoster* and herpes complement binding titres were not increased. The opthalmological examination showed no signs of ocular toxoplasmosis or interstitial keratitis and the eye background was normal with normal vessels. CT scanning revealed no intracerebral calcifications and polytomography showed no otospongiotic foci in the cochlear capsule.

In March 1984, two years after treatment, the audiogram was unchanged compared with that shown in Fig. 3. The caloric vestibular examination now showed normal responses on both sides. The patient was followed twice a year until 1988. The final audiogram was identical with the audiograms from 1982

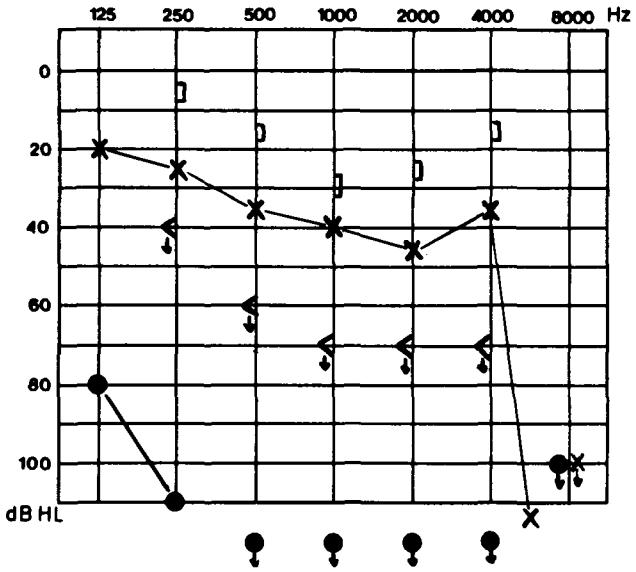

FIG. 2

Audiogram at time of the first anacusis. SRT: >100/35 dB HL. Discrimination: −/100 per cent

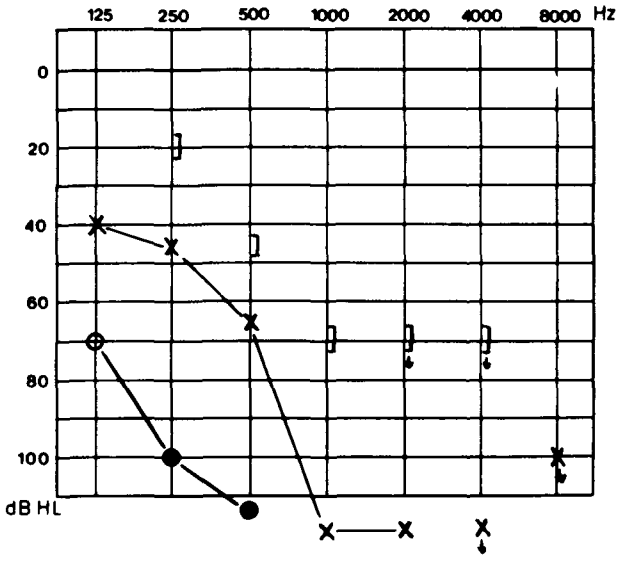

FIG. 3

Audiogram May 5 1982. SRT: > 110/95 dB HL. Discrimination: 0/0 per cent

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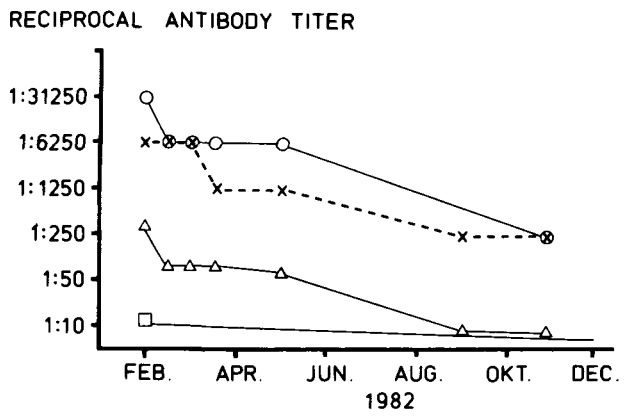

FIG. 4

Follow-up of Toxoplasma antibody titres. ○ IgG-indirect fluorescent antibody test, × Sabin-Feldman-Dye test. ▲ Complement fixation test. □ IgM-indirect fluorescent antibody test.

and 1984. Apart from the hearing loss the patient had no complaints and she was in good health.

Discussion

Although the patient reported here displayed Menière-like symptoms on the left side seven months prior to the occurrence of severe bilateral hearing loss, the further course of events was not typical (Stahle, 1976). Previous audiological examinations had shown indications of a conductive hearing loss eventually due to otosclerosis. Menière-like symptoms have been described in connection with cochlear otosclerosis (Paparella *et al.*, 1984); yet rapid deterioration of hearing or severe sensorineural hearing loss is mostly associated with pronounced otospongiotic changes of the cochlear capsule (Shambaugh, 1978).

Intra-cochlear membrane tears or rupture of the fenestra usually occur unilaterally in connection with trauma or sudden pressure changes (Goodhill, 1981), whereas the symptoms in the case presented here apparently emerged spontaneously. Politzer's test and Valsalva's manoeuvre never gave rise to dizziness and fistula test was negative. There was no ataxia. Polytomography and a CT scan of the skull and temporal bone revealed no perilymph fistula and gave no reason for suspecting developmental malformation which might conceivably cause or predispose to a fistula or membrane rupture.

The cause of the disease was not typical of autoimmune hearing loss in which case hearing deteriorates over a period of weeks or months rather than hours or days (McCabe, 1979). Our examinations revealed no signs of an autoimmune disorder; in particular there was no suspicion of Cogan's syndrome.

Our patient suffered from hay fever every summer but no allergic reactions were noted during the course of the hearing complaint; treatment with steroids had no effect on hearing ability. It is hardly to be expected that the bilateral cochleovestibular symptoms of this teenage patient showing no signs of a coagulatory disorder, vascular disease or conditions predisposing thereto were due to any cerebrovascular disorder affecting the inner ear. As toxoplasmosis is not uncommon in our country the simultaneous appearance of the infection and inner ear disease could be casual. Still we believe that toxoplasmosis was the cause of the severe hearing disorder in our patient; the following reasons support our assumption:

1) Bilateral anacusis and extinct vestibular function developed within three months indicating a systemic disorder rather than local pathology. The initial high values of toxoplasmosis antibody titres and their fluctuations (Fig. 4) are indicative of an actual infection; we have been unable to demonstrate any other systemic disorder than toxoplasmosis.
2) Following treatment with sulphadiazine and pyramethamine which has proved to be valuable in CNS-toxoplasmosis (Townsend *et al.*, 1975; Ruskin and Remington, 1976) the patient regained some hearing after two months of anacusis on the left ear and five on the right. Also the vestibular function returned to normal following treatment.

Previous reports have described cochleo-vestibular symptoms in connection with toxoplasmatic meningo-encephalitis (Grimaud *et al.*, 1961; Partsch and Rieder, 1962) which we had no reason to suspect in our patient. Wilke (1961) who reported on three patients with *T. gondii* and Menière's syndrome suggested that symptoms might be attributed to invasion of endothelium which by causing endothelial hypertrophy, diminishing of capillary lumina followed by vasculitis, perivasculitis and thrombosis would result in reduced blood flow in the inner ear. This might severely reduce the oxygen tension causing partial or total damage of sensory cells. Why just the cochleae of this apparently healthy young woman should become a target is uncertain. To quote Feldmann (1981) it is likely that SSHL in one ear is related to the state that this ear *and* the other ear were in before the acute loss. Our patient had for some time suffered from a disease in the left ear, presumably otosclerosis; the opposite apparently healthy ear which was attacked first by SSHL may also have been in a precarious state so that some incident, in this case toxoplasmosis, could trigger off the loss (Feldmann, 1981).

Conclusion

Toxoplasmosis may presumably cause cochleo-vestibular disease. Since effective treatment seems to be available it is important to establish the diagnosis. In sensorineural hearing loss of unknown origin—and particularly if bilateral—we therefore recommend examination for acute toxoplasmosis in order that chemotherapy can be instituted.

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Address for correspondence:
Morten Katholm,
M. D. Nordre Strandvej 50,
DK-3000 Helsingør,
Denmark.

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