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Hearing loss in sarcoidosis

by

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Introduction

Sarcoidosis is a systemic granulomatous disease of unknown aetiology. Any tissue may be affected but the organs most commonly involved include lymph nodes, lung, liver, spleen, bone, skin and eyes. Males in their thirties are most often affected. The course of the disease is usually chronic, with minimal constitutional upset, but occasionally acute phases with fever, malaise and weight loss are present.

Involvement of the nervous system, previously thought to be rare, now appears to occur in approximately 1–5 per cent of cases (Hook, 1954), or higher if cases with abnormalities of the cerebro-spinal fluid are included. Neurological involvement appears to carry a poorer prognosis (Hook, 1954). The most common neurological finding is a VIIth nerve palsy which may be bilateral. The VIIIth nerve is the fourth most commonly affected cranial nerve after the IInd nerve and the IXth and Xth nerves combined (Colover, 1948). Symptoms of VIIIth nerve involvement include hypoacusis, tinnitus, vertigo and vague unsteadiness. It may, therefore, mimic many other otological disorders. Disturbances of balance in sarcoidosis may also be due to involvement of the cerebellum.

Case 1

A 21-year-old male, British-born West Indian miner presented to the Eye Casualty Department with a five-week history of painful red eyes and diminished vision. He also complained of general lethargy and had lost two stones in weight over the same period. Uveitis was diagnosed and treated with topical steroids.

One month later he suddenly developed bilateral deafness which became worse in the course of the following few days. Deafness was preceded by a two-week history of unsteadiness which had gradually improved.

There was no significant past medical history.

On examination he was profoundly deaf bilaterally. The remainder of the ENT examination was normal, as was neurological examination. His spleen was slightly enlarged and he had a small palpable node (3×2.5×1.5 cm.) in his groin. He had signs of posterior uveitis.

General Investigations

Chest X-ray, skull X-ray, serum calcium, ESR and bone marrow biopsy were normal. The C.S.F. contained 24 cells/ml. in the first specimen and 4 cells/ml. in the second. The cells were mainly lymphocytes. C.S.F. protein was mildly raised, at 60 mg. per cent, but the sugar was normal.

Full blood count, urea and electrolytes, thyroid function tests, liver function tests, serology for syphilis, auto-antibodies, CPK and viral studies were all normal.

The Kveim test was negative. C.A.T. and gamma scans were normal. The enlarged inguinal lymph node was biopsied and showed granulomatous changes characteristic of sarcoidosis.

Otological Investigations

A pure tone audiogram showed a sub-total hearing loss in both ears (Fig. 1).

The tympanogram was normal on both sides, with absent stapedial reflexes.

Electronystagmography/caloric tests showed complete loss of labyrinthine function on the left, and diminished response on the right.

Cortical and Brain Stem Evoked Response Audiometry failed to show any response in either ear up to the maximum stimulus of 100 db. to 120 db.

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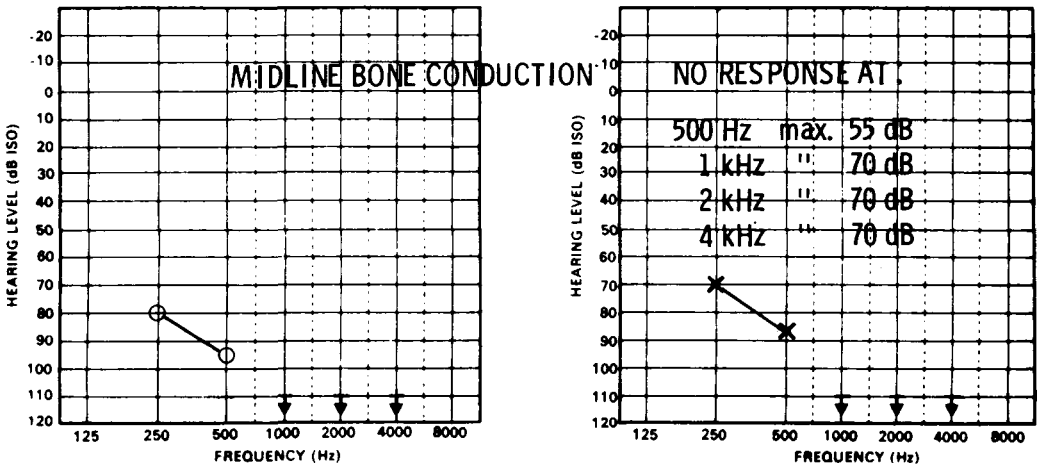


FIG. 1
Showing a pure tone audiogram of Case 1.

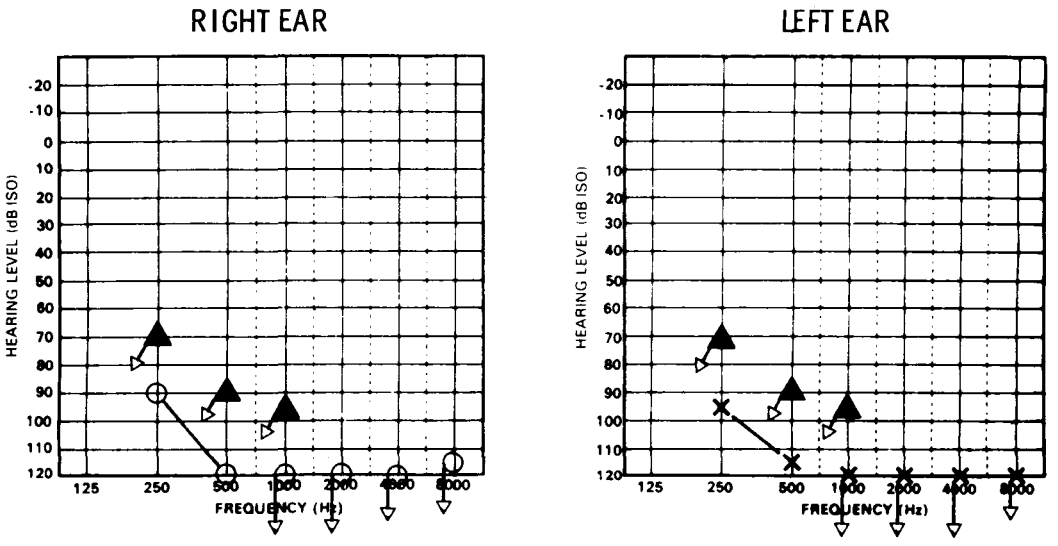


FIG. 2
Showing a pure tone audiogram of Case 2.

He was diagnosed as having sarcoidosis affecting the vestibulo-acoustic system and he was started on Prednisolone 60 mg. per day initially, reducing to a maintenance dose of 10 mg. per day.

After six months of steroid therapy he had noticed no improvement in his hearing. All audiological investigations, including Evoked Response Audiometry were repeated and found to be unchanged, as were vestibular function tests. At this time extra-tympanic electrocochleography was performed. The cochlear microphonics were recorded to be down to a level of 70 db. but no action potential was recorded up to 100 db.

Case 2

A 16-year-old British male presented to the Eye Casualty Department with a two-week history of having a painful red left eye. Over the

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same period he noticed a profound deterioration in his hearing, first in the left ear and then in the right. He also complained of feelings of unsteadiness and of hearing noises in both ears.

One month previous to this he had a similar episode of painful red eyes, which responded to eye drops prescribed by his General Practitioner.

There was no significant past medical history.

On examination he appeared profoundly deaf bilaterally and had a third degree spontaneous nystagmus to the right. The remainder of the ENT examination was normal, as was the neurological examination. He had a mild generalized lymphadenopathy and an acute iritis affecting the left eye.

General Investigations

His full blood count revealed a mild hypochromic microcytic anaemia (Haemoglobin 11.2 gm./100 ml.) and his ESR was slightly raised at 20 mm./hr., Westergren.

The C.S.F. contained 38 red blood cells/microlitre and 27 lymphocytes/microlitre. C.S.F. protein was raised to 140 mg. but the sugar was normal.

Chest X-ray, skull X-ray, urea and electrolytes, serum glucose, toxoplasma and viral serology were all normal. Serology for syphilis was negative and liver function tests were normal, other than a slightly raised Alkaline Phosphatase.

A small palpable lymph node from the posterior triangle on the right side of his neck was biopsied and showed granulomatous changes characteristic of sarcoidosis.

Otological Investigations

A pure tone audiogram showed a sub-total hearing loss in both ears (Fig. 2). Delayed speech feedback showed no evidence of non-organic hearing loss.

The tympanogram was normal on both sides, with absent stapedial reflexes.

Electronystagmography/caloric tests showed complete loss of labyrinthine function in both ears.

Cortical and Brain Stem Evoked Response Audiometry failed to show any response in either ear up to a maximum stimulus of 120 db.

Extratympanic electrocochleography showed a small cochlear microphonic at 100 db. but no action potential was recorded up to 100 db.

He was diagnosed as having sarcoidosis affecting the vestibulo-acoustic system and he was started on Prednisolibe 60 mg. per day reducing to 40 mg. per day after one week. When reviewed after one month on steroid therapy there was no lymphadenopathy and his ESR had returned to normal but there was no improvement in his hearing.

Discussion

Involvement of the acoustic system by sarcoidosis is uncommon. These are the first reported cases of bilateral total deafness of sudden onset in previously undiagnosed cases of sarcoidosis. The diagnosis is relatively easy when the features of systemic sarcoidosis are also present. If not, the cause of the hearing impairment may remain undiagnosed unless the presence of certain signs makes one consider sarcoid. These include uveitis, parotid swelling, VIIth nerve palsy (especially if bilateral) and lymphadenopathy, including hilar adenopathy. The diagnosis may then be confirmed or refuted by the Kveim test, scalene fat pad biopsy (Daniels, 1949), or excision biopsy of any palpable node.

Thirty-five cases, including our own, of deafness due to sarcoidosis have now been described in the English literature since it was first mentioned by Colover (1948). In only 26 cases has the diagnosis been confirmed by histology.

In the cases reviewed the mean age at presentation was 35 years, with no sex predominance.

The high incidence of associated uveitis noted by Gristwood (1958) was confirmed (72 per cent of patients) although the frequency was slightly lower than that reported by Kane (1976).

Sarcoidosis affecting the acoustic system without other signs of neurological involvement was previously thought to be uncommon (Tharp and Pfeiffer, 1969), but in 14 cases (40 per cent) reviewed, there was no evidence of any other neurological involvement. Facial nerve palsy was the most common associated neurological abnormality, occurring in 43 per cent of cases. In 17 per cent the palsy was bilateral.

In 55 per cent of cases hilar lymphadenopathy was noted on the chest X-ray, thus emphasizing the importance of this routine investigation.

Nine cases had parotid swellings, eight of which also had uveitis and could therefore be classified as uveo-parotid fever.

Sarcoidosis is not generally recognized as a cause of sudden deafness (Hybels and Rice, 1976). Fluctuating hearing loss has been described by Engberg and Jepsen (1962) and by Jahrsdoerfer *et al.* (1981). Impairment of hearing may be slight or severe. It may affect high or low

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frequencies and is usually bilateral, although one side is generally more severely affected. Caloric responses are usually hypo-active or absent.

Owing to the lack of fully investigated cases and the unavailability of temporal bones for histology, it is impossible to be sure at what site sarcoidosis affects the vestibulo-acoustic system but it appears from the cases reviewed that it may involve the system at any level.

Out of the 35 cases, 25 have insufficient data concerning the hearing loss. Ten are described as suffering from 'deafness', 11 from 'nerve' or 'sensory-neural deafness', one from 'mixed deafness', two from 'conductive deafness', and one from 'toxic or degenerative labyrinthitis'.

Engberg and Jepsen (1962) suggested that the labyrinth was the site at which sarcoid had its effect since recruitment was present in two of their cases. Cases described by Gristwood (1958) and Kane (1976) were also suggestive of labyrinthine affection.

Owing to the severity of the deafness in our own cases we were unable to test for recruitment. However, extra-tympanic electrocochleography showed a well-recorded cochlear microphonic suggesting normal hair cell function in Case 1. This, in association with an absent action potential, would indicate a retrocochlear lesion. Although only a small cochlear microphonic was recorded in Case 2, this indicates at least some hair cell function. This combined with the absent action potential suggests, as in Case 1, a retrocochlear lesion.

Hooper and Holden (1970) reported two cases. One had definite labyrinthine signs and the other had more central signs with associated brain-stem symptoms. The former recovered completely but the latter was left with residual hearing loss. This conforms with Hybels and Rice's (1976) observation that the more central the lesion, the poorer the prognosis.

The underlying pathology is unknown because of the shortage of histopathological specimens available for examination. Several possibilities have been suggested. Gristwood (1958) put forward three alternatives:

1. Toxaemia affecting the organ of Corti or ganglia.

2. Mechanical pressure on the VIIIth nerve by sarcoid in the basal meninges.

3. Invasion of neural tissue by sarcoid granuloma in

(a) VIIIth Nerve

(b) Brain-stem.

Hybels and Rice (1976) felt that granulomatous meningitis involving the posterior fossa was the most common mechanism resulting in acoustic dysfunction, but also suggested that a vasculitis in the vasa nervorum of the VIIIth nerve may be responsible for the otologic symptoms. This would explain the similarity with Cogan's syndrome, which is thought to be due to periarteritis (Tharp and Pfeiffer, 1969).

Recovery of hearing may range from little to complete improvement (Hybels and Rice, 1976). Caloric responses tend to remain hypo-active despite an absence of vestibular symptoms.

The only form of treatment available at present is steroids. Although they are of proven value in sarcoidosis it remains undecided whether they are beneficial with regard to hearing. Interpretation of the response to steroids is made difficult by the natural tendency to undergo spontaneous remissions, e.g. the improvement of hearing in Engberg and Jepsen's (1962) first case was thought to commence before steroids were prescribed. The deafness remained unchanged in our first patient, despite six months of steroid therapy, and there has been no improvement in the hearing of our second patient to date.

Nevertheless, since the possibility remains that steroids may improve hearing in cases of sarcoidosis, they should be prescribed and the results of such treatment will determine future therapy.

Conclusion

Sarcoidosis may cause bilateral sub-total deafness of sudden onset. Evoked Response Audiometry suggested a retrocochlear lesion with normal hair cell function. Thirty-three cases of sarcoid causing acoustic impairment are reviewed. Steroids remain the only form of treatment available at present but their effectiveness is not assured.

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