Deafness in sarcoidosis

By Kevin Kane (Manchester)

Sarcoidosis is a granulomatous disease of unknown aetiology which most commonly involves the lymph nodes, lung, liver, spleen, skin and eyes. Occasionally it involves the parotid gland and facial nerve, nasal cavity, larynx and rarely the organ of hearing.

The disorder usually runs a chronic course with little constitutional disturbance. Adult males in the third decade are more commonly involved and children very rarely.

The pathognomic feature of the disease is the presence of non-caseating tubercles on histological examination. The lesion is most often found in lymphoreticular tissue and is characterized by a collection of elongated epithelioid cells surrounding central uniform polyhedral cells. Giant cells of both the Langhans' and Foreign Body type are common but there is no central caseation as in the granuloma of tuberculosis.

The nervous system, however, is rarely involved and is affected in approximately five per cent of cases (Hook, 1954). This rises to fifty per cent if uveo-parotid fever is present (Colover, 1948). Any of the cranial nerves may be involved but the most common is the facial nerve. The stato-acoustic nerve is rarely involved and a search of the literature shows that twenty-seven cases have been reported. A further case is described in this paper.

Case report

Mrs. J.W. aged 44 years was admitted under the care of the Neurology Department in April 1974 with a suspected diagnosis of sarcoidosis.

Nine weeks prior to admission she had developed anorexia, general malaise, weight loss, nausea and vomiting.

This was followed by dizziness which she described as 'a swimminess and unsteadiness in walking' and bilateral deafness associated with buzzing tinnitus. She then noticed that her face had become paralysed and her deafness had increased. There was also some mistiness of vision in the right eye.

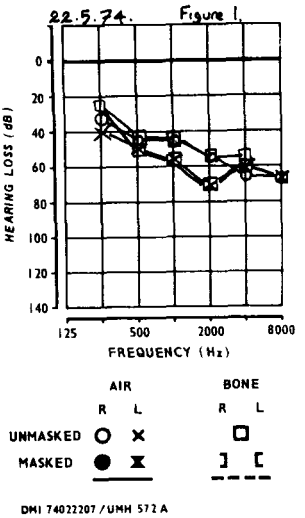
On examination she was afebrile. There was some irregularity of the right pupil with circumcorneal injection which was typical of iridocyclitis. A bilateral lower motor neurone facial nerve palsy was present and her gait was unsteady with a positive Rhomberg test.

There were no other otolaryngological findings apart from the facial nerve

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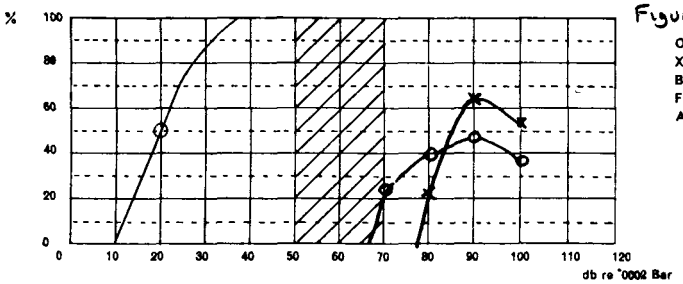
palsies and deafness. There was no demonstrable nystagmus and tuning fork tests showed a central Weber with bilateral positive Rinne tests.

Further investigations were carried out to substantiate the diagnosis of sarcoidosis. These included a full blood examination and ESR which were normal; a chest X-ray which showed glandular enlargement at the right hilum; lumbar puncture revealed a normal CSF pressure and there was a slight increase in the number of lymphocytes present. The sugar was slightly lowered and the CSF protein normal.



A blind deltoid muscle biopsy showed sarcoid tubercles on histological examination thus confirming the diagnosis.

A pure tone audiogram showed a moderate bilateral sensori-neural deafness, more marked on the left side. This correlated with the speech audiogram and the humped curve suggested that recruitment was occurring (Fig. 2).

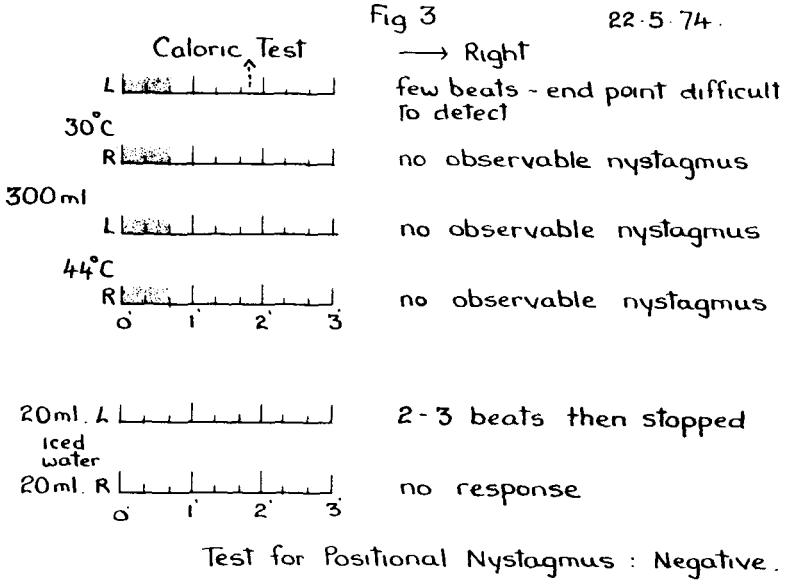


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Impedance audiometry showed excellent stapedial reflexes on the right with 95 decibels and excellent stapedial reflexes on the left at 80 decibels. This again was indicative that recruitment was occurring. There was no tone decay nor stapedial reflex decay.

The SISI was fifteen per cent on the left and forty per cent on the right.

Caloric tests showed an absent response on the right and only slight activity on the left (Fig. 3).



Thus despite a negative SISI, the tests showed that recruitment was present suggesting that the site of the lesion was in the cochlea.

She was placed on Prednisolone 60 mgm. daily initially and her facial nerve palsies and hearing rapidly improved (Fig. 7).

In the ensuing weeks, she showed signs of toxicity by developing Cushingoid facies and thus the dosage was reduced. This was finally stopped after five months.

She was reviewed three months later in July 1974 when the hearing in the right ear had greatly improved subjectively, although she was still very deaf in the left ear. A hissing tinnitus was also troublesome in the left ear.

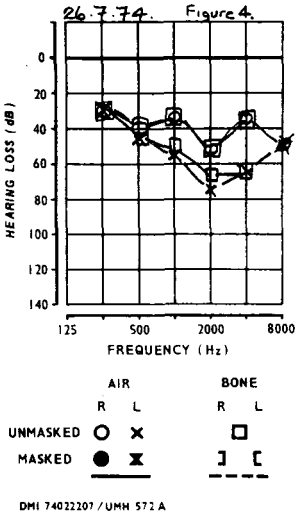
A pure tone audiogram showed some improvement on the right (Fig. 4), but there was no improvement in the speech audiogram (Fig. 5). In November 1974 a pure tone audiogram showed a 30–40 decibel loss on the right side and also some improvement on the left (Fig. 6).

Specialized audiometric testing showed no tone decay and good stapedial reflex with no decay.

Caloric tests with iced water stimulation showed no response.

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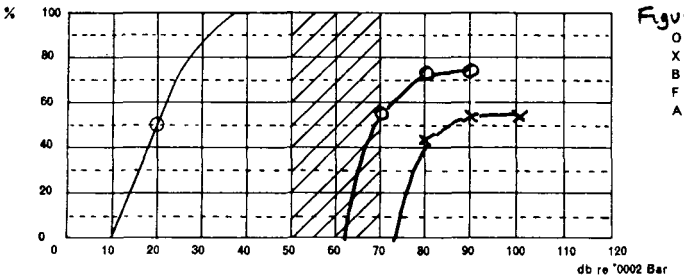
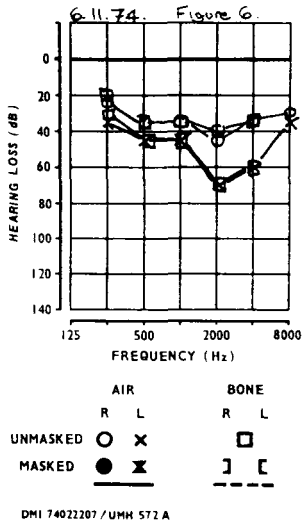


Figure 5. 26.7.74
O – Right
X – Left
B -- Binaural
F -- Free field
A -- Aided



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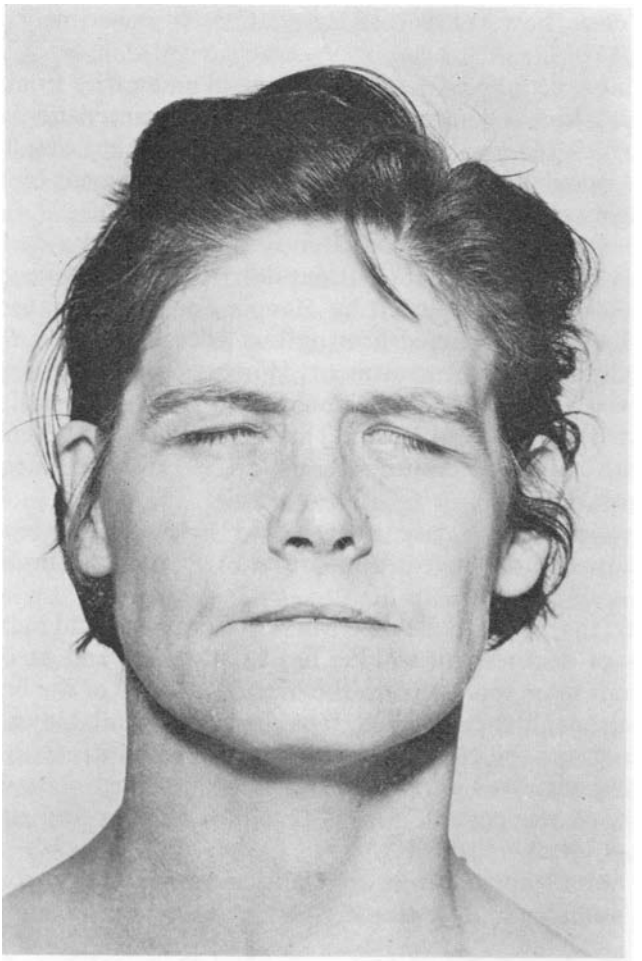


FIG. 7.
The patient showing minimal residual facial nerve palsies four weeks after treatment with Prednisolone.

Discussion

Very little is known about the mechanism of deafness in sarcoidosis. This has resulted both from the lack of cases fully investigated audiometrically and from the lack of pathological evidence.

Gristwood (1958) suggested four ways in which the deafness might result in sarcoidosis: by toxaemia affecting the organ of Corti or cells of the spiral ganglion; by mechanical pressure on the auditory nerve from deposits in the meninges; by direct invasion of the nerve itself by disease; or by infiltration of the brain stem by perivascular collections of sarcoid in the central auditory pathways.

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What is clear, however, is that the deafness is sensori-neural or mixed in type.

The deafness may be unilateral or bilateral and varies from a minimal to a profound loss. There appears to be no characteristic audiometric pattern in which the low or the high frequencies are maximally affected. It is also not possible to fix a consistent site of involvement by the disease process using more modern audiological techniques.

The patient under review consistently demonstrated a cochlear deafness and this was similar to the patient described by Gristwood (1958).

In the two patients reported by Hooper and Holden (1970) the first case showed a minimal mixed hearing loss affecting mainly the low frequencies with absence of recruitment. However, electronystagmographic studies showed a positional nystagmus which was readily fatiguable and thus suggested a peripheral lesion. The second case had a sensori-neural deafness with absent recruitment and an electronystagmograph that suggested a central lesion.

Similarly, in the four cases reported by Engberg and Jepson (1962) two cases showed complete recruitment and two cases incomplete recruitment.

Jefferson (1957) reported a case where the patient could induce vertigo and attacks of deafness by sudden head movement and at operation a granulomatous mass was found in the fourth ventricle of the brain.

Lastly, although there is no autopsy material available on the stato-acoustic nerve nor the temporal bone, patients with deafness or vertigo have been demonstrated at operation or at post mortem to have adhesive arachnoiditis of the posterior fossa (Erichson, Odom and Stern, 1942; Colover, 1948; Walker, 1961).

Thus it would appear from available evidence that sarcoidosis can involve the auditory and vestibular system at any level along their nerve axis.

In the twenty-eight patients of the series who have sarcoid involvement of the stato-acoustic nerve, eighty per cent also show eye changes and forty per cent have facial nerve palsies. Twenty per cent of cases were seen to have parotid swelling (contrast with five per cent overall).

It is still not clear if sarcoidosis can produce a conductive deafness. However, some of the cases described, including that of Hooper and Holden (1970) had a mixed deafness.

Walsh (1939) describes a fourteen year old boy with sarcoidosis who developed a left sided conductive deafness. Examination of the ear showed a bulging tympanic membrane and this was thought to be due to the presence of sarcoid tissue within the middle ear. However, this was not histologically confirmed.

Colover (1948) mentioned a patient where a conductive deafness was found on otological examination but no further details were given.

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Treatment

There is no doubting the efficacy of steroids in the treatment of sarcoidosis and in cases where the auditory mechanism is involved, hearing loss may recover to a degree. In the cases published most patients continued to have a high tone loss and calorics remained hypoactive despite the absence of any vestibular symptoms. This patient continues to have a moderate hearing loss and absent calorics after twelve months despite considerable improvement.

Prednisolone 60 mgm. daily was used initially in this case. The dose was reduced over the ensuing weeks and finally stopped five months later. In the case reported by Tharp and Pfeiffer (1969) Prednisolone 30 mgm. daily was used for two weeks and then reduced over the following three months giving good lasting results.

Summary

A case of sarcoidosis and sensori-neural deafness in which the lesion was thought to be cochlear in position is presented and the audiometric findings are discussed in detail. A review of the reported cases shows that sarcoid lesions may involve the hearing mechanism at any point along its neural axis.

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