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Clinical Records

Mesotympanic sarcoidosis

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Abstract
Sarcoidosis often presents with signs and symptoms within the head and neck. In this region, it most commonly affects the nose, larynx, parotid and cervical lymph nodes. Within the ear, it usually causes sensorineural hearing loss of varying severity. We report a patient in whom sarcoidosis involved the middle ear and presented with a conductive hearing loss. Granulomata were found in the middle ear during an exploratory tympanotomy. The granulomatous process had caused necrosis of the long process of incus and was also encasing the chorda tympani nerve. Histological examination confirmed the diagnosis.

Key words: Sarcoidosis; Middle Ear; Hearing Loss

Introduction

Sarcoidosis is a multisystem granulomatous disorder of an unknown aetiology that can affect any organ in the body (Table I). It presents most commonly as a palpable lymphadenopathy or hilar lymphadenopathy detected as an incidental finding on a chest X-ray.

The diagnosis of sarcoidosis is made on clinical grounds and is strengthened by an abnormal chest X-ray that shows hilar lymphadenopathy or pulmonary mottling. Raised serum angiotensin converting enzyme levels (ACE) are detected in most and 15 per cent of patients have abnormal liver function.[1] The Kveim and Mantoux tests were used but are no longer available because of the potential for prion contamination and HIV infection. Tissue diagnosis is now the gold standard and biopsy of minor salivary glands can achieve this in 58 per cent of cases.[2] Obviously, the definitive diagnosis can also be made by direct biopsy of the involved lymph nodes when this is practical. In these nodes, non-caseating granulomata composed of multiple epitheloid cells, Langerhan type giant cells with intra-cytoplasmic inclusions of Schumann and asteroid bodies are seen.

TABLE I

ORGAN/SYSTEM INVOLVEMENT OF SARCOID

Site	(%)
Lung (hilar lymphadenopathy)	38
Eyes (iridocyclitis)	26
Lymph nodes; lymphadenopathy	22
Skin (erythema nodosum)	18
Nervous system (basal meningitis and cranial neuropathy)	5

From an historical and otorhinolaryngological perspective, Boeck (1905) was the first to describe sarcoidosis of the mucous membrane of the upper respiratory tract. The otorhinolaryngological manifestations of sarcoidosis are summarized in Table II.[1–3]

Case report

A 55-year-old Caucasian man was referred to the ENT department with a right-sided hearing loss and tinnitus of approximately one-year's duration. There were no

TABLE II

SUMMARY OF OTORHINOLARYNGOLOGICAL MANIFESTATIONS OF SARCOIDOSIS

Region	Involvement/manifestation/symptoms	Sign/finding
Nose	Rhinitis, muco-purulent discharge, epistaxsis	Nodule/granuloma/polypoidal mass, destruction of paranasal bones
Larynx	Voice change, progressive stridor	Generalized thickening of laryngeal mucosa, nodule/granuloma
Salivary glands	Facial nerve palsy (Heerfordt syndrome), recurrent sialadenitis	Enlargement of involved gland, mass in gland
Cranial nerves	Facial nerve	Facial palsy
Ear	Cochlea nerve and pathways of hearing	Fluctuating sensorineural hearing loss
	Vestibular nerve and pathways	Vertigo

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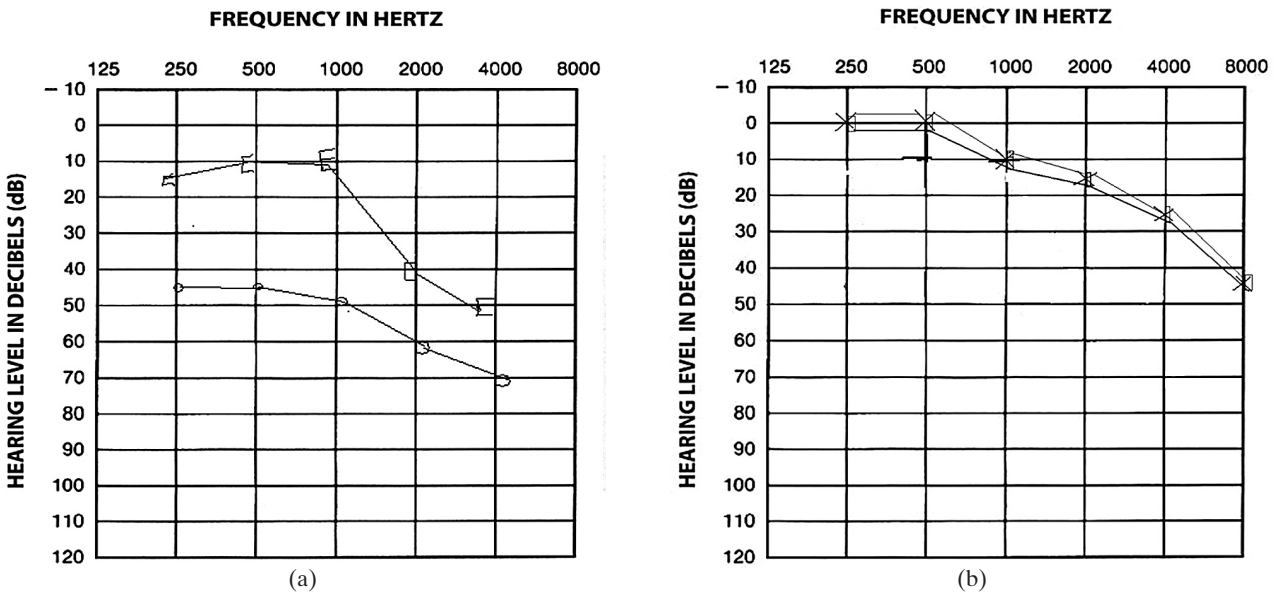

FIG. 1

Pre-operative audiograms of (a) right ear and (b) left ear.

vestibular symptoms and none related to his nose or throat. Fifteen years previously he had developed erythema nodosum and been treated with steroids for a period of time. Otherwise, he had a completely unremarkable history.

On examination of his ears, nose and throat there were no obvious abnormalities apart from a right-sided conductive hearing loss (Figure 1). A presumptive diagnosis of otoscelorosis was made and he agreed to surgical treatment. At tympanotomy, it was evident that the long process of incus had been eroded by a granulomatous process. Another granuloma was found encasing the chorda tympani nerve which was carefully removed and sent for histological examination. Reconstruction and restoration of hearing was achieved by a vertical transposition of his incus (Figure 2). He made a satisfactory recovery. Histological examination of the operative specimens showed a patchy lymphocytic infiltrate with poorly formed granuloma consistent with the diagnosis of sarcoidosis (Figure 3). A chest X-ray confirmed the diagnosis as he had a bilateral hilar adenopathy and upper lobe fibrosis. Serum ACE levels were slightly elevated at 27 IU (normal: 8.3–21.4 IU). A Mantoux test was performed and was negative. His serum calcium levels were within the normal range at 2.4 mmol/l. It was decided to treat him with a further course of steroids. He has now been under review for the last 12 years, is no longer being treated with steroids and has acquired further sensorineural hearing loss that cannot be accounted for purely by age (Figure 4).

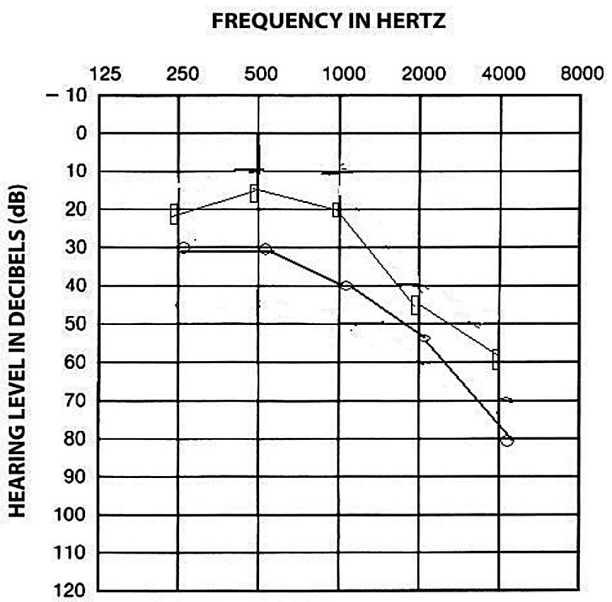

FIG. 2

Post-operative audiogram of the right ear.

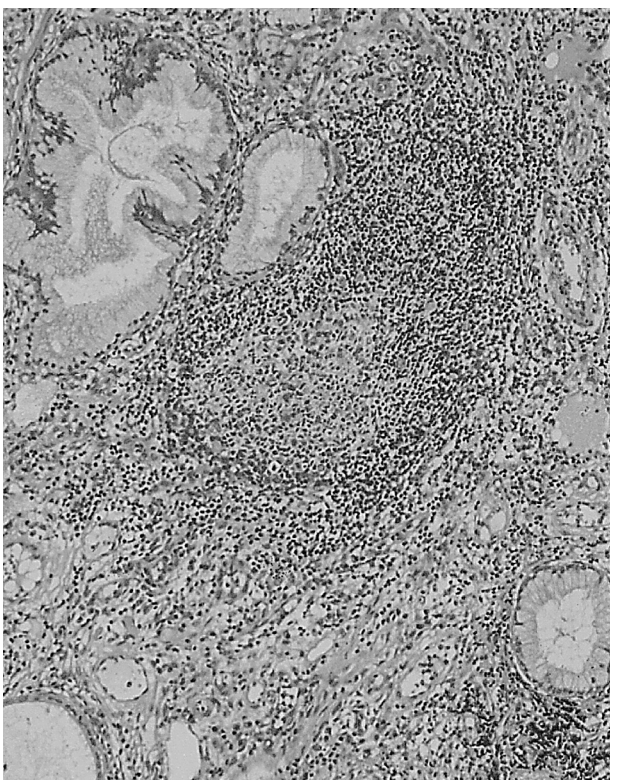

FIG. 3

Seromucinous glands with a dense chronic inflammatory infiltrate including lymphoid follicles with germinal centre (H&E stain, ×200).

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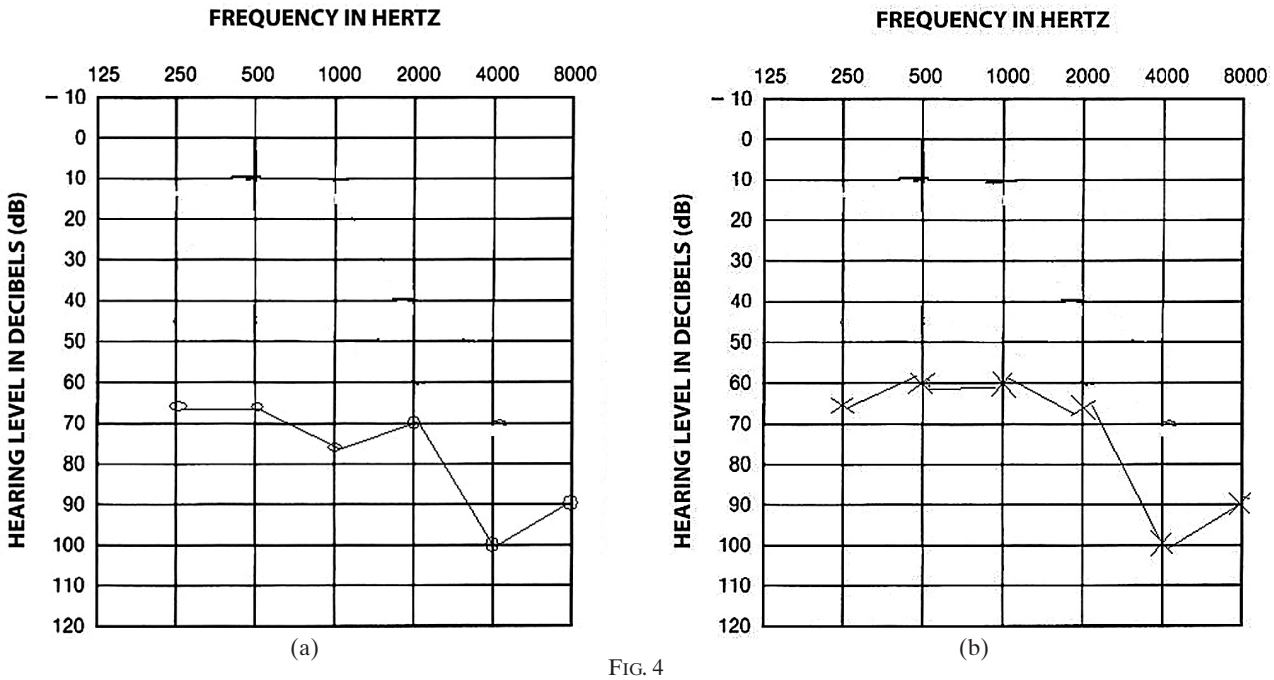

FIG. 4
Last audiograms of (a) right ear and (b) left ear.

Discussion

Sarcoidosis is a chronic granulomatous disorder of unknown aetiology. Approximately 9 per cent of patients have manifestations of this disease in the head and neck region,[4] with the nose being the most common site affected. The respiratory tract is commonly involved in sarcoidosis and, of course, the middle ear is a direct extension of it. It may well affect this site more often but go undetected.

The usual otological manifestations of sarcoidosis are infiltration of the skin and cartilage of the pinna, sensorineural hearing loss and vertigo.[5] Skin involvement is characterized by maculo-papular plaques and lupus pernio which is a chronic violaceous skin lesion that can be disfiguring. Neurotological symptoms and signs include fluctuating sensorineural hearing loss, paroxysmal vertigo and tinnitus. Facial palsy is usually associated with parotid infiltration and uveitis. The hypotheses proposed by Gristwood[6] in 1958 to explain the otological symptoms were: (1) toxaemia affecting organ of Corti or ganglion, (2) mechanical pressure on the VIIIth nerve in basal meningies, (3) invasion of neural tissue by granuloma. Hybels and Rice[7] suggested that granulomatous meningitis in the posterior fossa was one of the causes of sensorineural deafness in sarcoidosis. They also suggested that a vasculitis involving the vasanervorum of the VIIIth nerve could be another reason for sensorineural loss. Brihaye and Halama[8] supported the vasculitis theory, while Moine *et al.*[9] proposed infiltration of arachnoid vessels as yet another possible cause of hearing loss. Histological evidence of granulomatous changes in the posterior fossa was documented by Babin *et al.*[10] who reported perivascular infiltration around the VIIIth nerve within the internal auditory canal.

Conductive deafness has been reported before but in these cases the cause was secretory otitis media caused by nasal involvement, eustachian tube dysfunction or nasopharyngeal sarcoidosis.

Only two cases have been reported in literature where sarcoid has been found in the middle ear. The first was a patient with sarcoid who had experienced unilateral otalgia, aural fullness, tinnitus and had a mild conductive deafness. At tympanotomy, a soft tissue mass was found in the tympanum extending into aditus towards the sinus tympani. Biopsy of this tissue confirmed sarcoidosis. The same patient later developed nasal disease and was treated with steroids. This failed to improve her hearing or tinnitus.[11] The other case had intermittent aural discharge and a granuloma was found during mastoidectomy which was subsequently confirmed as sarcoid.[12] Although rare, sarcoid should be considered as a potential cause of conductive hearing loss and our experience emphasizes the importance of subjecting abnormal tissue found during middle-ear exploration to histological examination. Whether systemic steroids are effective in the management of middle-ear infiltration has to be debatable.

- **A rare case of sarcoidosis involving the middle ear is reported in this paper**
- **Presentation was with conductive deafness, with middle-ear biopsy being consistent with sarcoidosis**
- **The otological manifestations of sarcoidosis are discussed**

References

1 Page JR, Seth SM. Sarcoidosis of the upper respiratory tract. *J Laryngol Otol* 1969;**83**:1005–11
2 Lazarus AA. Sarcoidosis. *Otolaryngol Clin North Am* 1982;**15**:621–31
3 Miglets AW, Vial JH, Kataria VP. Sarcoidosis of the head and neck. *Laryngoscope* 1977;**87**:2038–48
4 Mccaffery TV, Mcdonald TJ. Sarcoidosis of the nose and paranasal sinuses. *Laryngoscope* 1983;**93**:1281–4
5 Postma D, Fry TL, Malenbaum BT. The nose, minor salivary glands and sarcoidosis. *Arch Otolaryngol* 1984;**110**:28–30
6 Majumdar M, Crowther J. Hearing loss in sarcoidosis. *J Laryngol Otol* 1983;**97**:635–9
7 Hybels RL, Rice DH. Neuro-otological manifestation of sarcoidosis. *Laryngoscope* 1976;**86**:1873–8

https://doi.org/10.1258/0022215054797907 Published online by Cambridge University Press

8 Brihaye P, Halama AR. Fluctuating hearing loss in sarcoidosis. *Acta Otorhinolaryngol Belg* 1993;**47**:23–6
9 Moine A, Frachet B, Van Den Abbeel T, Tison P, Battesti JP. Deafness and sarcoidosis. *Ann Otolaryngol Chir Cervicofac* 1990;**107**:469–73
10 Babin RW, Liu C, Aschenbrener C. Histopathology of neurosensory deafness in sarcoidosis. *Ann Otol Rhinol Laryngol* 1984;**93**:389–93
11 Tyndel FJ, Davidson GS, Birman H, Modzelewski ZA, Acker JJ. Sarcoidosis of the middle ear. *Chest* 1994;**105**:1582–3
12 Verse T, Pirsig W, Heymer B. A case of sarcoidosis of the mastoid. *Laryngorhinootologie* 1997;**76**:312–14

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Mr A S Banerjee takes responsibility for the integrity of the content of the paper.
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