

A Rare Cause of Sudden Hearing Loss in Chinese: Cochlear Involvement of Sarcoidosis

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Citation

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Abstract

Sudden sensorineural hearing loss is a common problem presented to otorhinolaryngologists. Sarcoidosis is a rare condition among the Chinese population. Sudden sensorineural hearing loss as a manifestation of sarcoidosis in Chinese patients has not been reported in the literature. This is the first reported case of sarcoidosis with cochlear involvement on a 42-year-old Hong Kong Chinese man who presented with sudden unilateral sensorineural hearing loss. The diagnosis was made after he presented with hearing loss.

All the work of this article should be attributed to the Division of Otorhinolaryngology, Department of Surgery, Queen Mary Hospital, University of Hong Kong Medical Center

INTRODUCTION

Sudden sensorineural hearing loss is a common otologic problem. It is defined as sensorineural hearing loss of 30 dB or more in three contiguous pure tone frequencies within 72 hours. The reported incidences are 5-20 per 100,000 population per year.¹ Persons of all ages may be affected, and there is no sex predilection.² Partial or complete spontaneous recovery occurs in 30% to 65% of cases.³ Extended evaluation may reveal occult underlying diseases (e.g. acoustic neuromas, and infectious diseases) in 10% of cases.²

Sarcoidosis is a disease of worldwide distribution but is rare among Chinese. In the Hong Kong Chinese population, only 12 confirmed cases of sarcoidosis have been reported.^{4,5,6} None of the cases reported in Hong Kong was characterized by central nervous system involvement, which affects approximately 5% of all cases. In fact, only one report exists that describes sarcoid-related cranial neuropathies in a Chinese patient.⁷

As Hong Kong physicians rarely encounter patients with sarcoid-induced cranial neuropathies, the diagnosis may be missed or delayed in the Hong Kong community. In this report, we describe the clinical features of the first reported case of sarcoid-induced sensorineural hearing loss in a Hong

Kong Chinese patient and discuss salient features of its presentation, diagnosis, and treatment.

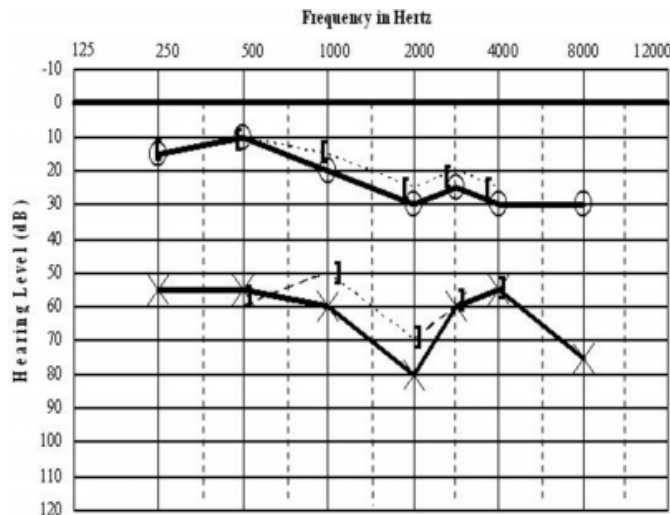
CASE REPORT

A 42-year-old Hong Kong Chinese man was referred for otolaryngological consultation for cervical lymphadenopathy and sudden sensorineural hearing loss of the left ear discovered incidentally on admission to our hospital. The patient presented to the physician with complaints of palpitations but was soon discharged, as thorough work up for cardiovascular pathology did not reveal any abnormality. The patient reported no recent illness, fever, night sweats, and malaise. In addition, he reported no tinnitus, vertigo, or head trauma. His past health was remarkable for remote history of rheumatic fever, an episode of facial palsy of the right-side one month prior to admission that improved with oral steroids (treated by an outside physician), and bilateral uveitis. Current medications included prednisolone and timolol eye drops. The patient is a non-smoker and non-drinker.

Physical examination was notable for one palpable, non-tender, firm, and mobile enlarged lymph node measuring approximately 2 cm x 2 cm in the right supraclavicular region. An audiogram revealed moderate to severe sensorineural hearing loss in the left ear and normal to mild hearing loss in the right ear (Figure 1). The remainder of the otolaryngologic and neurologic examination was normal.

Figure 1

Figure 1: Audiogram at presentation showing left sensorineural hearing loss. X & O symbols represent left & right air conduction thresholds respectively;] & [symbols represent left & right bone conduction thresholds respectively.



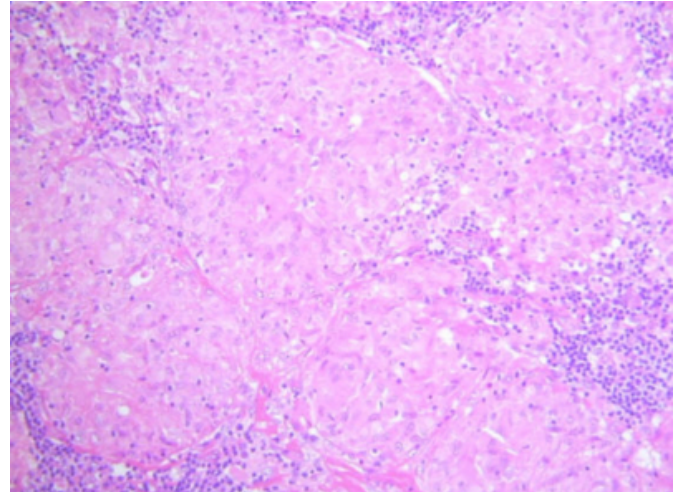
Laboratory investigations showed normal leukocyte count, slight anaemia with hemoglobin at 12.8 g/dL (normal range, 13.0-18.0 g/dL), and lymphocytopenia at $0.50 \times 10^9/L$ (normal range, $1.50-4.00 \times 10^9/L$). His serum chemistry was unremarkable. The erythrocyte sedimentation rate (ESR) was elevated at 50 mm/hr (normal range, <10mm/hr). Both the albumin adjusted serum calcium level and the urinary calcium excretion were normal. The liver enzyme panel was within normal range as were the serum globulin level and serum immunoglobulin patterns. Tests for autoimmune antibodies, including antibodies to neutrophilic cytoplasmic antigens (ANCA), were all negative. Spirometry recorded a reduced 1-second forced expiratory volume of 1.84 L (69% predicted volume) but the forced vital and diffusing capacities were within normal. Mantoux test performed during the hospital admission was negative. Kveim test was not available. Angiotensin converting enzyme level was 63 units (normal range, 14-70).

Bilateral hilar and paratracheal masses were seen on chest X-ray. Computerized tomography of the chest showed extensive mediastinal and hilar lymphadenopathy. X-ray films of the hands and feet did not reveal the presence of lytic lesions. Panendoscopy showed no significant findings, and nasopharyngeal biopsies were negative for malignancy. Ultrasound scan of the neck located two enlarged lymph nodes: one each in the right and left supraclavicular regions. Excision biopsy of the right supraclavicular lymph node and endobronchial biopsy of the left upper lobe revealed florid

epithelioid granulomatous inflammation with small areas of central necrosis and giant cell formation, suggestive of sarcoidosis (Figure 2). No acid-fast bacilli or fungi were noted on either biopsy. Magnetic resonance imaging (MRI) with gadolinium enhancement of the brain was normal.

Figure 2

Figure 2: Biopsy of the enlarged right supraclavicular lymph node showing coalescing non-caseating granulomas composed of large epithelioid histiocytes with rare multinucleated giant cells.



In audiologic investigations, both transient evoked otoacoustic emissions (TEOAEs) and distortion product otoacoustic emissions (DPOAEs) were absent in the affected ear. This represented an impairment of normal outer hair cells function in the affected cochlea. Auditory brainstem response (ABR) tests showed a consistent waveform and the interwave latency measurements were within normal range. The results indicated normal functioning auditory nerves.

Based on the serologic, radiographic, and histopathologic findings consistent with sarcoidosis, the patient was diagnosed with sarcoid-induced hearing loss. Steroid therapy was initiated at two weeks after the onset of hearing loss until the diagnosis of sarcoidosis was substantiated by chemical and histological investigations. Oral prednisolone of 1mg/kg was given for 4 weeks period and then tailed off in one-week time. The patient responded by showing a reduction of ESR to 10mm/hr, and significant reduction in size of hilar lymphadenopathy in chest XR with improvement of lung function parameters. However, pure tone threshold of the affected ear did not show any improvement.

DISCUSSION

Sarcoidosis is rare, and central nervous system involvement

has been virtually unheard of among the Chinese population. Here we report the first case of sarcoid-induced sensorineural hearing loss in a Hong Kong Chinese man. The 12 previously reported cases of sarcoidosis among the Hong Kong Chinese population were almost asymptomatic at presentation while others presented with uveitis, respiratory symptoms, malaise, or diabetes insipidus. In general, pulmonary (88%), lymphatic (25-50%), ocular (15-25%), dermatologic (20%), hepatic and splenic manifestations are common.⁸ Otolaryngologic signs of sarcoidosis are seen in 10% to 15% of patients, most commonly in the form of neck mass, parotid swelling, and facial paralysis.⁹ Facial palsy is the most common neurological disorder afflicting nearly half of patients with neurological involvement.^{10, 11} The optic nerve is the second cranial nerve most affected while cranial nerves IX and X combined are the third most commonly affected.¹¹ Sarcoid-induced hearing loss affects less than 1% of patients and is often sudden, asymmetrical, and fluctuating.^{9, 12} Involvement of cranial nerve VIII is usually associated with other cranial neuropathies. However, a few cases of patients whose sole initial manifestation of sarcoidosis was sudden hearing loss have been reported.¹³

Most studies report a strong association between VIIIth cranial neuropathy and uveitis and/or facial paralysis.^{10, 13, 14} Hybels and Rice,¹¹ found that of 12 patients with sensorineural hearing loss due to sarcoidosis, 11 patients had uveitis and 8 had facial palsy. Consistent with those reports, our patient was being treated for uveitis at the time of presentation and reported a recent history of facial palsy.

The diagnosis of sarcoidosis is made based on compatible clinical, serologic, and radiological findings, histological evidence of noncaseating granulomas, and exclusion of other diseases capable of producing a similar clinical or histological picture. Common laboratory abnormalities seen in sarcoidosis include anaemia, lymphocytopenia, elevated alkaline phosphatase, hypercalcemia, hypercalciuria, hypergammaglobulinemia, and an elevated ESR. The case presented here demonstrated the presence of many of these abnormalities. Because the cells that make up granulomas secrete large amounts of angiotensin converting enzyme (ACE), high ACE enzyme levels are observed in up to 80% - 90% of patients with sarcoidosis. Although not pathognomonic, elevated ACE levels are suggestive of sarcoidosis. Both ESR and ACE levels can be used to monitor the progress of the disease. The Kveim test, which involves intradermal injection of heat-treated sarcoid tissue to test for

host granulomatous reaction, is fairly sensitive (approximately 80%) and highly specific (>95%), but the antigen is not available in Hong Kong due to the risk of disease transmission.

Radiographic findings such as bilateral hilar and mediastinal lymphadenopathy, pulmonary parenchymal infiltrates, or lytic lesions of the metacarpals and phalanges can support the diagnosis of sarcoidosis. Although gadolinium-enhanced MRI findings did not identify any lesions in the VIIIth cranial nerve in our patient, MRI findings of nerve lesions in patients with sarcoid-induced hearing loss and facial palsy have been reported.^{12, 15}

Although not pathognomonic, sarcoidosis is most strongly supported by histopathologic findings of noncaseating granulomas in involved tissues. If available and accessible, involved lymph nodes may be the simplest to biopsy. If lymph nodes are unavailable, mediastinoscopic or bronchoscopic biopsies offer the highest rates of specificity at 60% - 80% and 98% - 100%, respectively.⁹ Nonetheless, tuberculosis, lymphoma, fungal infections, vasculitides, rheumatic fever, and berylliosis can give a similar histopathologic picture and must be excluded before the diagnosis of sarcoidosis can be confirmed. Moreover, even in a patient with known sarcoidosis, it is important to rule out other possible causes of sensorineural hearing loss, such as infection, neoplasm, ototoxicity, and trauma, before a diagnosis of sarcoid-induced hearing loss is made.

Multiple theories have been proposed to explain the mechanism of hearing loss in sarcoidosis. These theories have included toxemia of the Organ of Corti, mechanical compression and/or direct infiltration of sarcoid deposits in the VIIIth cranial nerve and/or the brainstem, and inflammatory vasospasm or vasculitis of vessels serving the brainstem or cranial nerves.^{11, 12, 14} However, the exact pathophysiology of sarcoid-induced hearing loss remains unknown. In our patient, normal ABR and MRI findings supported an intact auditory nerve. TEOAEs and DPOAEs reflect the normal function of outer hair cells in a healthy cochlea, absence of which in our patient indicated cochlear impairment. As the onset of the hearing loss was abrupt and direct infiltration of sarcoid deposit to the cochlea was excluded in the MRI study, we postulated that the mechanism of sensorineural hearing loss in our patient was ischaemia of the cochlear organ secondary to inflammation of the supplying vessels. Whereas pulmonary manifestations of sarcoidosis are often asymptomatic and may not require

treatment, administration of steroids is the mainstay of treatment for those with central nervous system involvement and has been shown to improve central nervous system function.¹³ However, the steroid therapy was not started until the diagnosis of sarcoidosis was substantiated by chemical and histological investigations. There was a two-week delay of steroid treatment after the onset of hearing loss. As the ischaemia of the cochlear organ secondary to inflammation of the supplying vessels was suggested, there may be irreversible ischaemic damage to the inner ear if the treatment was delayed. This can help to explain the poor response to treatment in pure tone threshold of the affected ear despite of the favorable responses in ESR and lung function parameters. We propose that earlier administration of steroid therapy, hopefully before the irreversible ischaemic damage to the inner ear, is necessary for a better outcome.

CONCLUSION

Although sarcoidosis is rare among the Chinese, it must be considered in the differential diagnosis of cranial neuropathies. Sudden sensorineural hearing loss is a well-recognized manifestation of sarcoidosis, and when sudden asymmetric sensorineural hearing loss presents in association with uveitis, facial paralysis, and neck mass, sarcoidosis should be strongly considered. The diagnosis of sarcoidosis and sarcoid-induced hearing loss is most strongly supported by characteristic histopathologic findings of noncaseating granulomas in biopsy specimens, but diseases with similar presentations must be excluded. Treatment involves administration of systemic steroids. ESR, ACE levels, and follow-up audiograms are useful in the monitoring of disease progression.

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