

An Unusual Presentation Of Sarcoidosis With Progressive Hearing Loss

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Citation

J Williamson, V Singh, S Lobo, R Goodfellow. *An Unusual Presentation Of Sarcoidosis With Progressive Hearing Loss*. The Internet Journal of Otorhinolaryngology. 2010 Volume 13 Number 1.

Abstract

ObjectiveWe present a case report of a 52 year old female in whom the first and only presenting symptom of sarcoidosis was hearing loss. Neurosarcoidosis is a rare condition and published data are limited to case reports and small case series. A small number of cases with hearing loss as an initial symptom have been described, however all have also had other neurological disturbances. **Case Report**The patient presented to emergency outpatient clinic with sudden deterioration in hearing but was otherwise asymptomatic. Initial investigations suggested a diagnosis of ossicular fixation, however, her hearing rapidly and progressively deteriorated over the following months. Further investigation revealed raised serum angiotensin converting enzyme and anti-nuclear factor and suggested a diagnosis of sarcoidosis, however, there was no discrete lesion to make a tissue diagnosis. Treatment with high dose oral steroids failed to improve her symptoms, the efficacy of which has been debated in the literature. **Conclusion**The Otolaryngologist should consider an autoimmune cause in any patient with unexplained hearing loss.

INTRODUCTION

Sarcoidosis is a chronic, multi-system disease of unknown aetiology. It is a rare disorder, with a prevalence of between 10-50 per 100000 population.¹

It is characterised by non caseating granulomas which affect the lungs predominantly, but the disease may also affect many other systems including the eyes, skin, lymphoid tissue, bone, solid organs and in approximately 5% of cases, the nervous system.²

Hearing loss as a presenting symptom of neurosarcoidosis in patients with established sarcoidosis has previously been described.³

There have been few reports describing hearing loss as one of the initial presenting symptoms of sarcoidosis in otherwise undiagnosed patients, however these were also associated with other neurological or ENT disturbances.⁵⁻¹²

To date, there have been no cases in which hearing loss alone was the sole and initial presenting symptom in a patient with a de-novo diagnosis of sarcoidosis.

CASE REPORT

Mrs BM, a 52 year old lady presented in March 2008 to the ENT department at Royal Glamorgan Hospital, South

Wales.

She complained of a 6 month history of gradually worsening right sided hearing loss, which became noticeably worse over the course of 3 days. There was no associated tinnitus, vertigo or aural discharge. There were no other neurological or ocular symptoms. On questioning, she recalled a transient sharp pain within the right ear which lasted less than one day, occurring several weeks preceding her appointment which had resolved completely by the time she attended ENT clinic.

She was otherwise well with no systemic symptoms. She had no significant medical history and was on no regular medications. She had no allergies and there was no family history of note.

Full ENT examination revealed normal ear canals and tympanic membranes bilaterally. Examination of the nose and throat was normal. Tuning fork tests revealed Rinnes positive bilaterally and she was unable to localise Webers. Stapedial reflexes were absent bilaterally. Tympanometry was normal bilaterally, however, a Pure Tone Audiogram (PTA) revealed a moderate mixed hearing loss in the right ear up to 4KHz, with a severe dip for higher frequencies. In the left ear, there was a mild sensorineural hearing loss up to 4KHz with a similar dip for higher frequencies.

Fig 1. Pure Tone Audiogram 5/3/8

It was felt the conductive component of the right sided hearing loss was due to ossicular fixation. After discussing treatment options including exploratory tympanotomy, it was decided to provide a digital hearing aid for the right ear.

Upon planned review 3 months later, she had noticed a subjective deterioration in her hearing. A repeat PTA revealed worsening thresholds bilaterally, with severe hearing loss up to 4KHz, with a further dip for higher frequencies in the right ear and borderline moderate to severe hearing loss up to 3KHz with a dip for higher frequencies in the left ear.

Fig 2. Pure Tone Audiogram 11/6/8

To reassess the hearing, a repeat PTA was arranged for 2 weeks.

At this review, she had noticed some benefit using her newly fitted right sided hearing aid. Clinical examination revealed Rinne's negative on the right side and positive on the left, Webers was central. A repeat PTA revealed her right sided hearing loss was marginally worse, with a further deterioration in the left ear indicating profound hearing loss.

Fig 3. Pure Tone Audiogram 2/7/8

An auditory brain stem response test was booked to obtain an objective recording of this progressive bilateral hearing loss. Furthermore, a left sided hearing aid was fitted.

ABR performed in August revealed profound bilateral sensorineural hearing loss. MRI scan of the head and routine blood tests, including immune profile were requested. She was also empirically treated with oral prednisolone at a dose of 40mg once daily for one week, followed by 30mg once daily for 4 days and 20mg once daily for 3 days.

On review in September 2008 her hearing loss had deteriorated still in both ears, with bilateral profound hearing loss in all frequencies. ENT examination remained unchanged. The MRI scan revealed no intracranial abnormality but did however demonstrate diffusely enlarged parotid glands bilaterally. A follow up ultrasound of the parotids demonstrated fatty infiltration with no obvious abnormal area which could be biopsied. A chest X-Ray was normal with no evidence of hilar lymphadenopathy.

A full bank of autoimmune screen results was available which revealed a raised serum angiotensin converting

enzyme at 87 (normal range 20-54) and a weakly positive antinuclear factor. (Assays for antimitochondrial antibody, smooth muscle antibody, extractible nuclear antigen, anti-neutrophil cytoplasmic antibody were negative)

Despite the positive ANF the patient displayed no features of an autoimmune connective tissue disease, questioning also failed to elucidate any other common presenting features of sarcoidosis namely erythema nodosum, sicca symptoms, respiratory symptoms nor an arthropathy. In view, however, of the raised ACE with no other obvious cause, a diagnosis of sarcoidosis was deemed to be the most probable cause of her symptoms. Unfortunately her hearing did not improve with conventional high dose steroids and therefore she was not further immunosuppressed.

She remains under regular ENT review and has also been referred for consideration for a cochlear implant.

DISCUSSION

Hearing loss in Sarcoidosis is extremely rare. James and Sharma¹³ observed that over a 10 year period, of 1080 attendees to a sarcoidosis clinic, only 7% had neurological manifestations and only a single patient had deafness associated with the disease.

The commonest sarcoid related neurological disturbance involves the 7th cranial nerve, followed by the 2nd, 9th and 10th nerves with the 8th nerve least frequently involved.¹⁴

There have been few cases of sarcoidosis involving the 8th cranial nerve reported to date and most have been associated with other neurological, ophthalmological or systemic involvement.⁵⁻¹²

Cases in which sarcoidosis affect purely the audiovestibular system alone are extremely rare.

Souilere described a patient who developed sudden SNHL, dysequilibrium and tinnitus but was otherwise asymptomatic. Initial investigations were inconclusive, however, an air contrast CT scan revealed an inflammatory cerebellopontine angle mass and widening of the internal auditory canal. Further examination of the patient revealed a supraclavicular fullness and a biopsy of this revealed sarcoid granuloma. The patients hearing markedly improved with systemic steroids.⁶

A patient reported by Banerjee presented with unilateral conductive hearing loss and tinnitus. Investigation revealed ossicular erosion by granulomatous inflammation and

mesotympanic sarcoidosis was diagnosed. Hearing was restored with ossicular reconstruction and a course of systemic steroids.¹⁵

There have been several other proposed mechanisms by which sarcoidosis affects the hearing. Hybels suggested sarcoidosis may result in diffuse microgranulomas in the brain substance which may progress to larger confluent granulomas, or localised meningeal granulomatosis.³ This may result in symptoms by release of a 'toxic factor' from inflamed tissue, direct invasion of neuronal tissue or the vasa neuronium or by mass effect.¹⁶

It has been reported that there has historically been very limited post mortem histological material for research and further work is required in this area.¹⁷ Histological examination by Babin of a patient with advanced CNS sarcoidosis revealed extensive perivascular lymphocytic infiltration of the 7th and 8th cranial nerves resulting in axonal degeneration.¹⁸

A case report by Markert et al presented a patient with right sided hearing loss, dizziness, balance disturbance and tinnitus. A brain MRI revealed a right cerebellopontine angle mass. A biopsy of this revealed necrotizing granulomatous inflammation and treatment with corticosteroids resulted in significant improvement in hearing.²¹

In our patient the imaging performed did not provide an anatomical explanation for the cause of her neurological symptoms. Other studies⁹ have reported abnormalities of MRI gadolinium uptake in patients with neurosarcoid, however, in our patient it was felt further imaging or biopsy would add little to the management as steroid therapy was unsuccessful and she was otherwise asymptomatic. There is some evidence that the use of the immunosuppressant drug Methotrexate may act as a steroid sparing agent²², however, we did not consider its use appropriate in this case as there was no response to prednisolone initially. Furthermore, there is presently no compelling evidence supporting its use in isolated neurosarcoidosis and in view of its potential severe toxicity and after discussion with the patient, we opted not to use it in her case.

CONCLUSION

We report a case of progressive hearing loss attributable to sarcoidosis. There have been several documented cases of varying presentations of neurological sarcoidosis. This is the first documented case to date of sarcoidosis solely affecting the audiovestibular nerve with no other neurological or

systemic manifestations. Histological diagnosis in this case was not possible as there was no discrete lesion to biopsy. Treatment with corticosteroids in this case was minimally effective, the role of which has only been investigated on a case by case and series review basis. Further research in the form of larger well controlled clinical trials is required

In dealing with patients with atypical or rapidly progressive hearing loss, the Otolaryngologist should consider to the possibility of sarcoidosis and conduct a full neurological and general examination for evidence of this condition.

BULLET POINT SUMMARY

- Neurosarcoidosis is rare therefore no robust clinical data upon which to base diagnosis and treatment
- Rapidly progressive hearing loss alone, with biochemical evidence of Sarcoidosis, without other clinical signs or symptoms of autoimmune disease has not previously been described
- No identifiable pathological lesion to biopsy for tissue diagnosis
- Treatment with high dose oral steroids not effective in this case

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