Baird Samantha (Orcid ID: 0000-0002-5274-8761)

**Full Title:** Hearing recovery following repair of otic capsule defect secondary to semicircular canal erosion from Langerhans cell histiocytosis

**Short Title:** Otic capsule Langerhans cell histiocytosis

Samantha M. Baird, MBBS (Hons) GDipSurgAnat
Benjamin P. C. Wei, MBBS PhD FRACS
Kevin Nguyen, MBBS BMedSc, GDipSurgAnat FRACS
Robert Briggs, MBBS FRACS

1Department of Otolaryngology, The Royal Victorian Eye and Ear Hospital
2Department of Surgery, Otolaryngology, University of Melbourne

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**Corresponding Author:**
Dr Samantha M. Baird
[dr.sm.baird@gmail.com](mailto:dr.sm.baird@gmail.com)
Address: Department of Otolaryngology
Royal Victorian Eye and Ear Hospital

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Case Report

A 51-year-old male presented with a two-month history of sudden onset right-sided hearing loss, aural fullness and pulsatile tinnitus, and vertigo, autophony and Tullio’s phenomenon. Past medical history included treated hepatitis C infection and he was a current smoker (~15 pack-year history).

Otoscopy was unremarkable and fistula test was negative bilaterally. Tuning fork tests revealed a Weber that lateralised to the left with positive Rinne bilaterally. Complete neurological examination including cerebellar testing, head impulse test and Dix-hallpike test were normal. Audiogram demonstrated right mild-to-moderate sensorineural hearing loss (SNHL) with conductive overlay at 500Hz and 1000Hz, and a word recognition score (WRS) of 60% at 70dB. There was mild left high frequency SNHL with a WRS of 100% at 60dB. Tympanograms were type A bilaterally (Figure 1A).

Computed tomography (CT) revealed an ill-defined lucent mass in the superior right petrous temporal bone (PTB) that extended into the mastoid antrum with thinning of the tegmen mastoideum and bone erosion involving superior and posterior semicircular canals (SCCs) (Figure 2A & 2B). Contrast magnetic resonance imaging (MRI) showed a 1.4x0.6x0.5cm enhancing mass in the superior right PTB with associated superior and posterior SCC erosion. Vestibular function testing was normal bilaterally without decreased
threshold nor increased amplitude on cVEMP testing to suggest third window effect. PET/CT showed no focal FDG uptake in the right otic capsule nor evidence of primary malignancy.

The patient underwent transmastoid excisional biopsy of the lesion. Intraoperatively, a soft tissue mass eroding the posterior and superior bony SCCs was identified. The lesion was removed and the superior semicircular canal (SSCC) occluded by packing the bony defect and lumen with fascia. The bony canal of the posterior SCC and common crura were partially eroded but underlying membranous labyrinth was not exposed. The residual soft tissue lesion adherent to posterior fossa dura was left in situ. Formal histopathology was consistent with otic capsule Langerhans cell histiocytosis (LCH).

Postoperatively, initially radiotherapy was considered. However, on further staging high resolution CT lungs showed multiple right-sided small nodules in the mid-to-upper lobe without cavitation consistent with pulmonary LCH. Bone marrow aspirate and trephine and skeletal survey excluded involvement and BRAF mutation was negative on molecular studies. It was subsequently recommended the patient trial conservative management with smoking cessation.

Remarkably, six months later the patient’s right-sided hearing loss had recovered (Figure 1B). Eight months postoperatively repeat MRI showed near complete resolution of the lesion. The patient successfully stopped smoking and remained asymptomatic with repeat CT lungs showing resolution of the lung nodules. 2.5 years postoperatively CT
demonstrated reossification in the site of the previous lucent lesion (Figure 2C & 2D). The patient has not required further treatment and continues yearly follow-up.

Discussion

LCH is a rare neoplastic proliferation of Langerhans cells of unknown aetiology\(^1,2\) but smoking is recognised as the main risk factor for pulmonary disease\(^3,4\). In our reported case the patient had asymptomatic pulmonary LCH with extrapulmonary otic capsule involvement\(^4\). Temporal bone involvement occurs in 15-61% of all LCH cases\(^5\) and lesions are usually isolated to the mastoid or squama, or rarely as part of multifocal disease\(^2\). The petrous portion is rarely involved and, in this event, the otic capsule is usually spared\(^6\). As a result, conductive hearing loss is common whereas SNHL has been reported in only 17 cases to date\(^1,3,6,7\). In our reported case, otic capsule involvement was mainly confined to the SCCs without middle ear involvement. The patient presented with SNHL with conductive overlay which, in conjunction with his symptomatology, is presumed related to labyrinthine inflammation secondary to LCH, and third window effect from SSCC erosion. Interestingly, cVEMP testing was equivocal and the authors postulate that this is because the hearing loss is likely multifactorial.

Smoking cessation and pulmonary LCH disease resolution as reported in our case has been previously described\(^4\). Treatment options for temporal bone LCH include intralesional or systemic steroids, systemic chemotherapy and surgical excision\(^1,2,6\). Surgical debridement has been reported to carry a high risk of complications such as postauricular fistula and facial nerve palsy\(^4\). Thus, surgical management has been controversial and is limited to obtaining
histological diagnosis, access for intralesional steroid injection or curettage. However, we present the first reported case of hearing recovery following surgical removal of an LCH lesion with SCC erosion, in turn isolating the labyrinth from the disease process and likely correcting the third window effect. Furthermore reossification of osteolytic temporal bone LCH lesions is associated with hearing recovery and the CT performed 2.5 years postoperatively for our patient supports this.

References


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**Figure Legends:**

**Figure 1:** Patient audiograms. 1A: Preoperative audiogram shows mild to moderate right sensorineural hearing loss with conductive overlay at 500Hz and 1000Hz and mild high frequency sensorineural hearing loss in the left ear. 1B: Audiogram six months postoperative showing bilateral, near-symmetrical mild high frequency SNHL.
Figure 2: Axial (2A) and coronal (2B) views of CT petrous temporal bone showing an ill-defined lucent mass in the superior aspect of the PTB apex with mastoid antrum extension, thinning of the tegmen mastoideum and cortex destruction including of the superior and posterior semicircular canals. Axial (2C) and coronal (2D) CT petrous temporal bone 2.5 years postoperatively showing reossification in the site of the previously demonstrated lucent lesion.
LCH Figure 1.png
Hearing recovery following repair of otic capsule defect secondary to semicircular canal erosion from Langerhans cell histiocytosis