CASE REPORT

A case of bilateral sensorineural hearing loss from Langerhans cell histiocytosis

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SUMMARY

Langerhans cell histiocytosis is a rare disorder, with resultant bilateral sensorineural hearing loss unreported in adults. A 42-year-old man presented with 4 months of right-sided tinnitus and hearing loss treated initially as otitis media with effusion. He re-presented 5 months later with progressive bilateral hearing loss—sensorineural (>100 dB) on pure tone audiogram. CT showed bilateral petrous temporal bone and calvarial lesions. Biopsy confirmed diagnosis of Langerhans cell histiocytosis and chemotherapy was started. Though uncommon, Langerhans cell histiocytosis should be considered among the differentials of persistent otological symptoms, as its progressive nature can cause bilateral irreversible sensorineural hearing loss.

BACKGROUND

Langerhans cell histiocytosis (LCH) is a rare disorder of unknown aetiology in which accumulation of pathological Langerhans cells leads to local tissue destruction. It can present as many otological symptoms including otorrhoea, periauricular mass and stenosis of the external auditory canal, or as polyps and eczematous lesions. Involvement of the temporal bone has been described in between 15% and 61% of cases,¹ however, sensorineural hearing loss (SNHL) is very rare.² We report the diagnostic challenge of a patient who developed bilateral SNHL from bilateral temporal bone involvement.

CASE PRESENTATION

A 42-year-old man presented with a 4-month history of bilateral tinnitus and right-sided hearing loss. One week prior to his appointment, he had developed mild imbalance and a further deterioration in the hearing in his right ear. His only medication was fluoxetine.

Initial examination suggested the possibility of middle ear fluid on the right with a normal left eardrum and a normal postnasal space on nasendoscopic examination. Pure tone audiogram (PTA) suggested a low frequency conductive loss on the right (20–30 dB) with mild high-tone loss on both sides (15–25 dB); impedance audiometry revealed normal middle ear pressures.

Nasal steroid sprays were started for a provisional diagnosis of otitis media with effusion (OME), despite the normal impedance result. On review 3 weeks later, the patient reported fluctuating hearing levels in the right ear and some very non-specific retroauricular pain. PTA again showed a conductive loss in the low frequencies (20–30 dB), but on this occasion, the impedance test

was flat on the right, and the patient was booked for right-sided myringotomy and grommet insertion.

Following several missed appointments, he had a mini grommet inserted 6 months after initial presentation (at which point his PTA showed no deterioration in hearing levels); it was noted during surgery that he had no middle ear effusion.

At a 3-month postoperative clinic follow-up, the grommet remained in situ, however, PTA now demonstrated severe SNHL on the right (70–80 dB), which was confirmed on auditory evoked brainstem response. The patient had concurrently been referred by his general practitioner to a private surgical clinic in the local area for assessment and possible excision of ‘cysts’ in the calvarium. Unfortunately, no access to these medical records was possible; however, partial drainage was attempted on one of the lesions, with rapid recurrence. No specimen was sent for histological analysis. In light of the calvarial cysts, he underwent CT of the head, which revealed two well-circumscribed lytic lesions in the skull vault ([figure 1](#)), and an expansile lytic lesion in the right mastoid temporal bone with invasion of both bony labyrinth and basal turn of cochlea.

After further failures to attend, including for MRI, the patient was reviewed at the 1 year stage. He had become increasingly deaf in the right ear and also reported crackling noises in the left ear. A decision was made to biopsy a lesion, however, the patient failed to attend for surgery. He eventually presented to a different hospital 6 months later, 18 months in total post initial presentation.

Figure 1 Bilateral calvarial lesions.

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Figure 2 MRI showing extensive oedematous signal change within the lytic petrous bone lesions.

At this time, PTA showed a profound hearing loss on the right (>100 dB), now with moderate SNHL of the left side (45–55 dB). Only 2 weeks later, PTA showed the left side to have deteriorated to the same level as the right (>100 dB). Repeat CT showed worsening of the bony lesions in the calvarium, with the changes previously noted in the right mastoid now also affecting the left; further discussions with neurosurgery resulted in urgent admission for biopsy. Specialist paediatric histopathological opinion was sought: the occipital bone biopsies showed infiltration with numerous, strongly S100 positive, eosinophils and mononuclear cells, some of which had curved and folded nuclei—from which a diagnosis of LCH was made. MRI showed no cerebellopontine angle region abnormality but there was extensive oedematous signal change in the lytic petrous bone lesions (figure 2). High resolution CT of the petrous bone confirmed bilateral LCH lesions invading the bony labyrinth, and invasion of the right vestibular aqueduct (figure 3A, B). Bone scan and chest CT revealed no other lesions; dermatological and respiratory specialist reviews at that stage revealed no evidence of further multisystem disease. Treatment with steroids produced only a very minimal improvement in hearing, and after further review by haemato-oncology, chemotherapy was started under specialist guidance.

DIFFERENTIAL DIAGNOSIS

Given the infrequent occurrence of LCH in adult ear, nose and throat clinics, it was not considered in the early stages of differential diagnosis. However, given the progression of symptoms and atypical findings on imaging, further consideration was warranted. In this case, the early differential of conductive hearing loss led to a diagnosis of Eustachian tube dysfunction/OME. This was in keeping with the flat impedance and middle ear fluid. The absence of fluid and the persistence of unilateral conductive hearing loss with a normal appearing middle ear may have steered the diagnosis initially toward otosclerosis. It was clear, however, that not only was there an absence of fluid in the middle ear at surgery (excluding otitis media as a cause), but the patient's symptoms had progressed. The combination of tinnitus, imbalance and progressive SNHL indicated inner ear or internal acoustic meatal

Figure 3 (A, B) High-resolution CT of the petrous bone confirmed bilateral Langerhans cell histiocytosis lesions invading the bony labyrinth. There was also invasion of the right vestibular aqueduct.

pathology, for which MRI was booked. In the first instance, this was to exclude acoustic neuroma or other cerebellopontine angle tumour, but it was also to assess for other structural inner ear abnormality. Development of multiple bony calvarial cysts, along with the findings on CT and MRI, and the progression to bilateral otological symptoms, was highly suggestive of an atypical or systemic condition not of primary otological origin. This presentation showed no sign of dermatological involvement, where dermatitis or xanthogranuloma and other histiocytoses may be considered among the differential. Further, the absence of respiratory and systemic disease was not immediately indicative of tuberculosis, sarcoidosis or a lymphoproliferative disorder. As in this case, the bony erosion of LCH in isolation can be difficult to differentiate from osteomyelitis or primary tumours of bone; all are uncommon presentations within the temporal bones/calvarium alone—and, as such, haematological and histological assessment was required for diagnosis. The expression of CD1a and S100 positive cells (as in this case) or the langerin protein are highly indicative of a diagnosis of LCH.

OUTCOME AND FOLLOW-UP

Postchemotherapeutic CT/MRIs have shown no evidence of further active temporal bone disease; however, there has been no improvement in the patient's hearing. Despite this, he remains under periodic review by dermatology and oncology to monitor for recurrence of disease.

DISCUSSION

This case demonstrates the diagnostic challenge of LCH-related hearing loss. In the adult population, pulmonary LCH is the

Learning points

- ▶ Langerhans cell histiocytosis (LCH) commonly involves the head and neck. Although rare in adults, it may present with a spectrum of otological problems, including sensorineural hearing loss.
- ▶ Initially, it may masquerade as more common conditions, and should thus be considered in the differential diagnosis of persistent or unusual patterns of otological symptoms.
- ▶ Owing to its progressive nature, LCH can produce severe disability with irreversible bilateral deafness, so early imaging and histopathological diagnosis are highly recommended.
- ▶ Owing to the often multisystem nature of the disease, early liaison with other specialities including dermatology, respiratory and haemato-oncology, is strongly advised.

most common form, and is usually seen in smokers. Although principally considered a paediatric disease, it can present in any age group, and may exhibit a bimodal distribution with a second peak around 30–39 years of age.³ The diagnosis of LCH for hearing loss may be suspected if the patient has otological symptoms as part of a multisystem disease; however, the temporal bone can be the sole bone involved.⁴

Conductive deafness caused by infiltrative changes in the middle ear, involvement of the ossicular chain and obstruction of the internal auditory canal have been described,¹ whereas SNHL and vertigo resulting from involvement of the bony

labyrinth are much more uncommon, at around 10% of cases.² Review of all available cases of LCH associated SNHL in the literature (19) by Saliba and Sidani,⁵ note the presence of temporal bone disease at around 20% of cases of LCH, with bilateral disease in around a quarter of these. Although bilateral disease has been seen in a very few cases in children, no report currently exists of bilateral SNHL in an adult, as this case demonstrates.

To start treatment, a definitive histopathological diagnosis is required. However, because the otological findings are similar to those of otomastoiditis or other otological pathology, diagnosis and treatment are often delayed.¹

Competing interests None declared.

Patient consent Obtained.

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