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CASE REPORT

Malignant lymphoma presenting as bilateral sensorineural hearing loss—A case report

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Abstract

Bilateral Sensorineural Hearing Loss is a rare disease that is often associated with other complex medical conditions. Primary central nervous system lymphoma is an uncommon and aggressive variant of non-Hodgkin lymphoma that can mimic many other neurological diseases. Herein, we present a rare case of lymphoma of the CNS as the etiology for progressive SSNHL. We describe a 58-year-old male with previous IgG4-disease presentation who was diagnosed with progressive sensorineural hearing loss. The condition evolved rapidly despite proper, conventional therapy. The patient acquired vestibular symptoms and other cranial nerve deficiencies and he was diagnosed with intracranial lymphoma, mainly in the cerebellar region. This case demonstrates that rare intracranial lymphoma can present initially as sensorineural hearing loss. A higher suspicion for malignancy should be held in mind for patients with a history of IgG4-related diseases and for patients presenting with progressing bilateral SSNHL that is not responding to therapy.

KEYWORDS

lymphoma, sudden sensorineural hearing loss

1 | INTRODUCTION

The incidence of sudden sensorineural hearing loss (SSNHL) is 5–20 per 100,000.[1] Bilateral SSNHL accounts for 1.7–4.9% of all patients with SSNHL.[2,3] Primary central nervous system lymphoma (PCNSL) has a varied clinical presentation, depending on the affected region and accounts for 3% of all intracranial primary tumors.[4] We present a case of progressive bilateral SSNHL where the etiology was PCNSL.

2 | CASE REPORT

A 58-year-old man was referred from the primary clinic to our hospital with bilateral sudden hearing loss following treatment of locked ears with antibiotics, pseudoephedrine and nasal corticosteroid. The patient had a history of asthma, sicca symptoms, immunoglobulin G4 (IgG4) autoimmune pancreatitis, and symptoms consistent with Mikulicz's disease. At presentation, his audiogram recorded deafness of the right ear and severe sensorineural

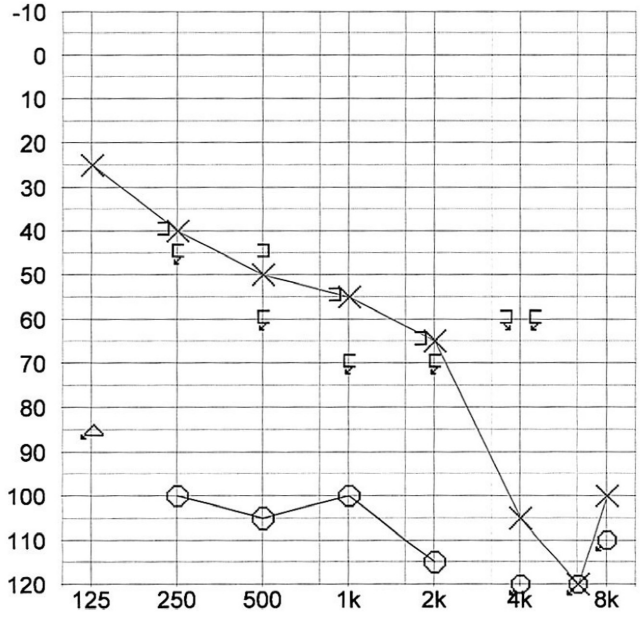

FIGURE 1 Audiogram taken at first admission presents SNHL in both ears, severe on the right. Pure tone audiometry 69/107 dB. Hearing threshold (dB) on *y*-axis, frequencies (Hz) on *x*-axis. O = right ear air conduction, X = left ear air conduction, [= right ear bone conduction,] = left ear bone conduction

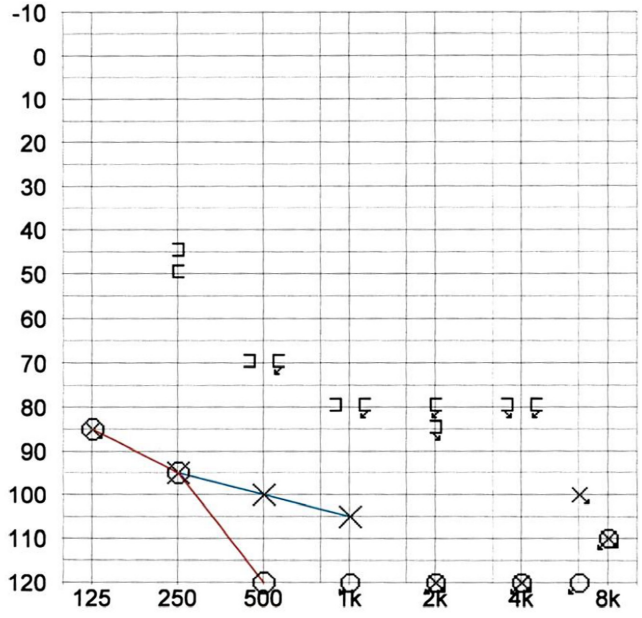

FIGURE 2 Audiogram taken 9 days after the first admission, severe SNHL in both ears. Hearing threshold (dB) on *y*-axis, frequencies (Hz) on *x*-axis. O = right ear air conduction, X = left ear air conduction, [= right ear bone conduction,] = left ear bone conduction. The bone conduction hearing measured here is most likely a mere tactile response from the patient, not an audiological finding

hearing impairment of the left ear (Pure tone audiometry (PTA) 69/107 dB) (Figure 1). The patient did not have a history of hearing loss, nor was there any particular hearing loss in the family. Physical examination of the upper respiratory tract was normal. Otomicroscopy findings of the ears were normal. The patient was administered methylprednisolone 64 mg daily.

Despite oral corticosteroids, the patient's hearing declined within a week to deafness of both ears (PTA 120/103 dB) (Figure 2). On the next physical examination, blisters were found on the lips, tongue and oral cavity. The head thrust test was positive to the left, and spontaneous nystagmus to the right could also be seen. Ramsay–Hunt syndrome was suspected; therefore, valaciclovir (1 g three times a day) and hyperbaric oxygen treatment were initiated, and methylprednisolone was continued (32 mg daily).

Within a week, the patient presented with left-sided facial nerve palsy and vertigo. The patient was admitted to the otorhinolaryngology ward for and a higher dose (64 mg daily) of methylprednisolone was administered. An extensive magnetic resonance imaging (MRI) of the brain showed left-sided periventricular T2 signal focuses in the area of the corpus callosum genu, fornix and temporal lobe. A similar lesion was seen in the right cerebellum. (Figure 3) Contrast enhancement was seen in the left facial nerve (VII), as well as in the left vestibulocochlear nerve (VIII), and in the right VII-VIII nerves at the site of the porus acusticus.

Neurologists and a rheumatologist specialized in IgG4 disease were consulted and a central nervous system (CNS) affiliation of the IgG4-related disease was suspected, with elevated serum IgG4 levels, 2.71 g/L (reference value 0.08–1.4 g/L). The patient was admitted to the neurological ward, where a cerebrospinal fluid (CSF) investigation showed leukocytosis, 22×10^6/L (reference value 0–5 $\times 10^6$/L), and elevated levels of total IgG, 55 mg/L (reference value 13–36 mg/L). Pulse steroid therapy (methylprednisolone 1 g daily) was administered without regression of the symptoms. A week later a control MRI showed progression of the expansions in the brain (Figure 4).

The patient was referred to a neurosurgeon for a stereotactic biopsy and the histopathology confirmed PCNSL with no signs of IgG4 (Figure 5). A full-body computer tomography (CT) scan showed no other neoplasms. Chemotherapy was planned, but the patient's state rapidly deteriorated and he presented with emesis, fatigue, headache, and finally he became unconscious. To exclude hemorrhage of the brain, a new CT scan (Figure 6) was administered, in which rapid growth of the tumors, progression of a hemorrhage and disturbed CSF circulation could be seen. The cerebral hemorrhage was interpreted as secondary to tumor growth, but could have been a complication following stereotactic biopsy.[5] Soon after, the patient succumbed. The autopsy stated herniation of the cerebellum as the acute cause of death and CNS lymphoma as the primary cause of death; no signs of IgG4 disease were found in the CNS.

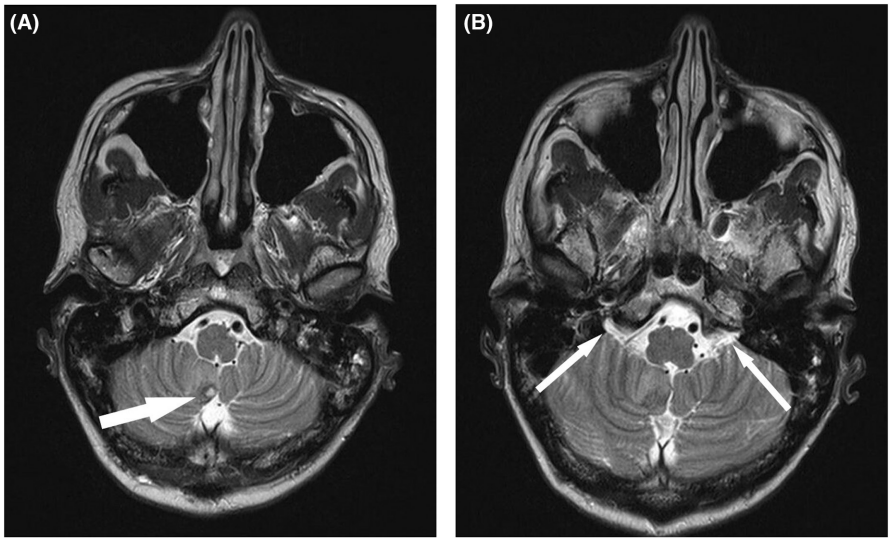

FIGURE 3 MRI taken 15 days after first admission. A lesion is present in the cerebellum (arrow). Other lesions were found in the corpus callosum genu, fornix and temporal lobe (A). Some enhancement was seen in the left facial nerve (VII), as well as in the left vestibulocochlear nerve (VIII), and in the right VII-VIII nerves at the site of the porus acusticus (arrow9 (B))

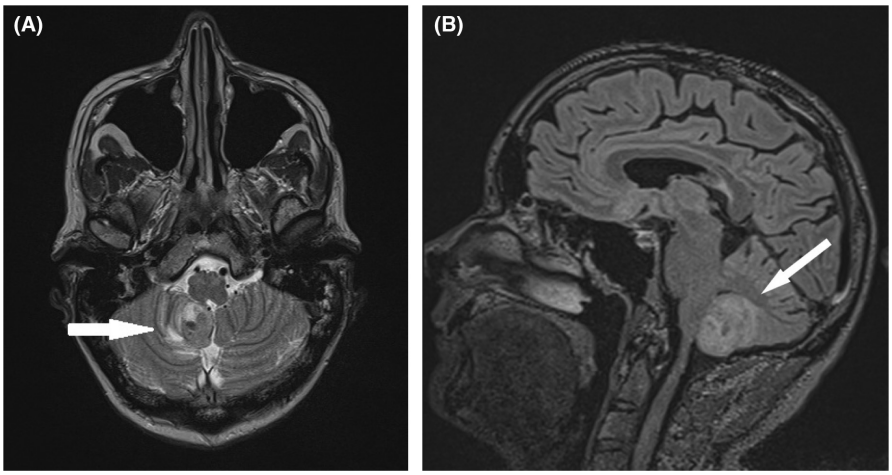

FIGURE 4 MRI taken 24 days after first presentation, 9 days after first MRI. The lesion in the cerebellum has grown substantially (arrow) and is compressing the brain stem. Axial plane (A), sagittal plane (B)

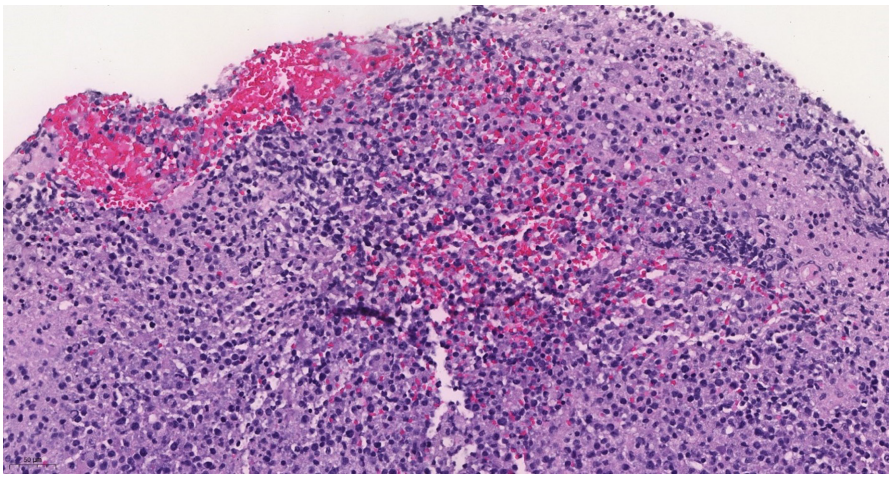

FIGURE 5 Hematoxylin–eosin stained (HE) brain tissue biopsy demonstrates diffuse infiltrate of large atypical lymphocytes with perivascular infiltration as well, consistent with the lymphoma diagnosis. Magnification ×200

3 | DISCUSSION AND CONCLUSION

The etiology of bilateral SSNHL is not fully identified. However, the disease is often associated with autoimmune, toxic, neoplastic and vascular conditions, whereas the unilateral form is predominantly idiopathic. Patients with bilateral SSNHL have more profound hearing loss, show poorer recovery and up to a 15–35% mortality rate, whereas up to 65% of patients with unilateral SSNHL spontaneously recover hearing. Corticosteroids are the most commonly used treatment in both forms; however,

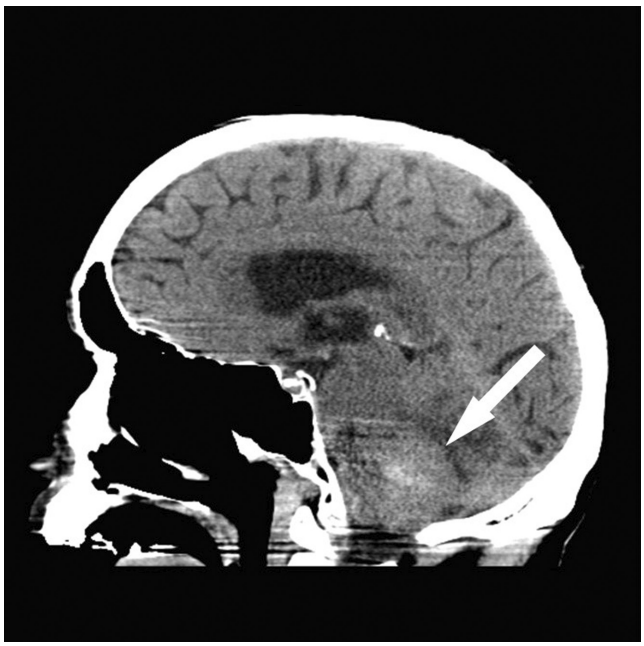

FIGURE 6 CT scan taken 6 weeks after the first presentation. The lesions in the cerebellum have grown (arrow), accompanied by hemorrhage, which led to the herniation of the tumor-infiltrated cerebellum into the foramen magnum and compression of the brain stem

in bilateral SSNHL, they are effective in only 50% of cases.[1]

In the case of progressive SSNHL, appropriate work-up includes MRI of the CNS, as well as blood autoantibody tests. The MRI is diagnostic for tumors, intracranial infection, and labyrinthitis. The autoantibody tests are diagnostic for autoimmune diseases (e.g., Cogan's syndrome).

PCNSL is an uncommon and aggressive variant of non-Hodgkin lymphoma with an incidence of 1.6 per million a year.[4] The malignancy can mimic many other neurological diseases. Risk factors include AIDS, other autoimmune diseases and immunosuppression. Clinical behavior is heterogeneous, but symptoms may include cognitive decline, personality changes, focal neurological deficits, increased intracranial pressure, seizures and ocular symptoms.[6] Work-up for PCNSL includes a complete blood count, electrolyte analysis, HIV serology, serum lactate dehydrogenase level, contrast-enhanced MRI, and a complete ophthalmology evaluation, as well as cytologic evaluation and flow cytometry of CSF through lumbar puncture. A biopsy will determine the diagnosis. Further studies in the work-up include a bone marrow biopsy and a CT scan of the chest, abdomen, and pelvis. For male patients, testicular examination, clinically and with ultrasound, is recommended. The use of positron emission tomography imaging is still under debate for PCNSL.[7]

IgG4-related disease is a relatively new diagnosis, where sclerosing masses are formed due to an unknown etiology and can be found almost anywhere in the human body, also in the CNS.[8] The disease presents with elevated serum IgG4 and characteristic histopathological features. Many previously described fibroinflammatory and sclerosing disorders are today considered as IgG4-related diseases. Patients with IgG4 disease have an increased risk for malignancy compared to the rest of the population.[9,10] The work-up for IgG4-related disease of the nervous system includes clinical examination, MRI, tissue sample, and serum IgG4.

In our case, it was first believed that IgG4 was the primary cause of the symptoms presented by the patient and treatment was administered accordingly. Despite oral corticosteroids, within a week the patient's hearing declined to deafness of both ears and he presented with left-sided facial nerve palsy and vertigo. The head thrust test was positive to the left, and spontaneous nystagmus to the right could also be seen. These multiple cranial nerve involvements were a sign of lesions in the brain stem. An MRI showed a rapid progression of the expansions of the brain and it was only after a spinothalamic tract biopsy that the PCNSL diagnosis was achieved. Unfortunately, the lymphoma was an aggressive type and in a fatal location and the patient passed away despite treatment.

This case highlights a rare lymphoma of the CNS as the etiology for progressive SSNHL. When bilateral SSNHL is not responding to therapy or is progressing, differential diagnostics should be reconsidered and further work-up is needed with MRI. The poorly responding or progressive SSNHL might be a sign of a malignant disease with fast progression; therefore, a high suspicion is to be held in mind and collaboration between specialties is essential. If a central nervous lesion is discovered, biopsy should be taken promptly to verify the diagnosis and begin the proper therapy. A higher suspicion of malignant diseases should be held in mind for patients with a history of IgG4 disease.

AUTHOR CONTRIBUTIONS

LH: Conceived the idea of the study. LH: Performed data collection. FR: Primary author (wrote most of the paper). LH, JK: Provided revisions to scientific content of manuscript. FR, LH: Provided stylistic/grammatical revisions to manuscript. JK: Provided access to crucial research components. LH: Principal investigator (advisor, manager).

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Not applicable.

CONFLICT OF INTEREST

The authors declare no conflicts of interest.

DATA AVAILABILITY STATEMENT

The data that supports the findings of this study are available in the supplementary material of this article.

ETHICAL APPROVAL

This article does not contain any studies involving human participants performed by any of the authors.

CONSENT

Written informed consent was obtained from the patient's next of kin to publish this report in accordance with the journal's patient consent policy.

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