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Case report

A sudden bilateral hearing loss caused by inner ear hemorrhage

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ABSTRACT

Introduction: Labyrinthine hemorrhage is a rare cause of sudden deafness and generally concerns only on one side.
Case summary: An 84-year-old man with a past medical history of myelomonocytic chronic leukemia (CMML) suffered from sudden bilateral hearing loss associated with vertigo. The audiogram revealed a left cophosis and a right profound deafness. Videonystagmography showed a left vestibular deficit. The MRI showed a spontaneous strong T1 weighted signal in the left and right labyrinths, corresponding to a bilateral inner ear hemorrhage (IEH). Dizziness resolved rapidly following vestibular physiotherapy, in contrast to hearing which did not improve at all and let the patient isolated in his environment. The patient successfully underwent cochlear implantation so that he could communicate.
Discussion: Most IEHs are unilateral and due to anticoagulants treatments and hematological diseases. Only rare cases have described bilateral labyrinth hemorrhage. This is the first case reported of bilateral labyrinth hemorrhage due to CMML.

1. Introduction

Sudden deafness represents a frequent cause of emergency consultation in otology. Most of the time, it concerns only one ear. Generally, no etiology can be identified and a viral infection is suspected. However, diagnosis requires a clinical workup including serologies, complete blood count and a MRI which detects pathologies such as tumor or inner ear hemorrhage (IEH). In these cases a spontaneous strong T1-weighted signal, no enhancement and no FLAIR extenuation are seen [1]. It is a rare cause of sudden deafness. Here we report a case of bilateral inner ear hemorrhage due to hematological conditions.

2. Case report

An 84-year-old man complained of bilateral deafness, mostly on the left side for two days. Vertigo started three weeks previously. The patient had a past medical history of prostate and colon cancers, high blood pressure, dyslipidemia and untreated chronic myelomonocytic leukemia. His treatment included only calcium blocker.

Otoscopy was normal. A spontaneous right horizontal nystagmus was observed with videonystagmoscopy. No neurological deficit was present.

The audiogram showed left ear cophosis and right ear subcophosis (Fig. 1). The patient underwent a five-day treatment of 1 mg per kilogram per day of intravenous corticosteroids and vasodilator Carbogen aerosols (mix of oxygen and 10% of carbon dioxide), combined with vestibular physiotherapy.

A cerebral CT scan was normal. Serologies were negative and complete blood count showed a subnormal count of platelets (126,000 per mm^3) but a high level of monocytes (3040 per mm^3). Coagulation results were normal.

The head shaking test confirmed the right horizontal nystagmus. Because of difficulties to communicate, the patient did not keep his eyes open, so that the caloric test could not be completed.

A control audiogram was performed seven days later and revealed no change in the left ear but aggravation in the right one, as the hearing threshold was 105 dB.

MRI showed a spontaneous strong T1 weighted signal of the left labyrinth and the posterior right labyrinth (Fig. 2A). There was no enhancement (Fig. 2B) and no attenuation on FLAIR. These elements were in favor of an intralabyrinth hemorrhage.

Dizziness regressed contrary to his deafness. Hearing aids were tried but were not sufficient for correct communication. He underwent cochlear implantation four months later on the left side. Bleeding was remarkably important during the procedure, but

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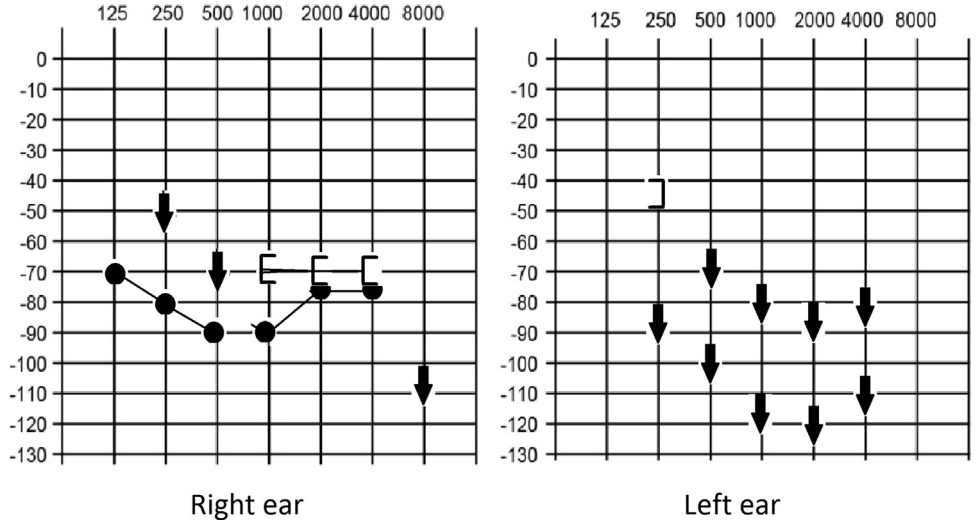

Fig. 1. Tonal audiogram on day 1.

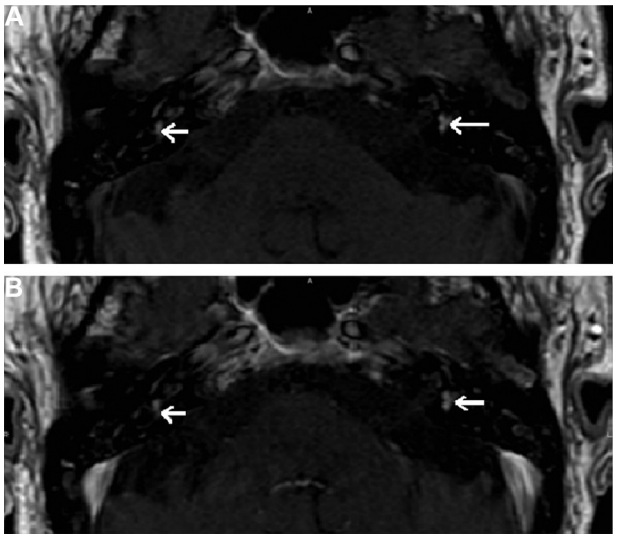

Fig. 2. Internal auditory canal MRI. A. T1-weighted image. B. Contrast-enhanced T1-weighted image. The arrows show the spontaneous strong signal of the right and left posterior labyrinths.

all electrodes could be implanted without any difficulty. This implantation was a success as tonal and vocal audiograms showed (Fig. 3). The patient understood 90% of words and 95% of sentences. He could participate in conversations with one person, but still had difficulties in noisy environments and when telephoning.

3. Discussion

Inner ear hemorrhage (IEH) is rare. As in our case, it induces sudden deafness with pronounced vertigo and tinnitus. These symptoms can occur simultaneously [2] or delayed [3]. Deafness is profound most of the time. Hearing level rises from 80 dB [4] to cophosis [1,2].

In our case, the two labyrinths were not affected in the same region, but the consequences on hearing were comparable. Wu et al. described that MRI [1] detected blood in the cochlea, the vestibule or the semicircular canal. Regardless of where the hemorrhage occurs, it always induces severe to profound deafness.

Various causes of IEH have been identified. Anticoagulant treatments are often implicated in cases of overdose [2]. Similarly, antiplatelet treatment is often involved [4]. However, our patient did not receive any treatment which could have a role in coagulation.

The second most frequent etiology is hematological diseases, as our case. IEH seems to be a frequent way of discovery of them. Myeloma[5] and Waldenstrom's disease [3] can also be encountered.

Auto-immune diseases such as systemic lupus erythematosus [6] can also induce sensorineural hearing loss by IEH. In the case of

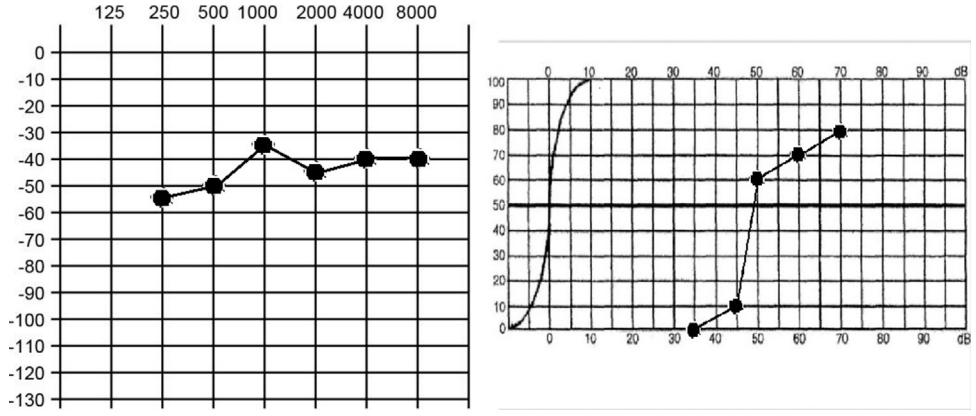

Fig. 3. Tonal and vocal audiograms seven months after implantation, with a left cochlear implant and right hearing aids.

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systemic lupus erythematosus, it is suspected to be a hemorrhagic transformation following thrombosis.

Head and neck radiotherapy is another cause [7]. IEH can happen up to twenty years after irradiation. Despite his oncological history, our patient did not undergo head and neck irradiation. Moreover, physical or chemical aggressions of the inner ear induce bleeding. Consumption of toxics products, such as cocaine with its vascular effects [8], can lead to IEH. Meningitis with bacterial diffusion can also be implicated [9] but the clinical presentation is different with neurological symptoms.

The other specificity of our patient's case is the bilaterality of the symptoms. The causes of bilateral IEH already described in the literature are Waldenstrom's disease [3], myeloma [5], systemic lupus erythematosus [6] and meningitis [9]. Our case report is the first one to describe bilateral deafness in this pathology. Cervantes et al. [10] reported a case of unilateral inner ear hemorrhage in a patient with chronic myelomonocytic leukemia.

In the other cases reported, no auditory recovery was observed and cochlear implant was sometimes proposed [1,9]. Cochlear implant was efficient. In our case, a second implantation can be considered since there was no ossification after four months.

4. Conclusion

Inner ear hemorrhage is a rare cause of bilateral sudden hearing loss. It can affect patients suffering from CMML.

Disclosure of interest

The authors declare that they have no competing interest.

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