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A CASE OF SENSORINEURAL HEARING LOSS DUE TO SUPERFICIAL SIDEROSIS OF THE CENTRAL NERVOUS SYSTEM: MULTIDISCIPLINARY EVALUATION

Contributions:
A Study design/planning
B Data collection/entry
C Data analysis/statistics
D Data interpretation
E Preparation of manuscript
F Literature analysis/search
G Funds collection

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Abstract

Introduction: Superficial siderosis of the central nervous system (SS-CNS) is a clinical condition characterized by a pathological accumulation of hemosiderin in the superficial or sub-superficial layer of the central nervous system, causing progressive and irreversible neurological alterations. The most common clinical presentation is characterized by hearing loss, cerebellar dysfunction (with dysarthria and ataxia), and cognitive impairment.

Case report: We describe the case of a man aged 53 with a sudden worsening of a pre-existing bilateral hearing loss. Clinical examination showed left head tilt, left upper limb paresis, and ataxic gait. A brain CT, US of supra-aortic trunks, and ECG ruled out cardiovascular and focal diseases. The patient was referred to the head-and-neck department for further investigations. Tonal audiometry showed a profound bilateral hearing loss greater on the left at low frequencies with no hearing above 1 kHz. Vestibular examination revealed the absence of nystagmus. The ABR revealed an unstructured trace without sign of any main waves. Transtympanic electrocochleography showed a clear bilateral first wave. A brain MRI revealed hypointensity due to hemosiderin deposition. A lumbar puncture demonstrated abundant red blood cells and xanthochromia, high ferritin, and protein levels in the cerebrospinal fluid.

Conclusions: From the multidisciplinary evaluation the final diagnosis was superficial siderosis of the central nervous system.

Key words: magnetic resonance imaging • superficial siderosis • sensorineural hearing loss • multidisciplinary approach

PRZYPADEK NIEDOSŁUCHU ODBIORCZEGO SPOWODOWANEGO POWIERZCHNIOWĄ HEMOSYDEROZĄ OŚRODKOWEGO UKŁADU NERWOWEGO: OCENA WIELOSPECJALISTYCZNA

Streszczenie

Wprowadzenie: Powierzchniowa hemosyderoza ośrodkowego układu nerwowego (SS-CNS) jest chorobą charakteryzującą się nieprawidłowym odkładaniem się hemosyderyny w powierzchniowej lub podpowierzchniowej warstwie ośrodkowego układu nerwowego, co powoduje postępujące i nieodwracalne zmiany neurologiczne. Do najczęstszych objawów klinicznych należą: niedosłuch, zaburzenia funkcji móżdżku (w tym dyzartria i ataksja) oraz zaburzenia poznawcze.

Opis przypadku: Opisujemy przypadek 53-letniego mężczyzny, u którego wystąpiło nagłe pogorszenie wcześniej obecnego obustronnego niedosłuchu. W badaniu klinicznym stwierdzono przechylenie głowy w lewo, porażenie lewej kończyny górnej i niezborny chód. Kolejne badania – tomografia komputerowa mózgu, ultrasonografia pni nadaortalnych i EEG – wykluczyły choroby układu krążenia i odogniskowe. Pacjent został skierowany do kliniki głowy i szyi w celu przeprowadzenia dalszych badań. Audiometria tonalna wykazała obustronny niedosłuch stopnia głębokiego w zakresie niskich częstotliwości, większy po stronie lewej, z całkowitym brakiem słyszenia powyżej 1 kHz. Badanie funkcji przedsionkowych wykazało brak oczopląsu. W badaniu ABR zarejestrowano bezkształtny wykres bez śladów głównych fal. Elektrokochleografia transtympanalna wykazała wyraźną falę pierwszą po obu stronach. W badaniu MRI widoczne były obszary o mniejszym natężeniu sygnału spowodowane złogami hemosyderyny. Punkcja lędźwiowa wykazała obecność w płynie mózgowo-rdzeniowym licznych krwinek czerwonych, ksantochromię oraz wysoki poziom ferrytyny i białek.

Wnioski: Na podstawie oceny wielospecjalistycznej postawiona została ostateczna diagnoza powierzchniowej hemosyderozy ośrodkowego układu nerwowego.

Słowa kluczowe: obrazowanie metodą rezonansu magnetycznego • powierzchniowa syderoza • niedosłuch odbiorczy • podejście multidyscyplinarne

Introduction

Superficial siderosis of the central nervous system (SS-CNS) is a clinical condition characterized by a pathological accumulation of hemosiderin in the superficial or sub-superficial layer of the CNS, causing progressive and irreversible neurological alterations [1]. The most common clinical presentation is characterized by hearing loss, cerebellar dysfunction (with dysarthria and ataxia), and cognitive impairment [2]. Frequent aetiologies of SS-CNS include head trauma, cerebral neoplasms, and aneurysms [3], but up to 50% of cases described in the literature are due to unknown causes of bleeding [4].

Case presentation

A man aged 53, after sudden worsening of a pre-existing bilateral hearing loss, came to the emergency room of our hospital. Anamnesis highlighted previous head trauma at a young age following a motorcycle accident, with associated lesion of the left brachial plexus resulting in hypoesthesia and paretic outcomes. Over the years, the patient had shown progressive bilateral hearing loss, more pronounced on the left. Repeated medical treatments, even with recent steroid therapy, had not yielded appreciable improvement.

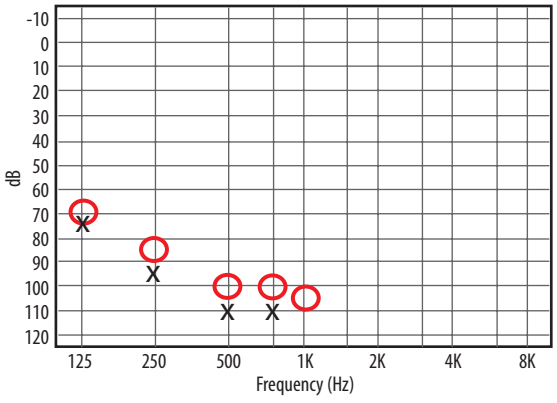

Figure 1. Tonal audiometry showing profound bilateral hearing loss greater on the left

The first clinical examination showed cervical hyperkyphosis associated with left head tilt, left upper limb paresis, and ataxic gait. The patient had a normal blood test including negative serological profile for recent infections and negative autoantibody profile. A brain CT, US of supra-aortic trunks, and ECG ruled out cardiovascular and focal diseases.

The patient was referred to the audiology and otology unit of the otorhinolaryngology department for further investigations. Tonal audiometry showed a profound bilateral hearing loss greater on the left at low frequencies with no hearing for frequencies above 1 kHz (**Figure 1**). Vocal audiometry demonstrated a partial articulation curve without reaching perception threshold. Impedance measurement revealed a bilateral type A tympanogram with absent acoustic reflexes in both ipsilateral and contralateral recordings. A vestibular examination revealed the absence of nystagmus and caloric labyrinth stimulation revealed normal bilateral reflexes.

ABR revealed an unstructured trace without any of the main waves being present (**Figure 2**). Cervical myogenic vestibular evoked potentials (cVEMPs) were not executable due to pre-existing cervical paramorphism with head tilt.

Transtympanic electrocochleography (ECoG) revealed a trace that showed distinct peaks that could be clearly identified bilaterally (**Figure 3**).

We brought the case to the attention of a neuroradiologist who carried out diagnostic imaging. MRI angiography highlighted normal morphology of the main intracranial vessels.

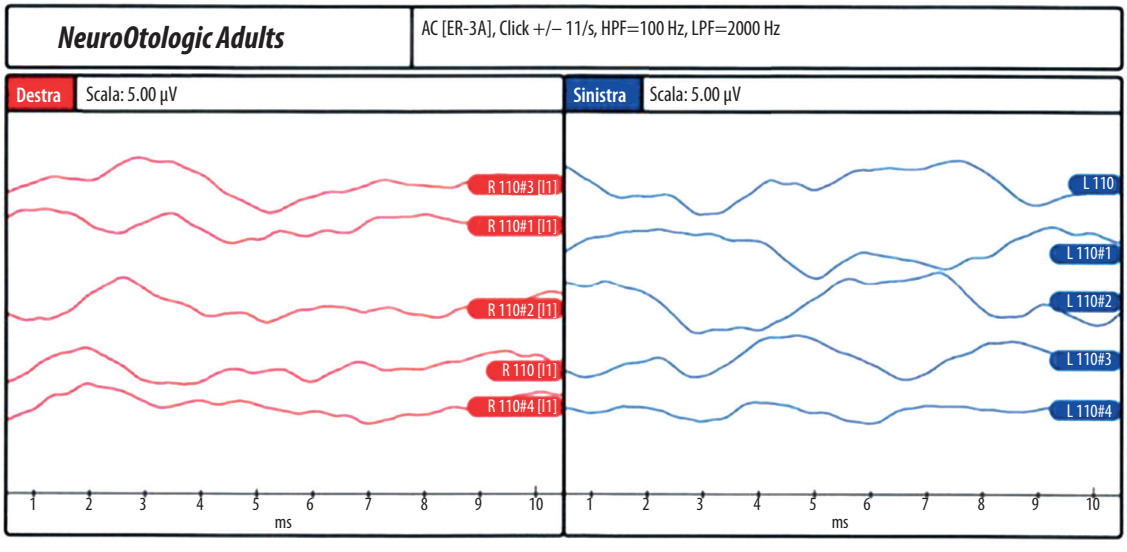

Figure 2. Auditory Brainstem Response (ABR) revealing an unstructured trace without any of the main waves

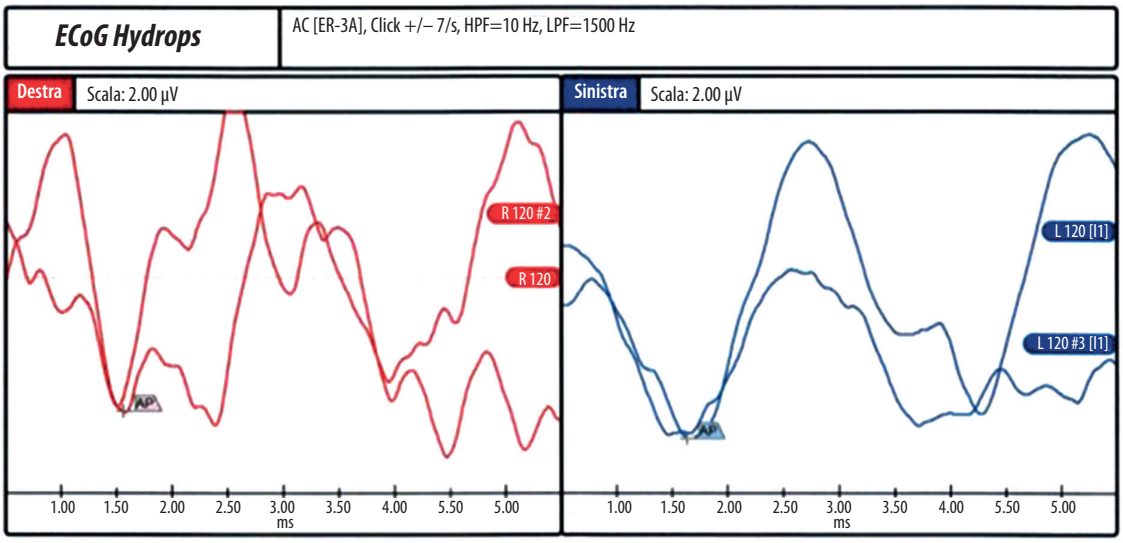

Figure 3. Transtympanic electrococleography (ECoG) revealing a trace with artifacts and the presence of a main wave clearly identifiable bilaterally

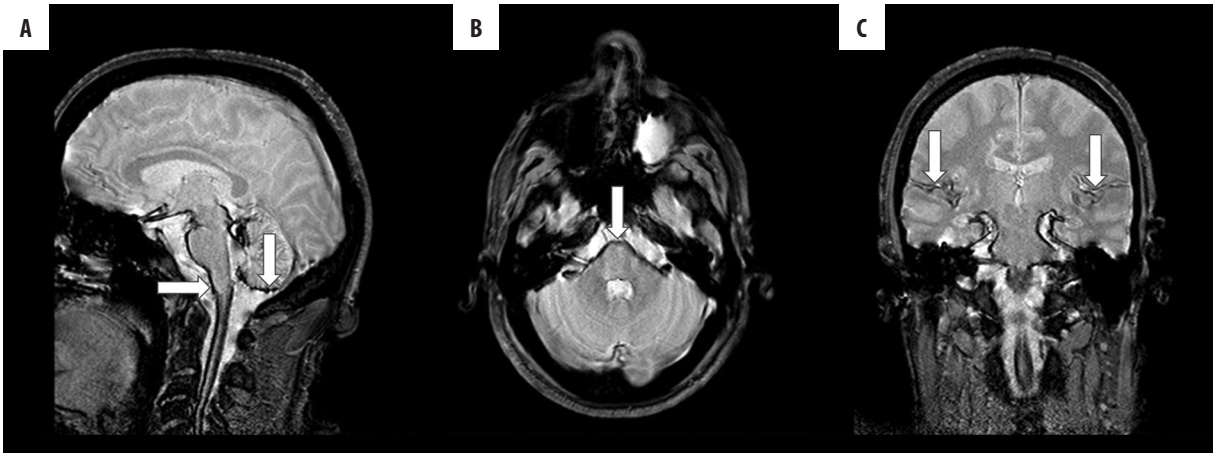

Figure 4. (**A–C**) MRI (T2*-GRE) showing bilateral hypointensity on the pial surface of the trunk, of the cerebellar sulci, and along the subarachnoid course of the main cranial nerves (arrows). This was attributed to hemosiderin deposition in the context of diffuse cortical involution

A brain MRI with and without contrast revealed bilateral hypointensity on the pial surface of the trunk, of the cerebellar sulci, and along the subarachnoid course of the main cranial nerves. This was attributed to hemosiderin deposition in the context of diffuse cortical involution (arrows in **Figure 4**).

The patient was then evaluated by a neurologist who performed a lumbar puncture. It revealed abundant red blood cells and xanthochromia, high ferritin, and high protein levels in the cerebrospinal fluid.

Discussion

This report underlines the importance of a multidisciplinary approach among different disciplines: otolaryngology, audiology, neurology, and neuroradiology. The role of the audiologist was to identify the type, severity, and location of the hearing loss. The neurologist excluded the presence of any related neurological syndromes and performed the lumbar puncture. The neuroradiologist revealed the typical hemosiderin deposition on MRI.

Most hearing tests revealed a deep bilateral sensorineural hearing loss. ABR examination and electrocochleography were therefore performed. In the first exam no main waves were detectable, in the second a first wave was present: such a discrepancy argues for a central nervous conduction problem.

Differential diagnosis in patients with sudden worsening of a pre-existing bilateral hearing loss includes several pathological conditions: primary causes are vascular insufficiency, viral infections, and autoimmune disorders; secondary causes may be stroke, neoplasm, or trauma [5].

The clinical history and age were initially suggestive of severe worsening of a cochlear disorder, probably of a vascular basis. However, the negative family history of cardiovascular disease or of diabetes, as well as the CT scan

of the head, the ECG, and the US of epi-aortic trunks excluded the hypothesis of vascular lesions.

Similarly, serological negativity excluded viral infection or autoimmune aetiology. Furthermore, after steroids the patient did not show any clinical benefit. The patient did not report recent exposure to noise or trauma, except for the early head trauma from the motorcycle accident.

The occurrence of the trauma, even though apparently not directly related to the present clinical presentation, led us to suspect a possible case of SS-CNS induced by a single-episode of previous traumatic subarachnoid haemorrhage, as has been described in the literature [6].

The brain MRI confirmed the absence of tumoral lesions but highlighted the presence of an hypointense signal in the weighted T2 sequences, involving the CNS at the level of the posterior cranial fossa and especially on the trunk, the cerebellum, and the main nervous structures, all of which are compatible with SS-CNS. In this context, Stabile and colleagues have described MRI finding of SS-CNS resulting from hemosiderin [7]; apparently hemosiderin creates a local magnetic field inhomogeneity generating a loss of signal on T2*-GRE and SWI sequences, with a low-intensity signal seen in those parts of the nervous system adjacent to CSF [7]. In addition, as well as the SS-CNS, atrophy and signal intensity abnormality of the involved regions also suggest tissue damage [8].

As a result of the growing use of MRI, SS-CNS is increasingly being recognized, including in asymptomatic patients. The most frequent aetiology is dural disease; in fact several authors believe that chronic bleeding originating from fragile blood vessels around the dural tear may be the likely underlying mechanism [9]. Imaging, although highly suggestive, is not pathognomonic of SS-CNS and it is advisable to extend the investigation to the whole rachis to exclude other possible causes. The presence of red blood cells in the cerebrospinal fluid is the defining feature of SS-CNS [10].

Regarding treatment options, currently there are no internationally validated therapies. The only reasonable therapeutic strategy is the management of bleeding of known aetiology. Additional medical therapies include iron alkylating agents or steroids. Another possible option is the use of acoustic prosthetic rehabilitation, based on the placement of a cochlear implant. In the literature, heterogeneous results have been reported, with poor, partial, or good auditory recovery [11]. Such inconsistent results are a consequence of the type of cochlear damage. This suggests that an indication for a cochlear implant should be guided by audiological results [12]. However, doubts remain about the long-term efficacy [13] because only a few studies have been reported with 5 years or more follow-up [14].

Conclusions

SS-CNS should be suspected in cases of patients with hearing impairment, especially if associated with ataxia, after exclusion of primary causes. A multidisciplinary approach is highly recommended. A brain MRI might show the presence of haemosiderin deposits, and lumbar puncture should be performed to confirm the diagnosis since the defining feature of SS-CNS is red blood cells in the cerebrospinal fluid.

Conflict of interest

The authors have no conflict of interest to declare.

Financial disclosure

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