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Sudden sensorineural hearing loss: when ophthalmology meets otolaryngology

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Abstract. *Sudden sensorineural hearing loss: when ophthalmology meets otolaryngology. Objectives*: Sudden sensorineural hearing loss is a perplexing entity in otology. Susac's syndrome (also called retinocochleocerebral vasculopathy) is a rare disorder that consists of microangiopathy of the brain, retina, and inner ear, and usually affects women in young adulthood. We describe the clinical aspects, radiographic findings, and management of one such case.
Case report: A 30-year-old woman was admitted to the hospital because of sudden onset of bilateral deafness and headache. During her hospitalization, she developed discrete right hemiparesis and hypoesthesia.
Results: Magnetic resonance imaging revealed multiple signal hyperintensities and atrophy of the corpus callosum. The differential diagnosis was a myelinating condition, such as multiple sclerosis or acute demyelinating encephalomyelitis.
Conclusion: Retinal fluorescein angiography helped the diagnosis of Susac's syndrome.

Introduction

An enthralling challenge for otologists is to determine the cause of and effectively treat sudden sensorineural hearing loss (SNHL). Most of the time, and despite numerous studies published on this topic, the cause remains uncertain and finding guidelines that address this problem in a comprehensive manner is difficult.[1-4] Susac's syndrome is a rare disorder of unknown etiology initially described in 1979 by John O. Susac.[5] He described two women with personality changes, paranoid psychosis, hearing loss, neurological dysfunction, and multiple branch retinal artery occlusions. In 1992, Schwitter *et al*.[6] proposed the acronym "SICRET" (small infarcts of cochlear, retinal, and encephalic tissues). Susac's syndrome mainly affects women without ethnic preference and has been described in patients aged 8 to 59 years.[7] The triad of fluctuating SNHL, loss of vision, and encephalopathy characterizes Susac's syndrome.[5-12] However, all three elements of the triad may not be present or not recognized as such, particularly at the beginning of the disease. The onset of involvement of each organ is variable, whereas early brain involvement is encountered most often. We report the case of a female patient initially admitted for sudden hearing loss. Some non-specific neurological complaints were noted in the patient's medical history, but without visual disturbances.

Case report

Three weeks after giving birth to a normal child, a 30-year-old woman was admitted to our hospital for sudden onset hearing loss, which affected the right ear more than the left. The clinical otolaryngological examination was normal. A pure tone audiogram revealed bilateral asymmetric SNHL, mainly involving the low and medium frequencies (Figure 1A). Speech audiometry revealed poor speech discrimination, particularly for the right ear (Figure 1B). The patient was treated with intravenous methylprednisolone (5 mg/kg/day for 3 days, followed by dosage tapering over 15 days). Partial auditory recovery was noted for the left ear. In the few days preceding the hearing disturbances, the patient complained of headache, attention deficit, personality and mental changes, and impaired cognition and memory. Three episodes of aseptic meningitis were noted in the patient's medical history, at the age of 5, 14, and 27 years. Between these events, the patient suffered from migraine-like headache and atypical polyarthritis.

Neurological examination showed non-specific signs of a light right pyramidal disorder: right hyper-

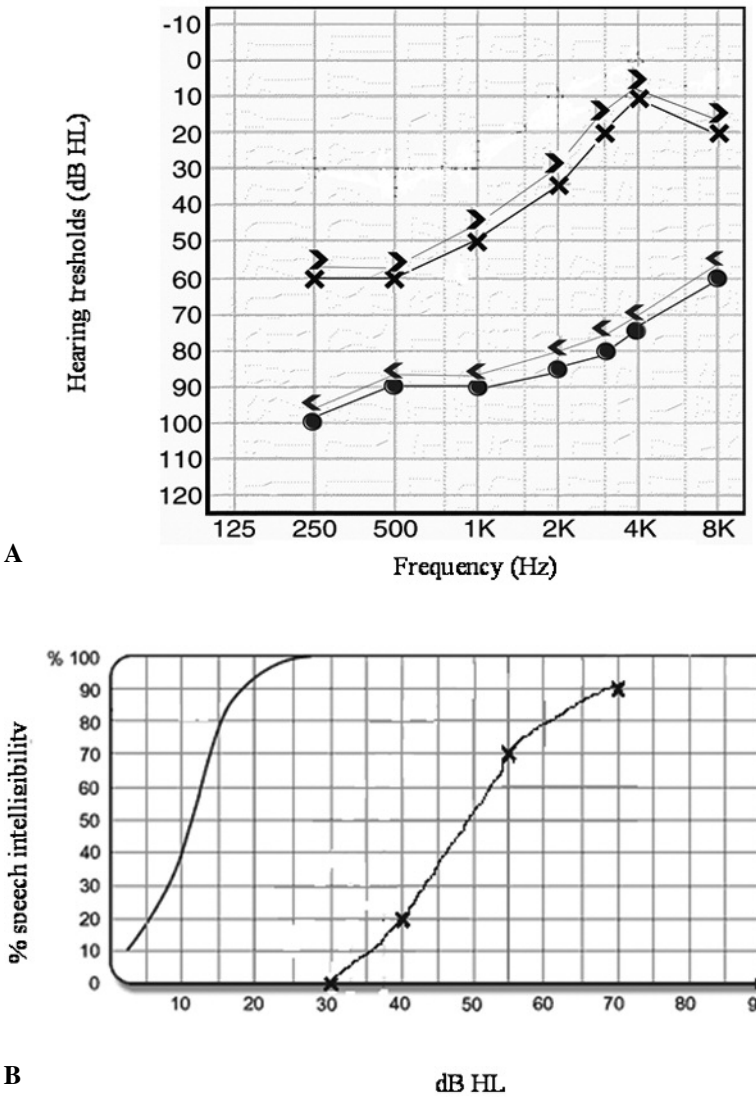

Figure 1
A: A pure tone audiogram revealed low-frequency SNHL in both ears. Hearing loss was moderate for the left ear and profound for the right ear. B: Speech audiometry showed poor speech discrimination, especially for the right ear.

reflexia and right Babinski and Hoffman signs. Laboratory analyses, including complete blood cell count, erythrocyte sedimentation rate, electrolytes, liver and kidney function, B12 and folic acid, Borrelia serology, anti-nuclear antibodies, and antineutrophil cytoplasmic antibodies, were normal. Cerebrospinal fluid analysis was also normal. In particular, no oligoclonal band was detected. Electroencephalography revealed runs of bihemispheric delta wave complexes. Visual-evoked potentials were abnormal; the responses were delayed and of reduced amplitude, indicating abnormal optic nerve function, particularly for the left eye. A CT scan of the brain, including temporal bone images, was normal. On magnetic resonance imaging (MRI), small hyperintensities appeared in a cluster pattern, particularly in the white matter, suggesting multiple demyelinating lesions. The corpus callosum was atrophic (Figure 2). This MRI was evocative of multiple sclerosis or acute disseminated encephalomyelitis, but it could not explain the hearing loss. In addition, cerebrospinal fluid analysis was not evocative of these diagnoses. Considering the persistent headache, slight neurological signs, SNHL, and electrophysiological and MRI abnormalities, a fundoscopy was performed with fluorescein retinal angiography, despite any visually related complaint. Retinal angiography revealed bilateral focal arteriolar occlusions, with retinal ischemia, arteriolar shunts, and small vascular dilatations (Figure 3). This triad of hearing loss, ophthalmic microangiopathy, and encephalopathy was evocative of Susac's syndrome. High doses of intravenous steroids were administered, followed by oral steroids. After the acute stage, salicylate (150 mg per day) was administrated to improve microvascular blood supply and calcium channel blockers were prescribed (nimodipine, 30 mg, three times per day) for their antivasospastic properties. Because Susac's syndrome occurs predominantly in women, and the patient developed this triad after giving birth, we preferred avoiding any additional hormonal treatment. After 6 months of treatment, retinal angiography showed obvious improvement in the arteriolar occlusions. However, the hearing loss did not improve, and hearing aids were fitted to both ears. Clinical follow-up at 6 months and one year showed no recurrence of the symptoms, and the patient had satisfactory hearing function.

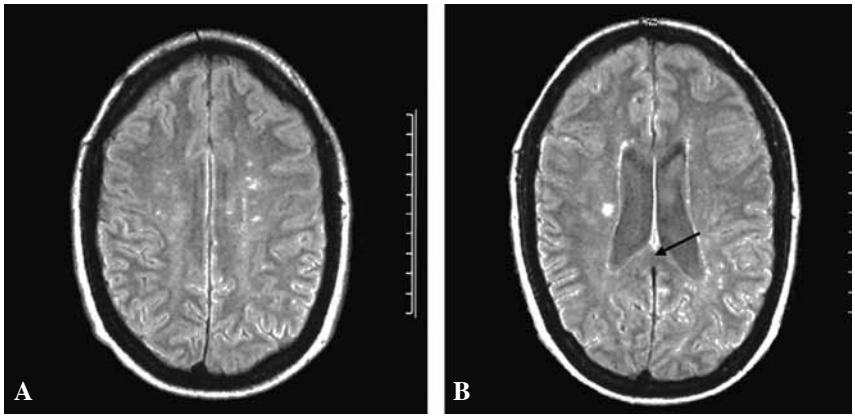

Figure 2
Fluid-attenuated inversion-recovery sequences of magnetic resonance imaging. A: Small hyperintensities appeared in a cluster pattern, especially in the white matter, and are described as multiple demyelinating lesions. B: The arrow shows right atrophy of the corpus callosum. This MRI was evocative of multiple sclerosis or acute disseminated encephalomyelitis.

Discussion

Sudden hearing loss is a deceptively simple concept, yet it defies strict definition. The most commonly used definition is a 30 dB loss over three contiguous frequencies within 3 days.[1-3] Because sudden hearing loss encompasses definite etiologies, as well as idiopathic causes, sifting through the myriad possibilities is a frustrating task.[2-4]

Susac's syndrome is a rare neurological disorder characterized by microangiopathy involving the brain, cochlea, and retina, and its etiology is elusive.[5] The most accepted hypotheses are immune-mediated pathogenesis,[7,9] infectious cause,[8] or a vascular disorder.[7-8] The exact prevalence is difficult to establish because Susac's syndrome is probably misdiagnosed. Only fewer than one hundred cases have been reported in the literature.

Our case is the first case of Susac's syndrome occurring after pregnancy. The association of pregnancy with inflammatory or immune-mediated diseases is particularly noteworthy in the literature. However, no cases of pregnancy in women with Susac's syndrome have been reported.

As in this case, Susac's syndrome affects predominantly middle-aged women.[7,9] No case of hereditary predominance has been described until now. The mode of onset and sequence of organ involvement are variable. Similar to the previous cases described in the literature, the hearing loss in our patient was sudden, bilateral, fluctuating, and asymmetric.[11] Otological manifestations likely result from apical cochlear arteriole occlusions, leading to the typical involvement of the low and medium frequencies.[6-8,13,14] Neurological and ophthalmological involvement must be investigated in the case of a rapidly progressive, often fluctuating, bilateral SNHL. Given the variability of ocular and vestibuloauditory dysfunction and neurological disease, Susac's syndrome should be suspected whenever close temporal association between hearing impairment and ocular or neurological symptoms exists. Neurological manifestations result from the multifocal microangiopathy affecting the brain. In our case, clinical signs revealed impairment of the corticospinal tract. Migraine-like headache is often a prodromal symptom that can occur several months before the onset of encephalopathy.[6-11,13] The severity of impaired cognition, memory loss, and clinical signs are variable. The change in mental status develops over several weeks.[9,11,15] MRI is often necessary to establish a diagnosis. During the acute stage, the MRI shows multiple white and grey matter lesions in the brain, especially in the corpus callosum, as well as the basal ganglia, middle cerebellar pedoncule, and cerebellar hemispheres.[7,8,12,13,15] Involvement of the corpus callosum suggests Susac's syndrome. Brain abnormalities on MRI could disappear after the acute stage.[15] In the course of the chronic stage, atrophy of the brain and the corpus callosum can lead to a residual corpus callosum,[9] which is responsible for variable symptoms of callosal disconnection.

The retinal artery occlusions lead to visual disorders, including scotoma or segmental visual field defects. Degrees of severity depend on the number and site of vessel occlusions and the presence of collateral arterioles.[15] Occlusions in posterior pole vessels can lead to profound visual loss, whereas peripheral arteriolar occlusions could be asymptomatic.[7,9,11] The key to the diagnosis of Susac's syndrome is the identification of retinal microangiopathy by fluorescein retinal angiography.[5,9]

The clinical course is usually self-limited and may improve

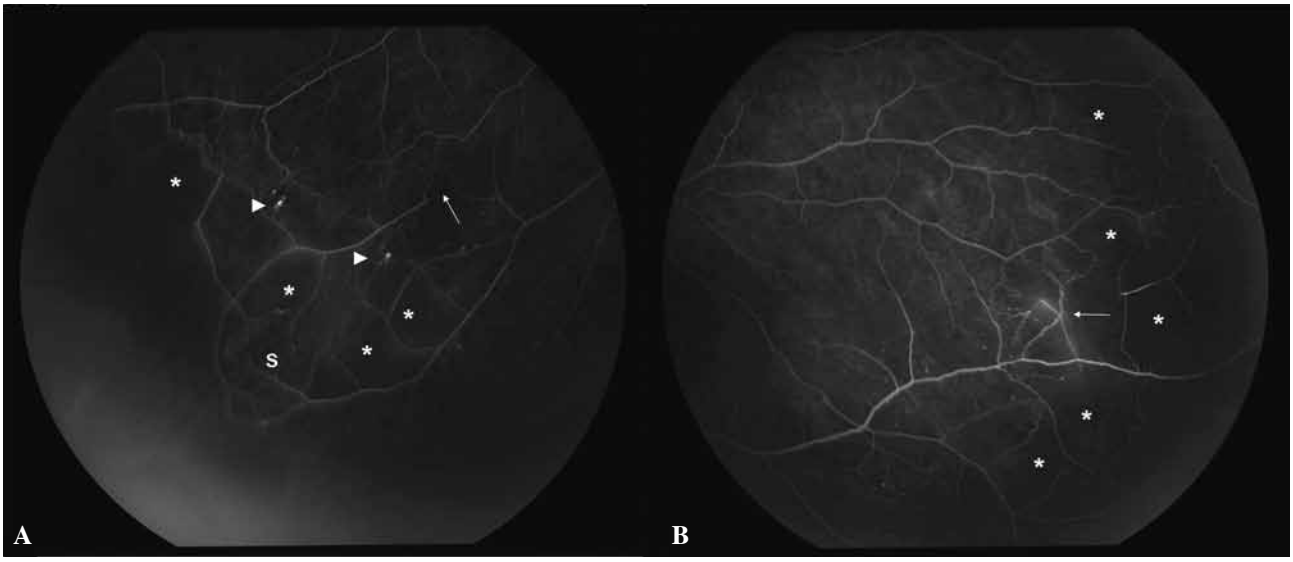

Figure 3
A: Fluorescein angiogram shows an infero-temporal arteriolar occlusion (arrow) with retrograde filling of the occluded arteriole, shunts (S), retinal ischemia (asterisk), and vascular dilatations (arrowhead) in the right eye. B: Fluorescein angiogram shows temporal microangiopathy with retinal ischemia (asterisk) and hyperfluorescence of the vessel wall (arrow) in the left eye.

spontaneously.[7,9,15] Outcome ranges from complete recovery to the association of deep deafness, blindness, and neuropsychiatric disorder. The progression or the reversibility of hearing loss is variable, but early diagnosis and treatment is known to reduce sequelae.[11] Papo *et al*.[15] reported that half of patients return to their normal lifestyle. The therapeutic approach is currently subject to discussion, and no proven or widely accepted treatment regimen is available for Susac's syndrome.

Given the hypothesis of immune-mediated disease, the usual treatment during the acute stage is high intravenous doses of steroids followed by oral steroids.[5] Cyclophosphamide, plasmapheresis, and intravenous immunoglobulin should be added when the clinical course does not improve or if it worsens.[7,9,13] Autoimmune mechanisms are still controversial, and symptoms may worsen after the steroid treatment.[8,11] In our case, immunosuppressive therapy failed to improve the patient's hearing. This lack of response may be related to the intensity of the hearing loss.

Given the hypothesis of vascular physiopathology, aspirin and anticoagulants have been used to improve the microvascular blood supply.[15] Calcium channel blockers, such as nimodipine, have been recommended for their antivasospastic properties.[14,15] Wildemann *et al*.[14] described improved hearing function and the regression of MRI and angiographic lesions. Most of the cases described in the literature have shown partial hearing improvement after treatment.[10,11]

Conclusions

Sudden hearing loss can be the first demonstration of a neurological syndrome. Susac's syndrome is an idiopathic microangiopathy characterized by the triad of hearing loss, visual loss, and encephalopathy. The syndrome predominantly affects women, and all three elements of the triad may not be present at the beginning of the disease. Patients with SNHL should be asked about ocular and neurological symptoms and immediately referred to an ophthalmologist. Retinal angiography plays a pivotal role in making the diagnosis. A FLAIR sequence of MRI must be used in all young people with hearing loss and neurological trouble. As the natural history and pathogenesis of Susac's syndrome are poorly defined, optimal management guidelines have not yet been established. Prompt recognition and treatment may reduce neurological, visual, and hearing sequelae.

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