



Case Report

A Rare Cause of Hearing Loss: Susac Syndrome

Büşra Yurtsever, Murat Çabalar, Hakan Kaya, Betül Tuğcu, Zahide Mine Yazıcı, Vildan Yayla

Clinic of Neurology, Bakırköy Training and Research Hospital, İstanbul, Turkey (BY, MÇ, VY)
Clinic of Ear Nose Throat, Bakırköy Training and Research Hospital, İstanbul, Turkey (HK, ZMY)
Clinic of Ophthalmology, Bakırköy Training and Research Hospital, İstanbul, Turkey (BT)

Susac's syndrome is a rare autoimmune disease, which is characterized by microangiopathic changes that affects the brain, retina, and cochlea. It is mainly characterized by asymptomatic cerebral infarctions, low-and mid-frequency sensorineural hearing loss, and bilateral distal retinal artery occlusions. Otolaryngologists should be familiar with Susac's syndrome because hearing loss may be the initial presenting symptom. The recommended treatment options vary from antithrombotic to immunomodulatory drugs. Although in its early stage, remission from the disease or self-limiting clinical course may be observed. Residual disabilities such as blindness, deafness, and dementia may also be presented in its late stages. Awareness of the condition and suspicion in selected patients will provide early diagnosis and treatment, which are both important to prevent the development of severe sequelae.

KEYWORDS: Encephalopathy, retinopathy, hearing loss

INTRODUCTION

Susac's syndrome (SS), first described by Susac et al. ^[1], is a rare autoimmune disease characterized by retinal, cochlear, and brain microangiopathy. The classic clinical triad consists of acute or subacute encephalopathy, branch retinal artery obstruction, and hearing loss without prominent systemic manifestations ^[2]. SS frequently affects young women (20–40 years of age), and its pathogenesis is not yet clear. Recent findings suggest that the pathogenesis is related to pre-capillary arteriole obstruction of the brain, retina, and inner ear because of damage from circulating anti-endothelial cell antibodies.

Sensorineural hearing loss (SNHL) in SS generally affects low and medium frequencies, suggesting that this manifestation is mainly caused by microinfarcts in the apical cochlea. The syndrome should be differentiated from a number of other diseases, including multiple sclerosis (MS), acute disseminated encephalomyelitis (ADEM), or systemic vasculitis ^[3]. Bilateral multiple retinal artery occlusions on fundoscopic examination or fluorescein angiography and SNHL typically involving low and medium frequencies are observed on audiological evaluation. Magnetic resonance imaging (MRI) findings include T2-weighted hyperintense small lesions in the corpus callosum and both grey and white matter of the infratentorial structures ^[2, 3]. We presented a case with SS because of its rarity.

CASE PRESENTATION

A 20-year-old female patient presented with headache, dizziness, hearing, and vision loss. Her medical history revealed treatment at an otolaryngology clinic for sudden hearing loss in the left ear 9 months ago. The following therapy regime was administered: methylprednisolone 1 mg/kg/day (Prednol®, Mustafa Nevzat, İstanbul, Turkey) for 14 days in decreasing dosage; 10 g of dextran and 0.9 g of NaCl/100 mL 500 mL/day (Rheomacrodex®, Eczacıbaşı-Baxter, İstanbul, Turkey) for 5 days; 20 mg of trimetazidine dihydrochloride (Vastarel®, Servier ilaç, İstanbul, Turkey) 3 times a day for 30 days; and hyperbaric oxygen therapy (2.5 atm, 100% O₂) once a day for 20 days. Partial improvement of hearing loss was obtained. After 5 months, the right ear presented with new beginning hearing loss. Also, the same sudden hearing loss treatment was administered, except hyperbaric oxygen therapy. After treatment, there was no improvement of SNHL in the right ear.

In addition, 4 months ago, ophthalmic examination was performed after the patient experienced sudden visual loss in both eyes. Fundoscopic examination revealed bilateral leaks due to retinal vessels vasculitis, and peripheral retinal ischemic areas related to occlusive vasculitis were detected on fundus angiography. Photocoagulation therapy was also performed.

On neurological examination, the patient was conscious, cooperative, and well-orientated, the muscle tone and muscle strength natural. The Romberg test was positive, and cranial nerve examination was normal. On ophthalmologic examination, visual acuity

This study was presented at the 7th Cochlear Implantation Otology and Neurotology and Audiology Congress, 5–8 September 2013, Gaziantep, Turkey.

Corresponding Address:

Zahide Mine Yazıcı, E-mail: minezahide@yahoo.com

Submitted: 27.02.2015

Revision received: 13.06.2015

Accepted: 15.06.2015

Copyright 2015 © The Mediterranean Society of Otology and Audiology

was found to be 6/10 in the right eye and 7/10 in the left eye. Fundoscopic examination revealed the expansion of the retinal vessels due to stasis, bilateral macular edema, and scars of the previously performed laser photocoagulation (Figure 1). In the otoscopic examination of both ears, the tympanic membrane was intact. Gadolinium-enhanced brain and ear MRIs and brain MR angiographic examination were normal. Carotid and vertebral artery Doppler ultrasonographic examination viewed the left vertebral artery as dominant. Transthoracic echocardiography was normal. Infectious and vasculitis markers were unremarkable. Cerebrospinal fluid (CSF) analysis showed a normal opening pressure and slight increase in protein concentration (38.8 mg/dL), without evidence of oligoclonal bands and normal Ig-G index (0.53). On audiological examination, the average hearing level was found to be 90 decibels in the right ear and 40 decibels in the left ear (Figure 2).

Based on these findings, the patient was admitted to the neurology clinic prediagnosed with SS and treated for 14 days with methylprednisolone 1 mg/kg/ day, followed by tapering. In the follow-up 2 weeks later, the neurological examination was unremarkable, except for ataxia. Eye examination showed full visual acuity due to regression of edema in the macula, and no additional pathology was found in the fundoscopic examination. No improvement in hearing was found in the audiological examination. Hearing aids were recommended to the patient. Informed consent was signed from patient for sharing his information.

DISCUSSION

Susac's syndrome is a rare disease characterized by non-inflammatory microangiopathy of the retina, brain, and cochlea. Because of the non-specific clinical findings, diagnosis is difficult and therefore delayed. The major clinical findings include headaches, dementia, hemiplegia, seizures, cranial nerve palsy, hearing loss, vertigo, tinnitus, and decreased or loss of the vision [4]. Generally, the first affected organ is the brain (45%), with 18% of retinal involvement during the initial phase of the disease. A total of 97% of patients do not have the classic clinical triad during the onset of symptoms. A high index of suspicion must be present before making a diagnosis because the triad may become complete after a delay of weeks to more than 2 years [5]. In particular, hearing loss is easily confused with idiopathic sudden hearing loss. Visual loss is generally due to localized retinal vasculitis rather than embolism, usually not at bifurcations [6]. Radiological examination was crucial for diagnosis. Typical MRI findings consist of small (3–7 mm) multifocal lesions. Snowball-like lesions in the center of the corpus callosum are very typical for SS. The lesions are most frequently observed in the centrum semiovale, subcortical regions, and periventricular areas [7]. Hearing loss can be a cause of serious and dramatic disability. The typical SNHL in SS, generally affecting low and middle frequencies, is mainly caused by microinfarcts in the apical cochlea and may be the first and only clinical feature of the disease [2]. Audiological symptoms are present in 22% of patients during onset. Hearing loss is fluctuated in 45% of patients and one-sided in 52% of patients [5].

Many diseases occur in the differential diagnosis. MS, ADEM, central nervous system vasculitis, and infectious encephalitis are the most common. Cerebral autosomal-dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL); Creutzfeldt–Jakob

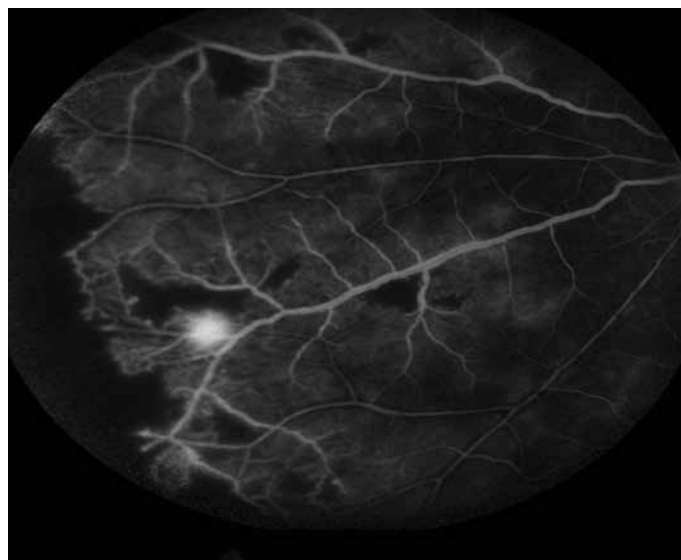


Figure 1. Fundoscopic examination revealed the expansion of the retinal vessels due to stasis

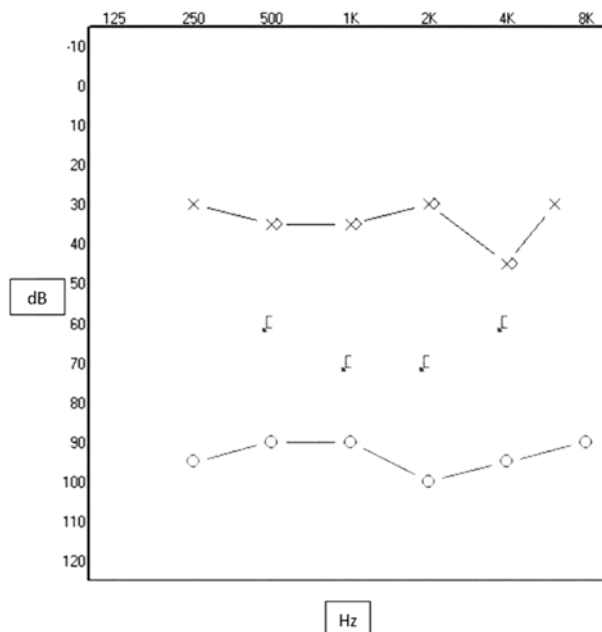


Figure 2. Audiological examination revealed that the average hearing level was found to be 90 decibels in the right ear and 40 decibels in the left ear (The symbol O stands for the right ear, the symbol X, represents the left ear)

disease; stroke; malignant tumors; migraine with aura; psychoses; and mitochondrial encephalomyopathy, lactic acidosis, and stroke-like episodes (MELAS) should also be considered in the differential diagnosis [3, 5].

Meniere's disease, Cogan's syndrome, idiopathic sudden hearing loss, viral diseases, autoimmune inner ear disease, and acoustic tumors should be considered in the differential diagnosis of the otologic involvement [2]. In our case, the imaging and laboratory findings performed for differential diagnosis were within normal limits. Cogan syndrome particularly occurred in the differential diagnosis because of the similar clinical symptoms, including sudden hearing loss and vi-

sual and neurological findings, but it was excluded because of the lack of systemic vasculitis and interstitial keratitis findings in the ophthalmic examination. In our case, ophthalmic and otologic findings were prominent without the availability of the brain involvement findings.

SS is generally self-limiting and monophasic, sometimes wavy, and rarely show recurrences in its clinical course ^[4]. In the literature, a small number of cases showing spontaneous resolution have been reported ^[6]. In addition, cases with progressive course resulting in death are also reported ^[8]. Similar to many autoimmune diseases, steroids are also used in the treatment of SS. In severe cases, intravenous immunoglobulin (IVIG) may be used. Immunosuppressants such as cyclophosphamide, mikofenolat mofetil, or rituximab can be added to steroids and IVIG treatment in patients with severe and possible sequelae ^[9]. Cochlear implant as a surgical treatment option is recommended in patients showing progressive hearing loss ^[10]. Our patient was treated with oral corticosteroids, and regression of the symptoms was evaluated during follow-up controls. Hearing aids were recommended.

As a result, SS is a rare syndrome consisting of neurological, audiological, and ophthalmological manifestations. Detection of early suspicious signs for diagnosis and early initiation of treatment is important in reducing the probable sequelae.

Informed Consent: Written informed consent was obtained from the patient who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - B.Y., M.C.; Design - M.C., H.K.; Supervision - Z.M.Y., V.Y.; Funding - H.K.; Materials - M.C., B.T.; Data Collection and/or Pro-

cessing - B.Y., H.K., B.T.; Analysis and/or Interpretation - Z.M.Y.; Literature Review - B.Y.; Writing - H.K., Z.M.Y., B.Y.; Critical Review - V.Y.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

REFERENCES

1. Susac JO, Hardman JM, Selhorst JB. Microangiopathy of the brain and retina. *Neurology* 1979; 29: 313-6. [\[CrossRef\]](#)
2. Lavinsky L, Scarton F, Lavinsky-Wolff M, Lavinsky J, Successful L H. Cochlear implantation in a Susac syndrome patient. *Braz J Otorhinolaryngol* 2012; 78: 123. [\[CrossRef\]](#)
3. Rennebohm R, Susac JO, Egan RA, Daroff RB. Susac's Syndrome--update. *J Neurol Sci* 2010; 299: 86-91. [\[CrossRef\]](#)
4. Ozturk A, Degirmenci Y, Tunc M, Kecci H. Susac Syndrome: A case of simultaneous development of all three components of triad. *J Neurol Sci* 2013; 324: 187-9. [\[CrossRef\]](#)
5. Aubart-Cohen F, Klein I, Alexandra JF, Bodaghi B, Doan S, Fardeau C, et al. Long-term outcome in Susac syndrome. *Medicine (Baltimore)* 2007; 86: 93-102. [\[CrossRef\]](#)
6. Petty GW, Matteson EL, Younge BR, Donald TJ. Recurrence of Susac syndrome (retinocochleocerebral vasculopathy) after remission of 18 years. *Mayo Clin Proc* 2001; 76: 958-60. [\[CrossRef\]](#)
7. Susac JO, Murtagh FR, Egan RA, Berger JR, Bakshi R, Lincoff N, et al. MRI findings in Susac's syndrome. *Neurology* 2003; 61: 1783-7. [\[CrossRef\]](#)
8. Saux A, Niango G, Charif M, Morales R, Mura F, Bonafe A, et al. Susac's syndrome, a rare, potentially severe or lethal neurological disease. *J Neurol Sciences*. 2010; 297: 71-3. [\[CrossRef\]](#)
9. Rennebohm RM, Susac JO. Treatment of Susac's syndrome. *Curr Treat Options Neurol* 2008; 10: 67-74. [\[CrossRef\]](#)
10. Grover N, Whiteside OJ, Ramsden JD. Susac syndrome: outcome of unilateral cochlear implantation. *J Laryngol Otol* 2011; 125: 856-8. [\[CrossRef\]](#)