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CASE REPORT

Susac Syndrome in a Girl Diagnosed with MS: A Case Report

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ABSTRACT
Susac syndrome (SS) is a rare disease attributed to a microangiopathy involving the arterioles of the brain, retina and cochlea. It was first described by John o susac in 1979 in two young women presenting with the classic clinical triad of subacute encephalopathy, retinal arteriolar branch occlusions and sensory-neural hearing loss. Susac syndrom can mimic several diseases. Differential diagnosis include demyelinatig disease, connective tissue disease, infection, neoplasm, procoagulant state and ischemic diseases. Rapid diagnosis is important for prompt therapy. In this article we present a patient whom she was treated on Multiple Sclerosis and finally the diagnosis of SS was established for her.
Keywords; *Susac syndrome, Multiple sclerosis, Demyelinating disease, Retinal arteriolar branch occlusion*

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INTRODUCTION

Susac syndrome (SS) is a rare disease of unknown etiology, which is clinically characterized by the triad of encephalopathy, visual and auditory loss. It can mimic several disorders. Its pathogenesis is not clearly defined yet but it seems micro angiopathy of the brain artrials, cochlea and retina results in the clinical triad of disease [1]. The diagnosis can be established by MRI findings, fluorescein angiography and audiometry accompanying clinical findings. The differentiation of acute disseminated encephalopathy (ADEM) and multiple sclerosis (MS) is particularly difficult [2].

CASE REPORT

In April 2014, a 22 years old female was admitted to our hospital with bilateral visual impairment. The patient was previously diagnosed with MS. She had no complaint of vertigo or unsteady gait. For the first time, her neurological symptoms were noted on 2010 with ataxia, vertigo and paresthesia in her lower limbs. At that time, her brain MRI showed extensive signal abnormalities in both periventricular white matters, thalami, cerebellar hemispheres with extensive Parenchymal enhancement. So diagnosis of MS was confirmed and she was treated by intravenous methylprednisolone and Interferon beta.

At this time, general physical examination, sensory and motor examination was unremarkable. On ophthalmic examination, her best corrected visual acuity was counting finger in both eyes. Visual field test showed bilateral temporal defect. Fundus examination showed significant swollen optic disc and splinter hemorrhage suggesting of branch retinal arterial occlusion. Laboratory findings were normal (table 1).

Table1- laboratory tests of the patient

Lab tests	Result	Normal Range
WBC	5700	4000-7000
HB	13.6	12-15
PLT	217000	140000-450000
ESR	10mm/HR	
CRP	<2	Up to 8 mg/l

RF	10	Up to 20 LU/ml
Angiotensin converting enzyme	46	8-65u/l
Pro C	98	70-150%
Pro S	126	55_160%
Ig A	80	70_400 mg/dl
Ig G	754.9	700_1600 mg/dl
C3	0.95	0.9_1.8 g/l
C4	18	10_40: mg/dl
Anti B2 micro globulin Ab (IgG)	4.4	positive> 8: u/ml
Anti B2 glycoprotein Ab (IgM)	4.2	positive> 8 u/ml
Burrellia burgdoferi Ab (Ig M) and (Ig G):	Negative	
Aquaporin 4 Ab (NMO)	Negative	

Brain and cervical MRI showed several merged and confluent periventricular and peri-callusal hyper intensities in both centrum semi oval, typical involvement of corpus callosum, temporooccipital lobe white matter.(fig 1-A). Considering these findings and suspecting MS relapse, patient received methylprednisolone. However, after 7 days of treatment there was no clinical improvement. in further history taking, left ear hearing loss was recognized which was started 2-3 months earlier.Audiometry showed left ear sensory neural hearing deficit in low and mid frequencies.

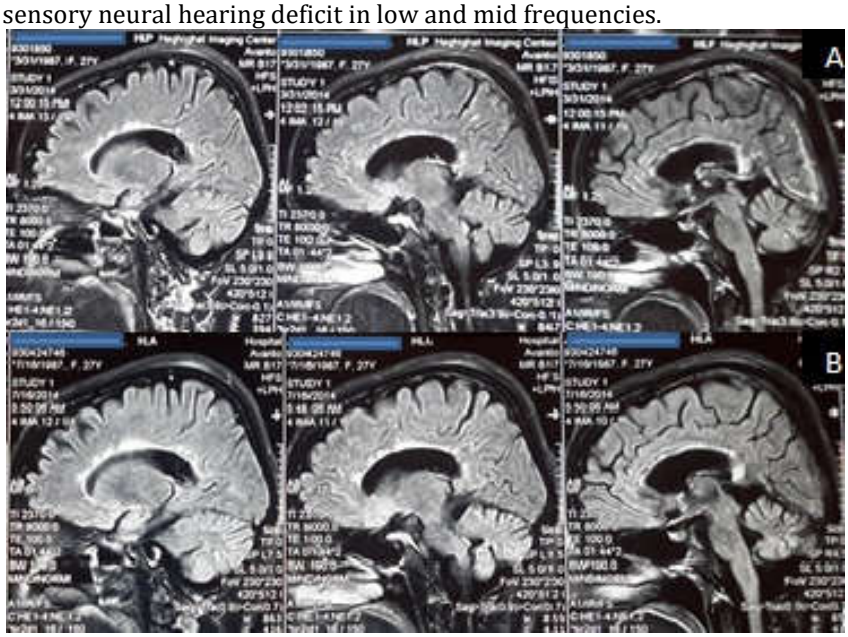

Fig 1- Sagittal T2-weighted MR image shows hyper intense of the corpus callosum. A: march2014 – B: four month later

Bilateral carotid artery color Doppler sonography showed normal with no significant stenosis.

Cervical MRA (magnetic resonance angiography) and MRV (magnetic resonance venography) showed normal.

Paranasal sinus, skull base and chest CT scan were unremarkable.

Fluorescein angiography showed focal arteriolar narrowing with areas of occlusions.

So the typical clinical hearing loss and retinal abnormalities, accompanied by the findings of MRI, confirmed the diagnosis of Susac syndrome.

So the treatment started by intravenous IVIG, Prednisolone 50 mg/day and Azathioprine 20 mg/day. Within 2 weeks left eye corrected visual acuity and visual field improvement were noted, but there was no recovery in her right eye visual acuity. Repeated MRI showed no new lesions. After discharge from hospital, predinisolone was tapered and she was prescribed Azathioprine 20 mg/d and monthly IVIG. On 1 year follow up, no new attack and no progression in hearing loss was noted. On examination her left eye corrected visual acuity was 9/10 with no visual field defect. Her brain MRI also had no new lesions (fig 1-B).

DISCUSSION

Susac syndrome is a rare clinical entity presenting with the typical triad of encephalopathy, hearing loss and visual disturbances.

Most cases of Susac syndrome are reported in young, healthy women, aged 18_40 years. No familial case has been reported [1, 2]. The etiology and pathogenesis of SS remains unknown and no procoagulant state or definite connective tissue disorder or vasospastic arterilar occlusion or micro embolization has been consistently reported [3-5].The disease frequently misdiagnosed as MS, Migraine, Lupus Erythematosus, Encephalitis, Meniere disease, thromboembolic stroke and even Schizophrenia [3, 4]. It is nessesery to exclude other entities. Characteristic findings on brain MRI and MRV, ophthalmic examination and audiogram can facilitate the diagnosis. A high index of suspicion must be present because a high percentage of patients do not have the clinical triad at the time of onset of symptoms(7, 8).Findings in MRI include multiple small foci of high T2 signal intensity and contrast enhancement throughout both gray and white matter in the cerebrum and infratentorial structures.corpus callosum is always involved [6-8].

Central callosal holes ensue as the active lesions resolve. In contrast to Susac syndrome, the callosal involvement in both MS and ADEM is on the undersurface of the corpus callosum at the septal interface. As encephalopathy abates, white matter lesions typically disappear, but atrophy becomes evident. The cranial nerves are intact. The hearing loss and vertigo are due to cochlear involvement and semicircular canal involvement respectively. There is a form fruste of the disease in which recurrent branch retinal artery occlusions and hearing loss occur in the absence of encephalopathy. Even in these cases, MR imaging may shows white matter changes, especially in the corpus callosum (6, 7).

The encephalopathy may be acute or sub-acute and is frequently seen with psychiatric features of personality change and bizarre behavior. It may be preceded by headache, often with migrainous features as a prominent symptom. The retinopathy is characterized by multiple peripheral retinal arteriolar branch occlusions that can be seen on ophthalmoscopy examination or retinal fluorescein angiography. Segmental loss of vision in one or both eyes and visual scintillating scotoma are typical complaints. Hearing loss is often acute, unilateral or bilateral and associated with tinnitus, vertigo, nausea, nystagmus and unsteady gait [7, 8]. Careful questioning may be necessary to obtain evidence of hearing loss [6-9].

Early recognition and appropriate therapy of this syndrome may reduce the permanent sequels. Treatment of SS remains largely based on anecdotal evidence. Various treatments have already been used: Corticosteroids, Cyclophosphamide, antiplatelet, antithrombotic agents, intravenous immunoglobulins, plasma pheresis, and hyperbaric oxygen [9].

We presented this case to emphasize that we should suspect Susac syndrome in any adult or young patients presenting with a sub-acute encephalopathy which are not common in multiple sclerosis, with or without visual or auditory symptoms and high T2 lesions at brain MR imaging. It appears that in patients in whom diagnosis and treatment are delayed, permanent morbidity is higher in terms of neurologic deficit, visual loss, and hearing loss. Rapid and early therapy should reduce sequels of this disease. Our patient has a good recovery following and appropriate therapy.

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