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Susac Syndrome in A Primigravida: A Rare Case Report

[*]Bhattu S R[1],Talib S.H.[2],Soni Anand[3],Deshmukh Shridhar[4],Vyawahare Suraj[5], Gunthe Sudarshan[6],Tamboli Gaurav[7],Agrawal Nishant[8]

[1]*Associate Professor, Department of medicine, MGM Medical College, Aurangabad, 431003, India*
[2]*Professor & Head, Department of medicine, MGM Medical College, Aurangabad, 431003, India*
[3]*Assistant Professor, Consultant Neurologist, MGM Medical College, Aurangabad, 431003, India*
[4,5,6,7,8]*Post Graduate Residents in Medicine, MGM Medical College, Aurangabad,431003, India*
Corresponding Author: Bhattu S R

Abstract: *Susac's syndrome is an uncommon neurologic disorder of unknown cause. It has been described as a clinical triad of encephalopathy, sensorineural hearing loss, and branch retinal artery occlusions (BRAO). Susac's syndrome is presumed to be an autoimmune endotheliopathy. We present a case of 21 year old 12 weeks primigravida female patient who presented with visual loss, sensorineural hearing loss, altered sensorium & missed abortion. A high index of suspicion leading to correct diagnosis and early appropriate therapy may reduce the permanent sequel seen with this disease. Because of its rarity and some similarities to other common neurological conditions such as multiple sclerosis and acute disseminated encephalomyelitis, it is often misdiagnosed and therefore mistreated. In patients in whom diagnosis and treatment are delayed, permanent morbidity is higher in terms of visual loss, hearing loss, and neurologic debility. In patients in whom rapid diagnosis has led to early administration of immunosuppressive therapy, recovery can be almost complete.* ***Key Keywords:*** *BRAO, Encephalopathy, Primigravida, Sensorineural hearing loss, Susac's syndrome*

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I. Introduction

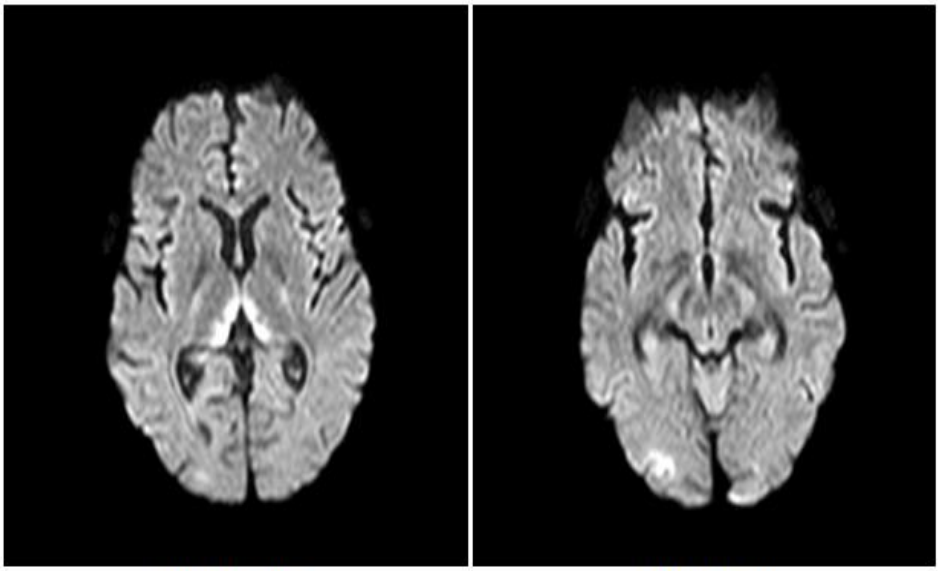
Susac's syndrome (SS) is a clinical triad of encephalopathy, branch retinal artery occlusion (BRAO) and sensory neural hearing loss (SNHL), first described by Susac.[1] He first reported two cases of brain and retinal vasculopathy in female patients with good response to corticosteroids in 1979.[2] It is a rare syndrome and, until now, slightly more than 300 cases have been reported in the literature.[3] SS is frequently seen in females of 20-40 years of age.[4,5] It usually presents with severe headache and behavioural changes, progressive cognitive decline, apathy and later by hearing loss, tinnitus and segmental visual loss.[4] The clinical triad of subacute encephalopathy, BRAO, and hearing loss is due to a precapillary arteriolar angiopathy of unknown origin, but most evidences are in favour of an immune-mediated endotheliopathy, and immunosuppressive therapy is the main mode of treatment.[6,7]

II. Case Report

A 21 year old female primigravida with 3 months amenorrhoea came to our hospital with chief complaints of 1) Diminution of vision since 8 days which worsened over last 4 days, 2) hearing loss since last 4 days 3) PV spotting since 2 days , was admitted under gynaecology ward for missed abortion. Fundus examination revealed B/L retinal edema along with B/L central retinal artery insufficiency. Audiometry was-s/o sensorineural hearing loss(SNHL). Patient was posted for suction & evacuation due to missed abortion which was done on 3rd day of hospitalisation. Post-op day 1, patient developed altered sensorium with dyskinetic movements & rise in blood pressures, in view of developing encephalopathy, patient was shifted under Medicine in medicine ICU. Patients neurological symptoms worsened with altered sensorium, drowsiness, dyskinetic movements, photophobia, apathy, aphasia associated with progressive loss of vision to the extent that only perception of light was possible, sensorineural deafness & hypertension.

Vitals: Pulse-120/min., BP-150/100mmHg, CNS examination: power 5/5 in all four limbs, DTR+, plantars b/l flexors. Lumbar puncture was done for CSF s/o raised proteins (CSF-Glucose=62mg/dl, Protenis=316mg/dl, Total WBC count=04 cells/cumm=all lymphocytes)

MRI Brain (P) was done s/o acute ischemic changes in B/L median thalamic region (Fig. A) & right occipital lobe (Fig. B)



(DWI Fig. A) (DWI Fig. B)

Fundus fluorescein angiography (FFA) was done s/o leakage from the inflamed retinal arterioles but did not reveal BRAO.

ANA, dsDNA was negative. Renal function tests, Liver function tests were within normal limits. Hemogram s/o Hb-10.9gm%, TLC-10,690/cmm, Platelets-1,84,000/cmm was within normal limits. Urine Albumin-Trace, Na-132, K-2.6 Patient was started with Pulse Inj. Methyl Prednisolone 1gm IV OD for 5 days considering the clinical possibility of Susac Syndrome i/v/o rapidly progressive visual loss, hearing loss & encephalopathy. Patient was also put on antihypertensive Tab. Lobet (labetalol) 100mg TDS for controlling hypertension. Patient responded clinically with improving vision, improving hearing by 5th day of Pulse therapy. She recovered completely from encephalopathy with no residual neurological deficit by around two weeks on oral steroids & subsequently hypertension also resolved. Though the MRI findings were not typical of Susac Syndrome, fundus showed microinfarcts on retina, our patient had a clinical triad of symptoms of Susac Syndrome as encephalopathy, vision loss & sensorineural hearing loss which responded very well to steroids.

III. Discussion

Susac's syndrome was first described by Susac in 1979[1]. The disease is also known as (a)-RED-M which stands for retinopathy, encephalopathy, and deafness associated microangiopathy, (b) SICRET (small infarct of cochlear, retinal, and encephalic tissues), and (c) retinocochleocerebral vasculopathy[8]. SS is frequently seen in women with female to male ratio of 3:1, it's onset varies in age of 9-58 years, but most of the patients are 20-40 years old.[4,5] Mateen et al. in a series of 29 patients with SS reported that 83% of their patients were female with a mean age at symptomatic presentation of 35 (19-65) years.[9]

Affected patients have multiple branch retinal occlusions that typically are bilateral, progressive hearing loss, and various neurologic presentations[10]. Arteriolar occlusions usually are caused by emboli that typically are fleeting in nature[10]. If the infarctions are extensive and involve the posterior pole, the patient will complain of impaired vision. If the occlusions occur in the more peripheral portion of the retina or if the patient is encephalopathic, he may not describe visual symptoms. Hearing loss can be a dramatic and severely disabling feature of SS. It often occurs overnight and may affect both ears. A loss of low or middle frequencies is typical, but losses of high frequencies can also occur[11]. The predominance of low and medium tone loss is suggestive of cochlear apical damage caused by occlusions of the cochlear end arterioles. Loss of high-frequency tone is a marker of more significant cochlear damage[10]. The hearing loss is often accompanied by vertigo and tinnitus[11]. The hearing loss is due to cochlear involvement and vertigo if present is due to semicircular canal involvement[12]. Neurologic symptoms and signs are diffuse and multifocal, acute or sub-acute in onset and progress during the active phase of the disease. Headache was a prominent feature initially in more than one half of the patients. Most patients have abnormalities in cognition, memory, behaviour with many neuropsychiatric manifestations[13]. The encephalopathy may progress to a stage where the patient is totally unable to communicate. Seizures and myoclonus may occur[14]. Susac's syndrome is presumed to be an autoimmune endotheliopathy. In biopsies of the brain, microinfarcts with loss of neurons, axons, and myelin in the white and grey matter could be detected. The microinfarcts are caused by microangiopathic process with arteriole wall

proliferation, lymphocyte infiltration, destruction of the capillary network, and basal lamina thickening[11]. Diagnosis and management often require a multidisciplinary effort involving a neurologist, neuroopthalmologist, otolaryngologist, neuroradiologist, and rheumatologist[15]. Brain magnetic resonance (MR) imaging plays an important role in evaluation of patients suspected of having SS[16]. On MRI, the typical lesions are small multifocal lesions of 3–7 mm.

The most important diagnostic sign and very typical for SS is the snowball-like lesions in the center of the corpus callosum. At the onset of the disease, before treatment contrast enhancement around small vessels representing a perivascular leakage and leptomeningeal contrast enhancement can be seen. The most important differential diagnoses are multiple sclerosis (MS) and acute disseminated encephalomyelitis (ADEM). In MS and ADEM, the lesions are on the undersurface and at the septal interface of the corpus callosum. In SS, the lesions are lying on the centre of the corpus callosum and are sparing the periphery of it. In ADEM and MS leptomeningeal enhancement is uncommon. Spinal fluid examination has invariably shown an elevated protein in the range of 100 to 800 mg/dL and minimal pleocytosis (5–15 lymphocytes) occurs occasionally[14]. EEG shows diffuse slowing[17] while cerebral arteriography findings are almost normal, because the involved precapillary arterioles (<100 μm) are beyond the resolution of arteriography. Fluorescein angiography however is extremely useful and will often show the branch retinal artery occlusion as well as the pathognomonic multifocal fluorescence of the branch arterioles[12].

The classical triad is pathognomonic for SS, but the three elements are not always present at the same time[18]. Patients may present with isolated encephalopathy, unexplained visual disturbance, or hearing loss and go undiagnosed until a full triad is assumed[15]. Based on the hypothesis of it being an autoimmune disease, treatment has to be immunosuppressive. Considering the often severe residual deficits of brain, eye, and ear functions in these mainly young patients, treatment has to be significantly aggressive to prevent further damage and relapses[11]. Treatment with IV methylprednisolone followed by oral steroids is recommended[12]. Intravenous immunoglobulin (IV IG) can be useful in severe cases. In severe and disabling cases, further immunosuppressants like cyclophosphamide, mycophenolate mofetil, or rituximab should be added to glucocorticoids and IV IG[11]. The clinical course of SS is usually self-limited, fluctuating, and monophasic. It lasts from two to four years but may be as short as six months or as long as five years in duration[12], but the exact duration in the individual patient is unpredictable. Relapses after decades, have been described. This requires monitoring for a lifelong time[11]. Although some patients recover with little or no residual disease, others are profoundly impaired with cognitive deficits, gait disturbance, and hearing loss.

A high index of suspicion, leading to correct diagnosis and early appropriate therapy may reduce the permanent sequel seen with this disease.

IV. Conclusion

Our patient who had presented with rapidly progressive visual loss, hearing loss & encephalopathy fit in the clinical triad of Susac syndrome, though her MRI findings were not typical. Our patient responded well to steroids & is doing well till last follow up at 4 months. With the paucity of cases reported from India, we report this case to sensitize the clinicians to suspect this disease & institute appropriate & timely treatment.

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