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Immunoadsorption as a Novel Strategy in Susac Syndrome

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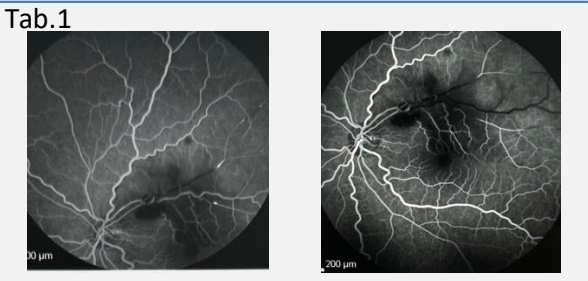
OBJECTIVES

To report the novel use of immunoadsorption (IA) in a patient with Susac syndrome (SuS) and evaluate its impact on clinical, neuropsychological, and radiological outcomes, in the context of limited therapeutic evidence for extracorporeal treatments in this rare condition.

MATERIALS

A 52-year-old woman presented with history of vague visual disturbance in right eye with **outpatience fluorescein angiography revealed bilateral branch retinal artery occlusions (BRAOs) (Tab.1)**, mild bilateral sensorineural hearing loss, intense headache and acute onset of severe vertigo, aphasia, behavioral changes, confusion, gait instability, and prosopagnosia.

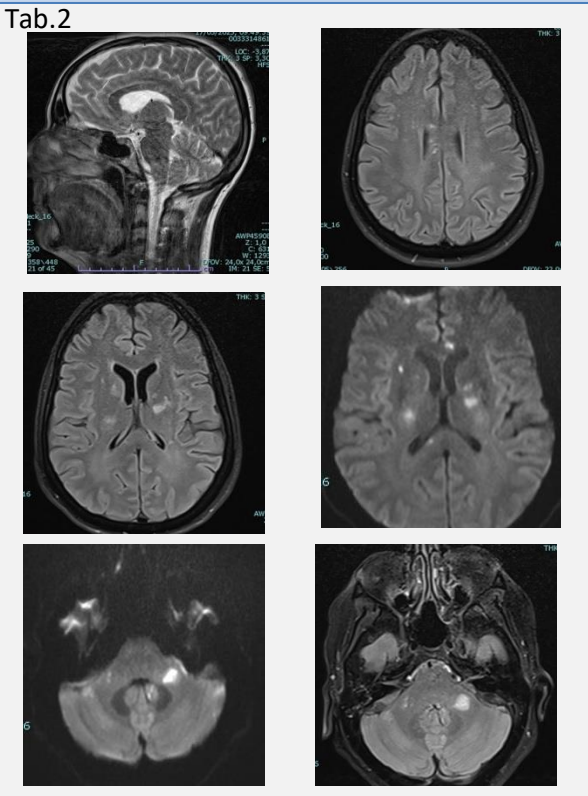
Tab.1



METHODS

Neurological examination revealed disorientation, limited and incongruent speech, ideomotor apraxia, marked perseveration, mild left-sided weakness, brisk reflexes (left > right), bilateral Babinski sign, and left-predominant dysmetria. Truncal control and gait were severely impaired. No overt sensory deficits were detected. Extensive laboratory testing ruled out infectious, metabolic, paraneoplastic and systemic autoimmune conditions. Lumbar puncture revealed mild hyperproteinorrachia and mononuclear pleocytosis. Cardiac ultrasound, total-body CT, and mammography were normal. **Brain MRI revealed multiple FLAIR hyperintensities in the corpus callosum, bilateral nucleo-capsular regions and left middle cerebellar peduncle with some callosal lesions showing restricted diffusion (Tab.2)**. ENT evaluation confirmed sensorineural hearing loss, predominantly on the left. Neuropsychological assessment revealed impairments in executive function, memory, and attention. Susac syndrome was suspected following exclusion of other diagnoses.

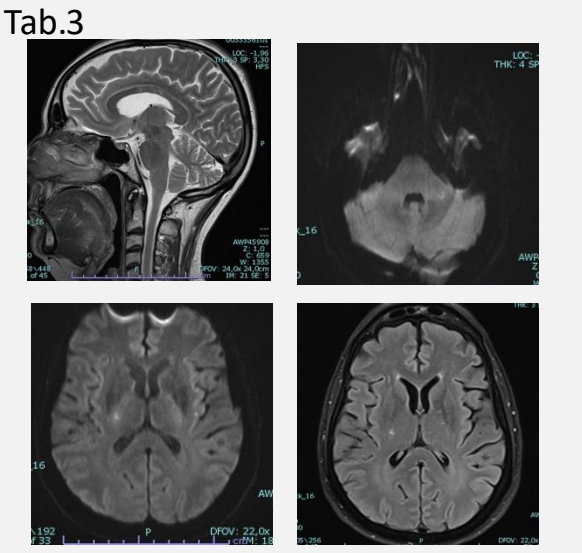
Tab.2



RESULTS

Initial treatment with IV methylprednisolone (1 g/day for 5 days) followed by high-dose oral prednisone resulted in partial clinical improvement. Due to the persistence of symptoms and the availability of therapeutic apheresis at our center, the patient underwent five sessions of immunoadsorption on alternate days. **Immunoadsorption was well tolerated and let to progressive improvement in cognitive and motor symptoms.** At discharge, the patient was alert, responsive, and behaviorally appropriate, with resolution of confusion. She regained upright posture and independent ambulation, with marked improvement in ataxia. Mild apraxia was still observed. Hearing loss and occasional vertigo persisted, while visual disturbances were infrequent and ill-defined. No motor or sensory deficits were found. **Follow-up brain MRI one month after discharge showed a reduction in lesion size with minimal residual diffusion restriction.(Tab.3)** Clinical status remained stable at subsequent evaluation.

Tab.3



DISCUSSION

Susac syndrome is a rare autoimmune endotheliopathy with a complex pathogenesis and limited evidence to guide treatment. While therapeutic plasma exchange (TPE) has been used as rescue therapy in SuS, requiring plasma replacement and carring risks of allergic reactions or infection, immunoadsorption allows selective removal of circulating immunoglobulins and immune complexes without the need for plasma substitution, offering a safer profile. Although not previously reported in SuS, IA has demonstrated efficacy in other antibody-mediated neurological conditions. In our patient, IA resulted in sustained clinical and radiological benefit.

CONCLUSION

Immunoadsorption proved to be a safe and effective therapeutic strategy in this case of Susac syndrome. This report highlights its potential as a rescue treatment in refractory cases, supporting the need for further investigation and inclusion in future treatment algorithms.

Boedecker SC, Luessi F, Engel S, Kraus D, Klimpke P, Holtz S, et al. Immunoadsorption and plasma exchange—Efficient treatment options for neurological autoimmune diseases. J Clin Apher. 2022;37(1):70–81

Rennebohm RM, Asdaghi N, Srivastava S, Gertner E. Guidelines for treatment of Susac syndrome – An update. Int J Stroke. 2020;15(5) 484–494

Akgun Y. Plasma exchange in Susac syndrome: Unlocking treatment for a rare disease. Vox Sang. 2025;1–3