

## Susac's syndrome, rare CNS vasculitis in a teenager: A case report

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Susac's syndrome is an autoimmune endotheliopathy of the microvasculature of the brain, retina and cochlea; characterised by rapid onset encephalopathy, visual loss with branch retinal artery occlusions and sensorineural deafness. A previously healthy 14-year-old girl presented in an acute confusional state. Her initial investigations including toxicology screen and inflammatory markers were unremarkable. Neuroimaging on day 4 showed multifocal bihemispheric and cerebellar white-matter high signal lesions with more confluent involvement of the corpus callosum, with restricted diffusion, suggestive of ischaemia. Her magnetic resonance angiography was normal. Cerebrospinal fluid examination revealed a white cell count of 7 cells/mm<sup>3</sup>, raised protein of 1.7g/l, with normal glucose, gram staining and culture and negative paired oligoclonal bands. Her EEG was encephalopathic. She received two pulses of methylprednisolone along with oral prednisolone with short lived improvement. She received plasmapheresis on week 3 and intravenous immunoglobulins on weeks 5, 7, 8 which resulted in transient improvement. She exhibited progressive cognitive and neurological deterioration and remained encephalopathic. Brain biopsy carried out in 9th week confirmed small vessel vasculitis. Susac syndrome was diagnosed by ophthalmic confirmation of multiple branch retinal artery occlusions and infarcts. Cyclical Cyclophosphamide and Rituximab were commenced. Further neuroimaging at 3 months shows generalised brain atrophy with confluent white matter high signal. Five months later, the patient remains conscious, communicates by switching and retains asymmetric spastic tetraplegia. Susac syndrome should be considered in young people with acute encephalopathy. Early brain biopsy to aid diagnosis and timely treatment with aggressive escalating immunomodulatory therapies may reduce morbidity.

### Biography

Aman Singh Sohal completed his MRCPCH from Royal College of Paediatrics and Child Health in 2007. He joined national training in paediatric Neurology in 2009 and is currently a resident at The Great North Children's Hospital, Newcastle-upon-Tyne. He has published papers in several peer reviewed journals.

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