A 21-year-old presented with a one-month history of confusion and headache, with collateral history suggestive of focal seizures. Examination revealed an encephalopathic patient with a bilateral internuclear ophthalmoplegia and a spastic ataxic quadraparesis. MRI demonstrated multiple non-enhancing hyperintense T2 lesions in the cerebellum, corpus callosum, grey and white matter, some with restricted diffusion. There was marked posterior fossa leptomeningeal enhancement. CSF protein was 2.32g, with 5 lymphocytes and negative oligoclonal bands. Brain biopsy showed non-specific inflammatory changes.

Extensive infective, inflammatory and malignancy screening was negative. Despite an initial clinical response to a steroid pulse, new lesions developed, accompanied by clinical deterioration.

Two months into the illness, whilst on high-dose oral steroids, he developed characteristic branch retinal artery occlusions. A diagnosis of Susac’s syndrome was made, despite normal audiometry. Treatment with aggressive immunosuppression was successful.

Susac’s (‘small infarctions of cochlear, retinal and encephalic tissue’ (SICRET)) syndrome, is a rare, presumed autoimmune, endotheliopathy. Microvascular occlusions lead to the pathognomonic triad of encephalopathy, sensorineural deafness and branch retinal artery occlusions, rarely all present at presentation. Seizures are an unusual feature. Temporal dispersion of symptoms may delay diagnosis for months in some patients. A literature review of clinical features and treatment will be presented.