Susac syndrome as a multiple sclerosis mimicer

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Susac syndrome (SS) is an autoimmune disorder characterized mainly by the clinical triad of encephalopathy, branch retinal artery occlusions (BRAOs), and sensorineural hearing loss (SNHL). Susac syndrome affects women more than men at a ratio of 3:1.The pathophysiology of Susac syndrome is still unclear; however, an immune-mediated injury involving the endothelium of retina, cochlea and cerebral vasculature is the leading hypothesis.
In cases with classical triad it can be readily distinguished from multiple sclerosis but when the other 2 components of the triad are not recognized – some clinicians fail to consider SS. Instead, a mistaken diagnosis of multiple sclerosis (MS) or acute disseminated encephalomyelitis (ADEM) is made and the patients are treated with a relatively brief course of corticosteroids. If the patient left untreated, persistant cognitive impairement can ensue.
MRI lesions occurring centrally in the corpus callosum, referred to as 'snowball' lesions and 'punched out holes' are thought to be pathognomonic for Susac's syndrome when the clinical triad is present. Retinal Fluorescein angiography is useful in evaluating BRAO. Early and Correct immunosuppressive therapy results in significant clinical and radiological improvement, and also may reduce relapses .
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