SUSAC'S SYNDROME AS A DIFFERENTIAL DIAGNOSIS OF RETINAL VASCULITIS

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BACKGROUND

A 27-year-old female patient, complaining of migraine with aura for the least 2 months, developed sudden vertigo, headache, tinnitus, ear fullness with hearing loss, and visual loss. There was no mental confusion. Fundoscopy demonstrated retinal detachment and central retinal artery branches occlusion.

CASE REPORT

Female, 27 years old, with a previous history of hypothyroidism, was admitted with hearing and visual loss, headache and vertigo. She denied fever, alopecia, arthritis, rash, livedo or weakness. Ophthalmological investigation showed campimetry with altitudinal deficit to the right, accompanied by ipsilateral visual deficit. Fundoscopy demonstrated retinal detachment and central retinal artery branches occlusion. Audiometry demonstrated sensorineural loss on the right, and with this, the Susac's syndrome triad (encephalopathy, retinal arterial occlusion and sensorineural hearing loss) was confirmed. Cerebrospinal fluid (CSF) examination was unremarkable, echocardiogram did not show vegetations, and carotid Doppler scan was normal. Imaging of central nervous system revealed a typical Susac's syndrome in CNS MRI ("snowball pattern").

CONCLUSION

Susac's syndrome is a rare entity, with less than 400 cases described worldwide. It comprises the triad of encephalopathy, sensorineural hearing loss and retinal artery occlusion. It is an immune-mediated endotheliopathy that affects the microvasculature of the brain, retina and inner ear, with antiendothelial cell antibodies playing an important role in the lesion. It may also present with myalgia, arthralgia, livedo reticularis, livedo racemosa, which makes this diagnosis of importance for the rheumatologist. It can be misdiagnosed as multiple sclerosis, disseminated encephalomyelitis, central nervous system vasculitis, and systemic lupus erythematosus. Treatment requires potent immunosuppression, with pulses of methylprednisolone associated with rituximab, cyclophosphamide or mycophenolate.

KEYWORDS

Susac's syndrome, Branch retinal artery occlusion, CNS vasculitis.

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