SUSAC SYNDROME: A CASE REPORT

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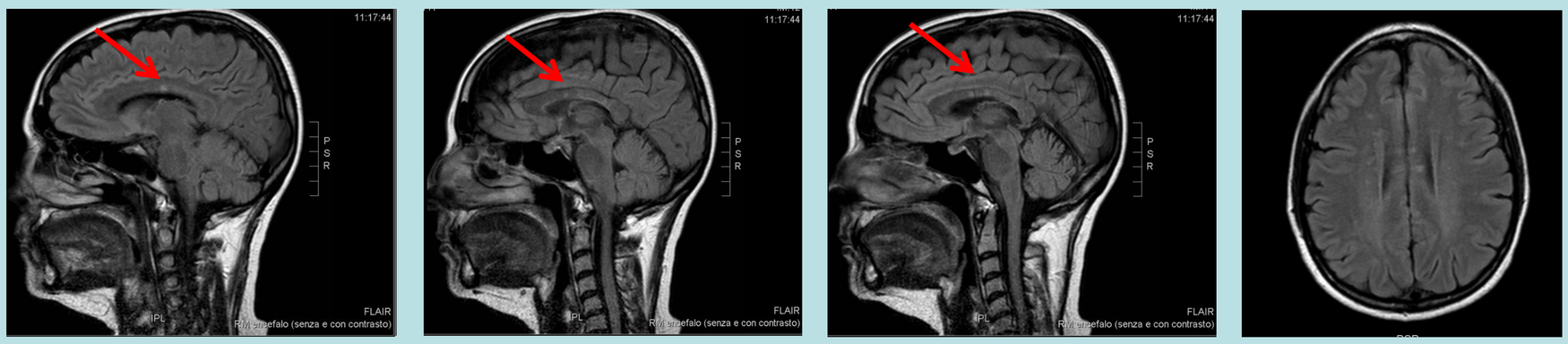
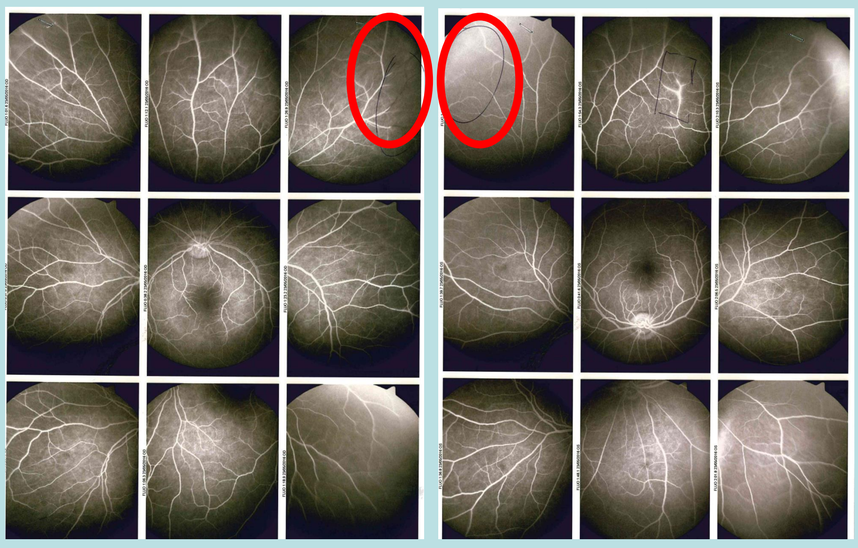
Susac syndrome is rare autoimmune microangiopathy affecting small vessels of the retina, inner ear and deep brain structures characterized typically by the triad of encephalopathy, retinopathy, and hearing loss. It affects women 3-4 times more often than men. Most individuals are 20-40 years old at presentation.

Case report

A 42-years-old woman presented in January 2016 acute confusional state and psychomotor agitation. In the next two weeks she had progressive memory loss, gait disturbance, vomiting and transient paresthesias in the limbs. A first brain MRI was completely normal. A two weeks later MRI revealed multiple hyperintense lesions on the FLAIR and T2-weighted images in the corpus callosum, cerebellum, pons.



On neurological examination at admission, she was confuse with mild memory deficit and disexecutive syndrome. Routine laboratory studies, cerebrospinal fluid, immunological and cancer screening were normal. During the recovery one focal seizure and acute hearing loss of the right ear occurred.
Audiometry confirmed perceptive hearing loss and fluorescein angiography revealed branch retinal artery occlusion (BRAO) in left eye, confirming the diagnosis of Susac syndrome.



She was treated with methylprednisolone 1000 mg/day replaced with dexamethasone 32 mg plus azathioprine, for the occurrence of severe bradycardia. After three weeks of azathioprine was suspended for liver toxicity and patient developed cognitive relapse while she was treated only with prednisone 27,5 mg/die. Intravenous immunoglobulin 2 g/kg in five consecutive days, for a total of 3 cycles, and oral cyclophosphamide (100 g/d) were prescribed. Follow-up brain MRI was almost completely normal as the cognitive status, tapering the daily dose of prednisone.

Susac syndrome is rare disease, but awareness of this condition has remarkably grown in the past several years. A high clinical suspicion must be maintained in order to diagnose as it can mimic other disorders. Rapid diagnosis is essential to ensure early immunosuppressive therapy. Cerebral magnetic resonance imaging and retinal fluorescein angiography play an important role in confirming the diagnosis.

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