Clinical Neurology and Neurosurgery 149 (2016) 64–67

Contents lists available at ScienceDirect

Clinical Neurology and Neurosurgery

journal homepage: www.elsevier.com/locate/clineuro

Case Report

Is Susac syndrome associated with bipolar disorder?

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ARTICLE INFO

Article history:
Received 5 February 2016
Received in revised form 4 July 2016
Accepted 10 July 2016
Available online 14 July 2016

Keywords:
Susac syndrome
Bipolar disorder
Corpus callosum
Magnetic resonance imaging
Psychiatric symptoms
Mood disorders

1. Introduction

Susac's Syndrome (SS) is a rare autoimmune disorder affecting the precapillary arterioles of the brain, retina, and cochlea. Young women, in particular, are at risk, but the disorder can also affect males (3.5 female/1 male) [1].

The clinical presentation is characterized by a triad of symptoms, including hearing and visual loss, and central nervous system (CNS) symptoms. CNS symptoms vary from cognitive impairment (48%), confusion (often with disorientation) (39%), emotional disturbances (16%), and behavioral changes (15%), to personality changes (12%), apathy (12%), psychosis (often paranoia) (10%), and reduced vigilance from light headedness to coma (9%). Furthermore, focal neurological signs may be present, and headache is a frequent prodromal symptom. Visual changes include scotoma and visual distortion. Hearing loss, usually in the low to mid-range frequencies, may be associated with tinnitus and vertigo [1]. At the time of diagnosis, the full clinical triad is found in only a minority of the patients. In two-thirds of the patients, the syndrome starts with CNS symptoms.

The diagnosis of SS is primarily based on clinical presentation, branch retinal artery occlusions (BRAO) on fluorescence angiography (FA) and characteristic brain lesions, especially in the corpus callosum (CC) on Magnetic Resonance Imaging (MRI) [1]. MRI typically shows multiple hyperintense foci centrally in the CC, often round (called "snowball lesions"), later evolving into punched-out lesions or "callosal holes". In the acute phase, callosal lesions may show diffusion restriction, reflecting microinfarctions. The CC is always involved in the encephalopathic form of SS, but white matter lesions are also commonly seen in the subcortical and deep white matter and internal capsule. Microinfarctions in the internal capsule typically have a "string of pearls" appearance, and in combination with the characteristic callosal lesions, they are considered pathognomonic for SS. Although white matter abnormalities predominate in SS, deep gray matter (basal ganglia and thalami) is involved in around 70% of the cases. Gray and white matter lesions variably enhance after Gadolinium administration. Leptomeningeal enhancement is found in around 33% [2].

The abnormalities on MRI in SS are often mistaken for demyelinating pathology such as multiple sclerosis (MS) or acute disseminated encephalomyelitis (ADEM). Although all three diseases exhibit multifocal white matter lesions, callosal lesions in SS typically involve the central fibers with relative sparing of the periphery, in contrast to MS, which typically involves the undersurface of the CC at the septal interface. Deep gray matter lesions are unusual in MS, and leptomeningeal enhancement and callosal holes are not seen in MS or ADEM [2].

Optical coherence tomography (OCT), which allows non-invasive in vivo measurement of retinal nerve fiber layer thickness,

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http://dx.doi.org/10.1016/j.clineuro.2016.07.017

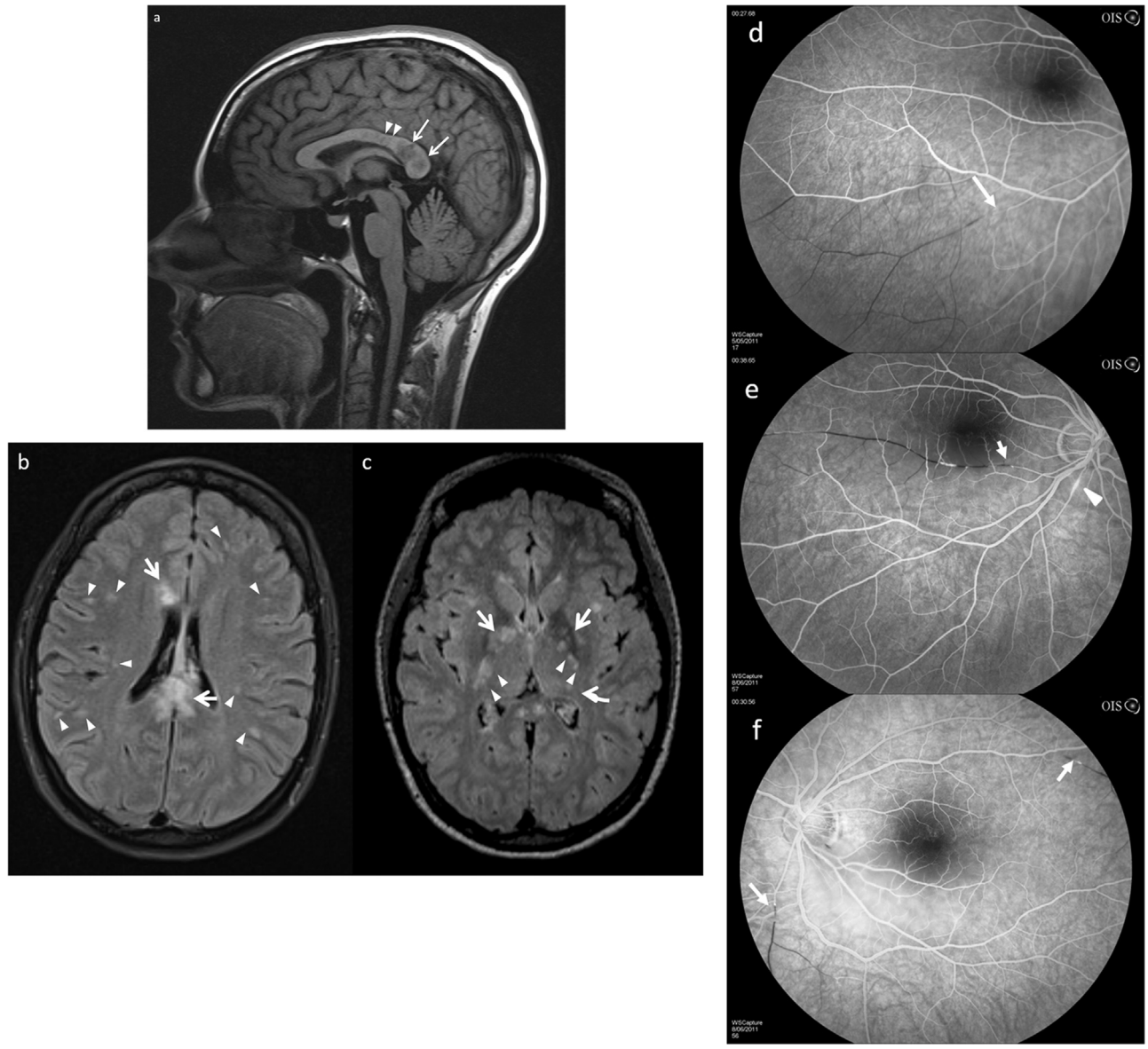

Fig. 1. Diagnostic investigations in the acute phase: MRI (a–c) and retinal fluoresceine angiography (d–f).
Sagittal T1 (a) and axial FLAIR (b and c) showing typical lesions in the acute phase:
a) midsagittal T1-weighted image shows multiple hypointense lesions located centrally in the corpus callosum with relative sparing of the periphery, characteristic of Susac's syndrome, in contrast to MS which usually affects the callososeptal interface. The larger lesions are spherical, appearing as "snowballs" (arrows). Smaller lesions show a "spoke"-configuration (arrowheads); b) axial FLAIR image again shows extensive lesions in the corpus callosum which are hyperintense on this sequence (arrows), as well as numerous smaller hyperintense foci scattered in the supratentorial white matter (arrrowheads), predominantly subcortical but sparing the U-fibers, in contrast to MS where juxtacortical lesions are typical. The callosal lesions show restricted diffusion, appearing hyperintense on B1000 diffusion-weighted images at the same level and black on the ADC-map (not shown here); c) Axial FLAIR image at a lower level shows multiple hyperintense lesions bilaterally in the globus pallidus (straight arrows) and in the left thalamus (curved arrow) which were not present on the initial images. Involvement of deep gray matter structures is common in Susac's syndrome but rarely seen in MS. New hyperintense lesions have also developed in the internal capsule (arrowheads) with a "string of pearls" appearance, another pathognomonic sign in Susac's syndrome. d) fluorescein angiography of right eye with branch retinal artery occlusion (arrow) and non-perfusion of the inferotemporal sector. These branch retinal artery occlusions could be observed in all quadrants of both eyes; e) right eye 1 month after diagnosis with new macular retinal artery occlusion (arrow) despite immunosuppresive treatment. Segmental arteritis (arrowhead) inferior of the optic nerve head; f) proximal branch retinal artery occlusion causing inner retinal edema in the inferior macula. Multiple additional vascular stops (arrows) on the fluorescein angiogram.

can also be used as a diagnostic tool. OCT could be useful for differentiating SS from MS and for disease monitoring, although it is rather aspecific and investigates small segments of the fundus. [3].

This case report presents a patient with SS with a co-morbid bipolar disorder (BD).

2. Case report

A 38 year-old Caucasian woman with Susac's Syndrome was referred to the Unit for Anxiety and Mood Disorders at Ghent University Hospital because of treatment-resistant depression. For one year, she had suffered from depressive mood and was unsuccesfully treated with different antidepressants with adequate dose and duration. Her psychiatric history revealed depressive symptoms after a job loss 4 years earlier and a manic-psychotic episode treated with olanzapine 13 months earlier. At the time, it was unclear whether the manic-psychotic episode was caused by an underlying BD, or by SS or its treatment consisting of high doses of corticosteroids. The family psychiatric history showed the suicide of her mother with a possibly undiagnosed BD, a maternal aunt