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## Short communication

# Retinal and optic disc vasculitis in Susac's syndrome ☆,☆☆

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### ABSTRACT

**Case report:** The case is presented of 42 year-old woman with no significant medical history, with severe headaches, nausea and vomiting, hearing loss, and alteration of mental status with disorientation and confusion. Ophthalmic examination showed optic disc hyperaemia in right eye, and focal areas of arteriolar occlusion in both eyes. Audiometry demonstrated bilateral neurosensory hypoacusis. Magnetic Resonance Imaging showed multiple small round hyperintense lesions located in the splenium, corpus callosum, basal ganglia, and white matter.

**Discussion:** Besides branch retinal arteriolar occlusion, the angiopathy in Susac's syndrome may affect the optic disc vessels.

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## Vasculitis retiniana y del disco óptico en el síndrome de Susac

### RESUMEN

**Caso clínico:** Mujer de 42 años, sin antecedentes de interés que acude con cefalea severa, náuseas, vómitos, pérdida de la audición y alteraciones mentales con desorientación y confusión. El examen oftalmológico reveló la presencia de hiperemia del disco óptico derecho y de áreas focales de oclusión arteriolar en ambos ojos. La audiometría puso de manifiesto una hipoacusia bilateral neurosensorial. En la resonancia magnética cerebral se detectaron

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múltiples lesiones de pequeño tamaño hiperintensas localizadas en el esplenio, cuerpo calloso, ganglios basales y sustancia blanca.

**Discusión:** Además de la oclusión de las arteriolas retinianas, la angiopatía del síndrome de Susac puede afectar a los vasos del disco óptico.

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## Introduction

Susac's syndrome (SS) is a multisystemic microvascular occlusive endothelial pathology with likely immune pathogeny involving the arterioles of the brain, the retina and the inner ear.<sup>1</sup>

First described in 1973,<sup>2,3</sup> the disease was named after John O. Susac in 1979, who published the cases of 2 young women with subacute encephalopathy, retinal arteriole occlusion and neurosensory hypo-acusia.<sup>4</sup>

The disease preferably affects females between 16 and 40 years of age,<sup>5</sup> although some cases were reported in females between 7 and 72 years of age.<sup>1</sup> It is characterized by the clinic triad of encephalopathy, retinal arterial branch occlusion (RABO) and loss of hearing. RABO constitutes the most common ophthalmological expression of SS and is present in all cases,<sup>6</sup> and may arise at any time in the course of the disease, although in many cases it remains asymptomatic because it involves small areas of the retinal periphery.<sup>1</sup>

However, optic disk (OD) angiopathy is rarely described. The case of a 42-year-old female with SS is described who, in addition to RABO, exhibited OD vasculitis.

## Clinic case report

Female, 42, without relevant history, referred by Neurology due to intense headaches with 2 months evolution accompanied by vomiting, mental alterations with disorientation and confusion, associated with tinnitus and hypoacusia at month 1 since the onset of symptoms.

Maximum corrected visual acuity (MCVA) was of 1 in both eyes. Anterior segment exploration and intraocular pressure were normal. Ocular fundus showed the presence of hyperemic papilla, without hemorrhages or exudates, in the right eye (RE). No additional vascular or retinal alterations were observed.

Fluorescein angiography (FAG) showed retinal arterial obstructions in the inferior nasal quadrant of the RE and in the temporal retina of the left eye (LE) as well as diffusion of vascular walls in the LE. In addition, it showed nearly complete hyperfluorescence in the RE, respecting 2 nasal focal areas up to late times. Macular optical coherence tomography was normal (Figs. 1 and 2).

Hemogram, biochemistry, self-immunity tests, tumor markers and serology were normal. Cerebrospinal fluid study identified 8 cells/mm<sup>3</sup> (100% mononuclear), proteins 1.21 g/l,

glucose 0.59 g/l, with negative serology, while pathological anatomy showed scarce and moderate lymphocytes.

Cerebral magnetic resonance (CMR) revealed 2 multiple small, hyperintense lesions without edema or mass effect that did not capture contrast and affected the *Corpus Callosum*, including splenius, white substance, basal ganglia and thalamus (Fig. 3). Cerebral angio-resonance and arteriography were normal.

Auditive measurements evidenced bilateral neurosensory hypoacusia with 94% hearing loss in the right ear and 60% in the left ear (Fig. 3). Auditive evoked potentials were normal.

The diagnostic of SS was established and managed with bolus comprising corticoids and immunoglobulin during 5 days with positive response, although tinnitus and hypoacusia persisted. Oral corticoids in descending pattern were prescribed for maintenance treatment during one year and mycophenolate up to this date (18 months follow-up).

## Discussion

SS appears to be a self-immune endotheliopathy that preferably involves the small vessels of the brain, retina and inner ear.<sup>7</sup> Characteristic findings include RABO, sometimes associated to segment plates of arteriolar walls and vascular wall hyperfluorescence in FAG; lesions in the center of the *Corpus Callosum* that can be observed in CMR and hearing loss detectable in audiometry.<sup>7,8</sup>

FAG is an extremely valuable diagnostic method in SS because retinal alterations could go unnoticed due to the absence of symptoms and because they affect small zones in peripheral areas, as in the case described herein. FAG enables the detection of up to 99% of RABO, including patients who do not refer visual alterations.<sup>5</sup> In addition, FAG is also useful in follow-up as it correlates with the activity of the disease.<sup>9</sup>

CMR is very important for diagnosing SS because the involvement of the *Corpus Callosum*, particularly the evidence of "snowball" lesions, is regarded as a highly characteristic sign.<sup>5</sup>

Although publications rarely report the involvement of the OD in SS, which would be the consequence of SS angiopathy, the presence of amyloaceous bodies has been described secondary to microinfarcts in the OD head capillaries and focal hyperfluorescence due to telangiectasiae.<sup>10</sup>

As with retinal vascular involvement, OD compromise in SS could go unnoticed due to the absence of symptoms. In these cases, only a detailed examination of the ocular fundus together with FAG can help detect existing alterations. Accordingly and due to the high clinic variability of SS, a high suspicion rate is necessary.

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