ATYPICAL OPHTHALMIC FINDINGS IN SUSAC SYNDROME

SÍNDROME DE SUSAC DE PRESENTACIÓN OCULAR ATÍPICA

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ABSTRACT

Case report: A 25-year-old woman with encephalopathy and sensorineural hearing loss was referred to us in order to confirm Susac syndrome, due to bilateral visual loss. Funduscopic examination revealed peripheral retinal microangiopathy.

Discussion: Susac syndrome is typically characterized by the clinical triad of encephalopathy, branch retinal artery occlusions, and hearing loss. In our case, ophthalmic findings were atypical. Interrupted vessels and microaneurisms accumulation in the peripheral retina of both eyes were observed (Arch Soc Esp Oftalmol 2007; 82: 179-182).

Key words: Branch retinal artery occlusion, microangiopathy, Susac syndrome, encephalopathy, hearing loss.

INTRODUCTION

The Susac syndrome consists in a microangiopathy of the brain, retina and cochlea, and is typically characterized by the clinical triad of encephalopathy, occlusion of retinal arterial branches and loss of hearing (1). Typically, the syndrome affects young women between 20 and 40. The syndrome has a very low prevalence, with under 100 cases described in the literature. Its etiology is unknown although it has been proposed to be in pathy, occlusion of retinal arterial branches and many other theories such as self-immune disorders, abnormalities in coagulation and microembolization (1,2). The

RESUMEN

Caso clínico: Mujer de 25 años de edad afecta de encefalopatía y pérdida auditiva neurosensorial, remitida a nuestro servicio para confirmar un síndrome de Susac por presentar una disminución de agudeza visual bilateral. En la exploración funduscópica se apreció una microangiopatía retiniana periférica.

Discusión: El síndrome de Susac se caracteriza típicamente por la tríada encefalopatía, pérdida auditiva neurosensorial, y oclusiones de ramas arteriales retinianas. En nuestro caso las alteraciones oculares eran atípicas, apreciándose amputación de vasos terminales con cúmulo de microaneurismas en retina periférica de ambos ojos.

Palabras clave: Oclusión de ramas arteriales retinianas, microangiopatía, síndrome de Susac, encefalopatía, pérdida auditiva.
most frequent finding in Susac syndrome is the ischemic retinal whitening caused by arterial occlusions. We present the case of a Susac syndrome patient with non-typical ocular findings.

**CASE REPORT**

A 25-year old woman was referred to our service to confirm the presence of a Susac syndrome due to bilateral visual loss. The patient had a history of alterations in her balance, ataxia, fecal and urinary incontinence and left side auditory loss, with a 4-year development. She was receiving corticoids, nimodipine and anticoagulants. The neurological assessment showed a spastic paraparesia, tendinous hyperreflexia and genital-urinary dysfunction. The blood analysis which include hemogram, general biochemistry, globleule sedimentation rate, angiotensin converting enzyme and coagulation gave no alterations. Serology for lues, HIV, herpes simple, hepatitis B and C, reactive protein C, rheumatoid factor, lupic anticoagulant, antinuclear antibodies, antcardiolipine and antiphospholipid was negative. Chest X-ray, electrocardiogram, echocardiography and cranial CAT scan did not reveal alterations. The cerebro-spinal liquid (CPL) was clear and with normal opening pressure, with slight protein increase and rest of the formula normal. The CPL serology for borrelia, Epstein Barr virus, citomegalovirus and lues were negative as were the usual cultures. The detection of oligoclonal strips was also negative.

A Nuclear Magnetic Resonance (NMR) of the brain revealed multiple focal areas of hyperintensity in the periventricular and pericallosal white substance, and in the regions of the semioval centre (fig. 1). The standard audiometric tests showed an asymmetric neurosensory hearing loss, with left side predominance, for low frequencies. The ophthalmological assessment produced a visual acuity of 20/25 in both eyes. Goldmann campimetry (HaagStreit, Bern, Switzerland) revealed a bitemporal inferior quadrantanopsia. The anterior segment biomicroscopy did not show noteworthy findings.

The funduscopy assessment revealed a peripheral retinal microangiopathy, with amputation of vessels with accumulation of microaneurisms in both eyes (fig. 2). The assessment did not reveal signs of inflammation or cells in the vitreous, or sheeting of retinal vessels. Fluorescein angiography (FA) showed signs of ischemia in the areas corresponding to the arteriole occlusions (fig. 3). The Susac syndrome diagnostic was based on the association of encephalopathy with typical findings in the NMR, retinal microangiopathy and neurosensory hearing loss. Due to the evident peripheral retinal ischemia shown by the FA, a laser retinal photocoagulation was made on the patient in the ischemic areas for prophylactic purposes, maintaining the stability of the visual condition since then. Two years later there have been no significant variations in the neurological and hearing condition.

**DISCUSSION**

Our patient exhibited a microangiopathic involvement of the brain, cochlea and retina which mat-
fusions with multiple sclerosis with which this syndrome shares a number of clinical and NMR signs. This screening is particularly important because the diagnostic of Susac syndrome is strictly clinical as there is no known specific marker.

The pathogeny is also unknown and the efficiency of treatment uncertain, although the most accepted treatments include corticoids and antiaggregating agents, such as in our case. Additional options include cyclophosphamide, calcium antagonists, plasmapheresis and immunoglobulines (2,4).

Ophthalmologists should know and be aware of this infrequent and many times undiagnosed syndrome of unknown original, because it could lead to visual loss. In addition, the ophthalmological findings can be very subtle, with minimum or virtually absent visual symptoms and without exhibiting the typical occlusions of retinal arterial branches. Accordingly, ophthalmologists must be very thorough in their funduscopic explorations when this syndrome is suspected.

REFERENCES