AUTOIMMUNE HEPATITIS IN A PATIENT WITH SERPIGINOUS CHOROIDITIS

HEPATITIS AUTOINMUNE EN UN PACIENTE CON COROIDITIS SERPIGINOSA

GÓMEZ-MAESTRA MJ¹, FRANCÉS E², AUSÍN E¹, MARTÍNEZ-COSTA R²

ABSTRACT

Case report: We report the case of a patient with serpiginous choroiditis who developed an autoimmune hepatitis.

Discussion: The etiology of serpiginous choroiditis remains uncertain. An immune-mediated mechanism has been reported. This case shows an association between serpiginous choroiditis and autoimmune processes (Arch Soc Esp Oftalmol 2007; 82: 773-776).

Key words: Serpiginous choroiditis, autoimmune hepatitis, autoimmunity, steroids, choriocapillaris.

RESUMEN

Caso clínico: Se presenta el caso clínico de una paciente afecta de coroiditis serpiginosa que presentó un cuadro de hepatitis autoinmune.

Discusión: La etiología de la coroiditis serpiginosa permanece desconocida. Se ha postulado una anormal respuesta de sistema inmunitario como mecanismo. Este caso muestra la asociación de la coroiditis serpiginosa con un proceso de tipo autoinmune.

Palabras clave: Coroiditis serpiginosa, hepatitis autoinmune, autoinmunidad, esteroides, coriocapilar.

INTRODUCTION

Serpiginous choroiditis is a rare inflammatory disease which affects the choriocapillaris and the retina’s pigmentary epithelium (RPE). It is also known as peripapillary helicoidal geographic choroiditis due to the typical clinical findings. It usually involves both eyes, but it can also be asymmetric. It usually evolves progressively and slowly, with multiple recurrences. It also expresses in the form of blurred vision and paracentral scotomae (1).

Serpiginous choroiditis is slightly more prevalent in men. The age bracket in which this disease expresses is between 30 and 70 years. In the absence of treatment, the prognosis is negative due to macular involvement.

Lesions generally begin in the papilla and disseminate following a winding pattern outwards and towards the macula. Acute lesions are subretinal grey-yellowish infiltrates. As the lesions advance, chorioretinal scarring occurs with variable hypertrophy of the RPE, progressive atrophy and fibrosis.
Choroidal neovascularization occurs in about 10% - 25% of patients. In relapses, the lesions appear adjacent to each other or radially around existing scars.

The diagnostic is based on clinical findings. Fluorescein angiography (FAG) is quite useful. In the initial stage, severe lesions exhibit hypofluorescence in the centre and hyperfluorescence in the edge. In the late stage, hyperfluorescence appears. Inactive lesions present hypofluorescence with belated tincture. Indocyanine green angiography is also useful to detect active lesions (2).

The treatment (3) which has proved to be the most efficient comprises a triple therapy with systemic corticoids, azathioprine and cyclosporine, although single therapy with cyclosporine can be adequate in the early stages.

**CASE REPORT**

A sixty two-year-old woman who refers multiple episodes of blurry vision in both eyes dating 13 years back with subsequent full recovery of visual acuity. Said episodes were treated with oral corticoid therapy, which was suspended after the respective remissions. She was diagnosed with Serpiginous choroiditis on the basis of clinical and angiographic findings.

Her family history includes a twin sister of her mother with a similar condition. Additional personal medical history is not relevant.

In the most recent ophthalmological assessment, the disease remained inactive and her visual acuity (VA) was of 1 in both eyes in despite of the important atrophy and chorioretinal scarring in the posterior pole, including the macula. The anterior segment did not exhibit relevant findings and the intraocular pressure was of 14 mmHg in both eyes.

The eye fundus exhibited multiple cicatricial and geographically extended lesions with the variable atrophy and hypertrophy of the RPE throughout the posterior pole (figs. 1 and 4). Fluorescein angiography showed multiple dot-shaped lesions, due to...
atrophy and hyperpigmentation with tincture in late stages (figs. 2, 3, 5 and 6).

Four months earlier the patient began to exhibit flatulent dyspepsia, vomits and itching. The laboratory studies revealed an increase of transaminase (GOT 110 and GPT 90), GGT (gammaglutamyl-transpeptidase) 110, hypergamma-globulinemia (IgG 1620), ANA (Antinuclear antibodies) 1/320. AMA (Antimitochondrial Antibodies) negative. VHB surface antigen, anti-HBs antibody, anti-HBc antibody and anti-VHC antibody, all negative. An echography of the liver was performed with normal results.

The patient responded positively to calculolytic biliar acids. The suspected diagnosis was self-immune hepatitis. It was decided not to carry out aggressive diagnostic tests such as a liver biopsy due to the slight activity of the digestive condition.

DISCUSSION

The etiology of Serpiginous choroiditis (SC) remains unknown, although it is believed that it could be caused by an abnormal response of the immune system. An increase in the HLA-B7 presentation has been found as well as associations with the retinal S antigen. In addition, a possible association has been proposed with herpes virus (1). Likewise, an increase of von Willebrand factor VIII was found in a small series of patients.

The histopathology of chorioretinal lesions of SC shows diffuse and localized infiltrates of lymphocytes in the choroids. The infiltrates are larger in the margins of the lesions, which are the most active areas.

This communication shows the presentation of a self immune hepatitis in a patient with a long-evolving SC. An abnormal response of the immune system of this patient could give rise to both processes. Fuentes-Paez et al. (4) presented the case of a patient with clinically stable systemic erythematosus lupus who later developed SC. Santos-Bueso et al. (5) described the case of a patient diagnosed with SC who three months earlier exhibited a joint infec-

Fig. 4: The funduscopy examination of the left eye shows similar lesions to that of the right one, and the patient also exhibits a VA of 1.

Fig. 5 and 6: FAG of the left eye shows in active lesions, with hypofluorescence in early stages and hyperfluorescence in later stages.
tion with hepatitis B and C virus. This case is not a self immune disease associated to SC because the existence of number of viral antigens could have given rise to an immunological alteration which eventually led to the appearance of a retinal lesion. These cases supports the hypotheses that an immunological alteration causes the retinal inflammatory lesions which are typical of SC.

REFERENCES


