ABSTRACT

Case report: A 45-year-old man was referred to us with a scotoma sensation in his right visual field. Funduscopic examination showed aneurysmal dilatations with lipid exudation in the superior and inferior temporal retinal arcades in his right eye. Angiofluorography showed vascular dilatations and late dye leakage.

Discussion: Leber’s disease is a primary retinal vasculopathy characterized by several aneurysms associated with intraretinal lipid exudates. Visual compromise depends on macular involvement by the exudates. Differential diagnosis must be established with other primary retinal vasculopathies and secondary aneurysms. Treatment consists of argon laser photocoagulation of the aneurysms (Arch Soc Esp Oftalmol 2008; 83: 669-672).

Key words: Aneurysm, Leber, miliary, exudate.

INTRODUCTION

Leber’s miliary aneurysm is an idiopathic retinal vasculopathy characterized by dilatations of venules and arterioles which cause chronic extravasation and intraretinal solid exudates. This aneurysm is part of primary retinal telangectasiae, together with juxtafoveal telangectasia and Coats disease. It is important to carry out a differential diagnostic of this disorder with other causes of primary and secondary retinal aneurysms because the visual prognosis varies considerably.

CASE REPORT

A 45-year-old man without relevant history attended the emergency ward due to scotoma in the right eye which began a few days back. Visual acuity was of 9/10 in both eyes, intra-ocular pressure of 12
mm Hg and biomicroscopy in both eyes was normal. The funduscopy assessment of the right eye revealed five aneurysm-like dilatations due to lipid exudates in the superior temporal arcade, and three in the inferior temporal arcade (figs. 1A and 1B). The left eye was normal. Fluorescein angiography (FAG) revealed vascular dilatation and the late extravasation of the dye (figs. 2A and 2B). Considering a possible diagnostic of Leber’s miliary aneurysm, it was decided to initially maintain an anticipative attitude.

Four months later the exudate did not diminish and treatment was established with argon laser photocoagulation of the aneurysms.

After two sessions of treatment, the exudation disappeared. Four years later, the eye fundus shows the laser scars (figs. 3A and 3B).

DISCUSSION

Primary retinal telangiectasiae retinal vascularization abnormalities characterized by multiple aneurysms which cause chronic extravasation and deposit of hard intraretinal exudates. Three entities are considered in this group: juxtafoveal

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Fig. 1: RE fundus image. A: 5 aneurysm-like dilatations in the superior temporal arcade with surrounding lipid exudation. B: 3 aneurysms in the inferior temporal arcade.

Fig. 2: A. Early times FAG (43 sec) showing the aneurysms in the superior temporal arcade of the RE. B: Image of the inferior temporal arcade (2 min.).
telangiectasias, Leber miliary aneurysm and Coats disease.

Leber aneurysms were first described in 1912. Initially they were considered to be different to Coats disease. Subsequently, Reese proposed to group them as «primary retinal telangiectasias» to become part of the spectrum of the same disease.

Leber’s disease in general appears in one eye in males around the fourth decade of life. The most frequent localization is the temporal hemi-retina between the posterior pole and the peripheral retina.

The diagnosis is made with a funduscopy and angiography assessment, showing multiple aneurysms with chronic and focalized extravasation which give rise to hard exudates. In the early stage of FAG there is hyper-fluorescence of vascular dilatations and at the later stage a diffusion of contrast from the telangiectasias, which may cause cystoid macular edema (1).

The differential diagnostic is approached with other primary and secondary retinal aneurysms. In contrast with Leber’s aneurysms, juxtafoveal telangiectasias appear in the posterior pole (causing an earlier reduction of vision) and generally in people over 40. The angiographic viewpoint is different: it exhibits small vascular dilatations in the perifoveal area (2). Coats disease is the most severe of all three primary vasculopathies, predominantly affecting the pediatric period of life. Visual compromise is greater and with a poorer prognosis because of larger and more extended exudates, which may evolve to retinal detachment (3). Forms of Coats disease have been described in adults with smaller functional repercussions, but the number of aneurysms and exudates is much greater than in Leber aneurysms (4). Retinal arterial macroaneurysm is more frequent in women, the third age and with high blood pressure. One or more arteriolar dilatations appear at an arterial-venous bifurcation, with more posterior localization. Diabetic retinopathy must be discarded through personal history or analytics. Aneurysms are smaller and include other signs of diabetic retinopathy such as cotton-like exudates or intraretinal hemorrhages, which do not appear in Leber’s disease.

The differential diagnostic must comprise other vascular diseases such as venous occlusion, retinal capillary and cavernous hemangioma as well as discarding past radiation as a cause of retinopathy with micro-aneurysms (1).

The case report described was labeled as Leber’s miliary aneurysms after discarding General diseases such as diabetes, high blood pressure or radiation. Coats disease was excluded because of the exudation was small and localized. The location far from the macula and the age of the patient made the diagnosis of juxtafoveal telangiectasia unlikely.

The development of this entity it usually slow and could go unnoticed up to later periods in life. The prognosis depends on the foveal compromise by the exudates, macular edema, vitreo-macular traction or epiretinal membrane (1,5). Other complications are possible such as thrombosis of the
aneurysms, neovascularization, hemorrhage in the vitreous and evolution to Coats disease.

The treatment consists in ablation of the aneurysms by argon laser photocoagulation if the exudate may compromise the macula. In some cases, triamcinolone or intra-vitreous antiangiogenics could be associated, as well as vitreo-retinal surgery in the presence of macular traction or epiretinal membrane. If the aneurysms are located in the periphery, cryotherapy could be considered (1).

In the instant case it was decided to applied photocoagulation to the aneurysms in spite of the focal nature and the poor exudation after considering that iatrogenia would be minimal and to avoid subsequent complications.

REFERENCES