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Central Retinal Vein Occlusion Revealing Anti Phospholipid Antibody Syndrome: Case Report

Dr. Rkia Azahrai
MD, Ophthalmology Department,
University Hospital Center Mohammed VIth
Oujda, Morocco. (Corresponding Author)

Dr. Manar Achargui
MD, Ophthalmology Department,
University Hospital Center Mohammed VIth
Oujda, Morocco.

Dr. Fatimazahra Mabrouki
PhD, Ophthalmology Department,
University Hospital Center Mohammed VIth
Oujda, Morocco.

Siham Chariba
PhD, Ophthalmology Department,
University Hospital Center Mohammed VIth
Oujda, Morocco.

Asmaae Maadane
PhD, Ophthalmology Department,
University Hospital Center Mohammed VIth
Oujda, Morocco.

Rachid Sekhsoukh
PhD, Ophthalmology Department,
University Hospital Center Mohammed VIth
Oujda, Morocco.

Abstract:- Anti-phospholipid antibody syndrome (APAS) is defined by the association of at least one clinical event (vascular occlusion, fetal loss) with the presence of antibodies to phospholipids, and may be isolated or associated with systemic lupus erythematosus. The vascular site of the eye has been described in the literature and may be the main manifestation.

We report the case of a 21-year-old woman admitted to the emergency department with a rapid unilateral decrease in visual acuity. Ophthalmological examination revealed an occlusion of the central retinal vein . The diagnosis of SAPL was made during the etiological work-up with the presence of anti-cardiolipin antibodies, the patient was put on anti-platelet aggregation treatment. The evolution was marked by the occurrence of a miscarriage, after which the patient was put on anticoagulant treatment.

When faced with an occlusion of the central retinal vein or artery in a young subject, in addition to the recommended cardiovascular check-up, a thrombophilia and SAPL check-up is essential in order to avoid a thrombotic recurrence which can be life-threatening if it is a cerebral location.

Keywords:- *Anti Phospholipid Antibody, Thrombosis, Central Retinal Vein.*

I. INTRODUCTION

Antiphospholipid antibody syndrome (APAS) is defined by the occurrence of arterial and/or venous thrombotic occlusions or recurrent miscarriages associated with the presence of antiphospholipid antibodies on several occasions(1), it is referred to as primary APAS when it is isolated, or secondary when it is associated with another pathology such as systemic lupus erythematosus (SLE), drug intake or certain infections(2.3). Ocular vascular damage (occlusion of the central retinal vein or artery, occlusion of the choroidal vessels) has been described in the literature, and can sometimes be the first sign of the disease, especially in young people (1). hence the interest of etiological investigations in order to diagnose this pathology and treat it early given the risk of recurrence and retinal damage or damage to new vascular sites, particularly the cerebral site.

II. OBSERVATION

This is a 21 year old female patient, whose father was treated for Behcet's disease with ocular involvement and without any particular pathological or obstetrical history, who was seen in emergency for a sudden and painless drop in visual acuity in the right eye 48 hours before her admission. The examination found a collapsed visual acuity limited to counting fingers at 1 metre in the right eye, a normal anterior segment and ocular tone, and in the fundus multiple haemorrhages converging towards the papilla, dilatation and generalised venous tortuosity with papillary oedema and macular oedema (figure 1) . Examination of the contralateral eye was unremarkable.

Fluorescein angiography confirmed the diagnosis of central retinal vein occlusion by showing venous circulatory slowing, macular oedema and numerous territories of non-perfusion in the periphery (figure 2) and OCT showed the extent of macular oedema (figure 3).

The general examination of the patient did not reveal any abnormalities, particularly in terms of blood pressure, and the cardiovascular examination, the echocardiogram and the ultrasound of the supra-aortic trunks did not show any emboligenic focus or atheromatous lesions. Biological examinations showed: a normal VS, blood sugar, lipid profile, blood count, haemostasis profile (TP, TCA, protein C, protein S, antithrombin), Anti-phospholipid syndrome (APS) was evoked by the positivity of IgG anti-cardiolipin bodies and anti-β2 glycoprotein G1 on two occasions and at an interval of six weeks.

The rest of the thrombophilia work-up was normal and the immunological work-up did not show any abnormality in favour of lupus or any other systemic disease. The diagnosis of primary APS was accepted and the patient was put on aspirin at a dose of 160 mg per day . Ocularly, the patient underwent pan-retinal photo coagulation (PPR) with IVT of anti-VEGF . The evolution was marked by an improvement of the visual acuity with a visual acuity raised to 1/10, after 1 year the patient presented 2 miscarriages for which she was put under anticoagulant treatment.

III. DISCUSSION

The causes of retinal vascular occlusions (arterial or venous) are multiple and they differ according to the age of onset and the patient's background.(4)

In the majority of cases (75%) an occlusion of the central retinal vein should lead to a search for vascular risk factors: arterial hypertension, diabetes, smoking, dyslipidemia(4), In some cases, and particularly in young people who have no risk factors for atheromatous pathology, aetiological investigations should be carried out to find more or less rare causes of CRVO, such as hyperviscosity pathologies (haemopathies, polycythemia), thrombophilias and autoimmune pathologies(4).

APS is characterised by the presence of thrombotic manifestations, arterial or venous, whatever the size of the vessels or repeated miscarriages, associated with the long-term presence of an anti-cardiolipin or anti-prothrombinase antibody (the positivity of the antibodies must be sought on several occasions with an interval of 6 weeks between 2 samples) (table 1).

APS can be primary, if it is isolated as in our patient's case, or it can be secondary if it is associated with another autoimmune disease, most often lupus (1)(6).

Retinal involvement in primary or secondary APS is rare, representing 3-8% of cases (5), (7). It occurs in the form of localized or diffuse retinal vascular occlusions (occlusion of the central retinal artery or vein(8), occlusion of the cilio retinal artery, occlusion of the multifocal arteriolar vessels); these attacks can be severe, especially in the case of bilateral forms: Optic neuropathy, conjunctival telangiectasia, limbal keratitis and episcleritis (1)(9), and the most frequently described sign is the tortuosity of the retinal veins: a prospective study of 17 patients with primary APS, in which fluorescein angiography was carried out, showed a tortuous aspect of the retinal veins in 29% of cases (9).

Physiopathologically, a link between anti-phospholipid antibodies and the phospholipids of platelets and endothelial cells is formed, this complex will trigger the activation of haemostasis and endothelial cells, which will lead to a cascade of events (secretion of adhesion molecules and inflammatory cytokines) resulting in a thrombotic event (9)

Through this case, we insist on the interest of haematological investigations especially in the young patient without vascular risk factors who presents a retinal vascular occlusion, it is necessary to detect a SAPL and differentiate a primary SAPL from a secondary SAPL.

An early and accurate diagnosis of APS allows the patient to have a better management and a good prognosis, after a spontaneous thrombotic episode, the risk of recurrence is high and requires a treatment based on long-term anticoagulation (1), so the occurrence of a severe occlusion is a sign of seriousness, both by the functional impairment following retinal ischemia, and by the risk of extension of the thrombus to other vessels

The evolution of APS is characterised by the occurrence of complications, sometimes even with anticoagulant treatment the mortality rate can be as high as 30% (9) (10)

IV. CONCLUSION

A picture of retinal vascular occlusion in a young subject should lead to a search for the anti-phospholipid antibody syndrome, essentially when vascular risk factors are absent or in the course of a lupus.

One should not hesitate to increase the number of immunological tests for anti-phospholipid antibodies and to check their persistence in the presence of such cases

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TABLES

Clinical criteria	Biological criteria
- 1 or more episodes of vascular thrombosis - 1 or more unexplained fetal deaths >10SA - 3 successive unexplained spontaneous abortions < 10 days' gestation - 1 or more premature deliveries < 34 days' gestation on a background of severe pre-eclampsia or severe placental insufficiency	- Presence of elevated anticardiolipin IGG or IGM on two samples taken at least 6 weeks apart - Presence of antithrombinase in two samples taken at least 6 weeks apart

Table 1: Clinical and biological criteria for APAS

FIGURES

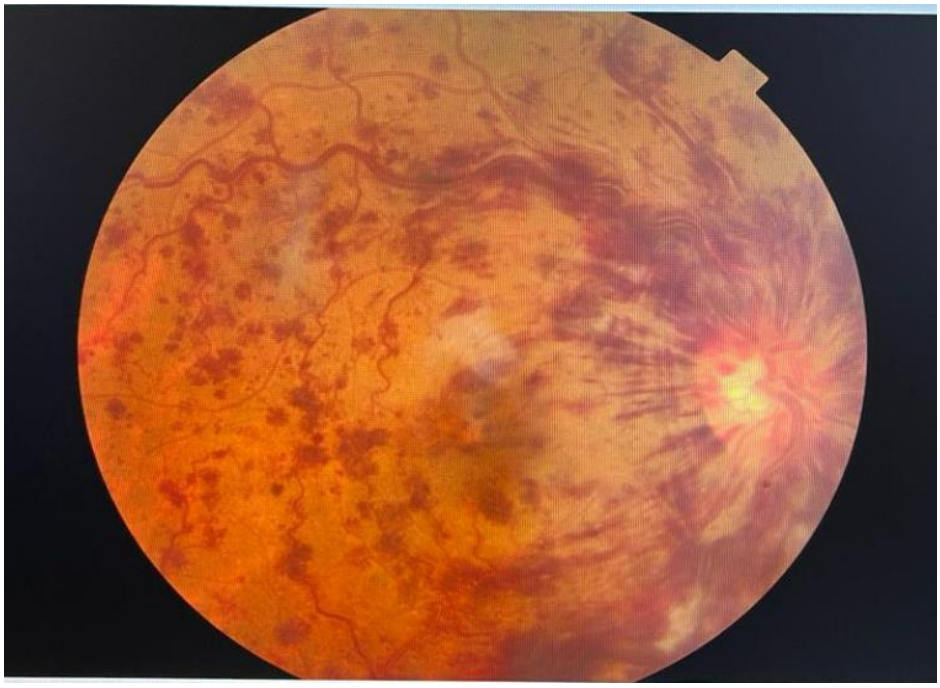

Fig . 1: Fundus of the right eye showing multiple haemorrhages converging towards the papilla, dilatation and generalised venous tortuosity with papilledema and macular oedema

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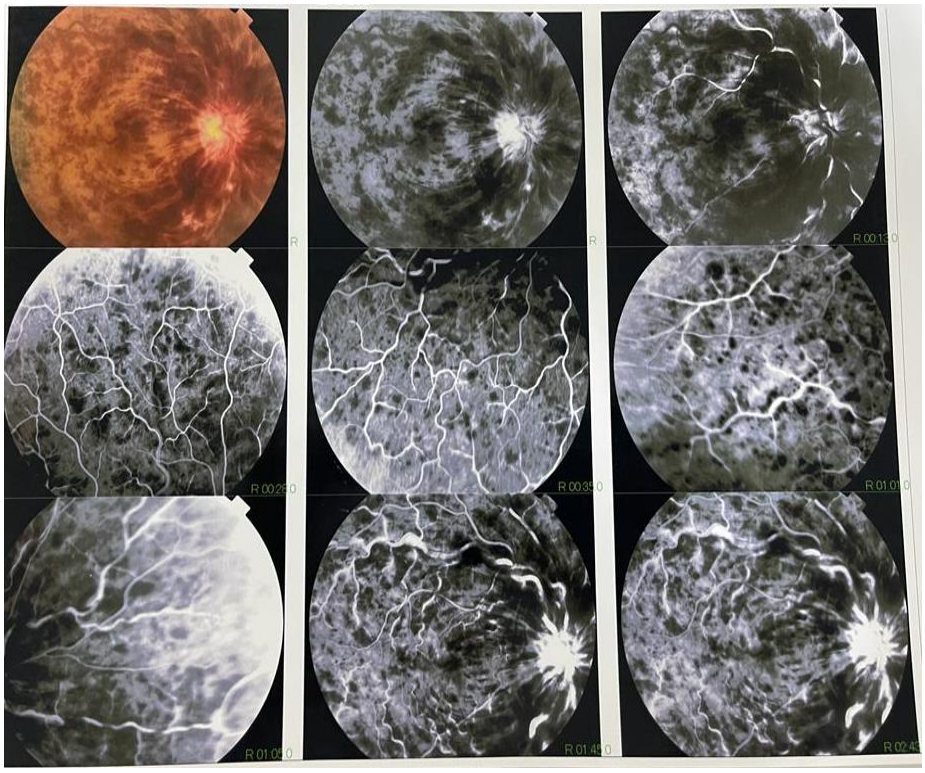

Fig .2: Retinal angiography showing venous circulatory slowdown, macular oedema and numerous non-perfusion territories in the periphery

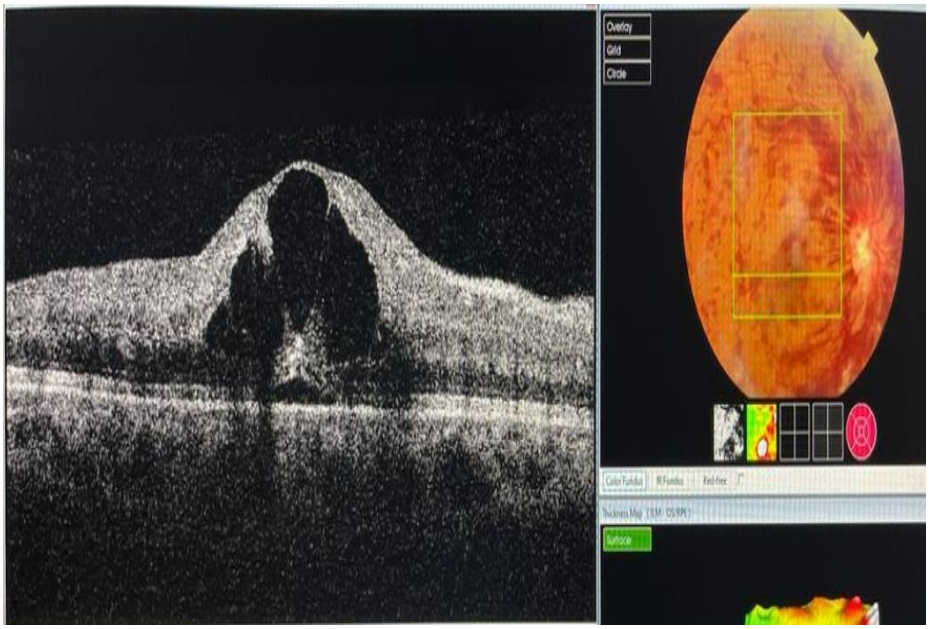

Fig .3: Macular OCT showing severe macular oedema