

Retinal Vascular Occlusion and Primary Antiphospholipid Antibody Syndrome: A Case Report

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Abstract

Case Report

The antiphospholipid syndrome is characterized by the combination of antiphospholipid antibodies and arterial and / or venous thrombotic phenomena. Ocular vascular involvement is described in the literature and may be indicative of the syndrome. His diagnosis is all the more important as the ocular and even vital prognosis of these often young patients can be engaged. We report an observation illustrating this situation. A young people aged 32 presented with a sudden unilateral loss of vision. Fundus examination and fluorescein angiography revealed a severe retinal vascular occlusion. an antiphospholipid syndrome was discovered during etiological check-up. It was associated with a heterozygot state mutation of Leiden V factor. In most cases central artery or vein occlusion means atherosclerosis. However, other etiologies must be studied in young patients. According to several studies: antiphospholipid syndrome has been detected in 5% to 33% of the patients showing a major vascular retinal obstruction. The association of thrombophilia must be considered because it increases the risk of thrombotic recurrence. The antiphospholipid syndrome must be studied in cases of numerous and severe retinal vascular obstruction occurring in young patients, even if vascular risks exist. This diagnosis is important because it may imply a long-lasting anticoagulative or an antiaggregative treatment to significantly reduce the risk of recurrent thrombotic events.

Keywords: Retinal vascular occlusion, primary antiphospholipid antibody syndrome.

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INTRODUCTION

The antiphospholipid syndrome is characterized by the combination of antiphospholipid antibodies and arterial and / or venous thrombotic phenomena. Ocular vascular involvement is described in the literature and may be indicative of the syndrome. His diagnosis is all the more important as the ocular and even vital prognosis of these often young patients can be engaged. We report an observation illustrating this situation.

CASE REPORT

Patient aged 32 years old who consulted for a unilateral decrease in visual sharpness. Fundus examination and fluorescein angiography revealed severe retinal vascular occlusion.

Biological examinations revealed thrombocytopenia at $51\ 000 / \text{mm}^3$, spontaneous TCA at 45 seconds for a control at 32 seconds. The hematocrit and fibrinogen counts were normal, as was blood glucose, lipid status with LDL 0.85 g/l, blood ionogram, protidemia, serum calcium, liver function and renal function.

The association of atypical thrombotic events in a young woman with few vascular risk factors, thrombocytopenia and a spontaneous increase in TCA has been hypothesized as an SAPL. The diagnosis was asserted by the presence of an antiprothrombinase, an IgG anticardiolipin level of 62 units, a level of anti- β 2 glycoprotein I IgG at 79 AU (significance threshold above 5 AU) and dissociated syphilitic serology (VDRL +, TPHA -). There were no anticardiolipin antibodies or anti- β 2 glycoprotein 1 IgM. All these results were confirmed 3 months later. The rest of the immunoassay did not show any abnormality in favor of associated lupus disease (absence of native and denatured anti-DNA antibodies). As the number of lupus disease criteria was insufficient, it was therefore a primary SAPL revealed by retinal vascular occlusion. The remainder of the thrombophilia assessment revealed a heterozygous factor V Leiden mutation.

DISCUSSION

The antiphospholipid syndrome SAPL is characterized by the association of arterial thrombosis and/or venous thrombosis of any size vessel or repeated abortions with the durable presence of an

antiprothrombinase or anti-cardiolipin antibody. The presence of an antiphospholipid antibody must be confirmed several times to be able to be diagnosed with SAPL [1, 2].

Thus the eye can be affected by this syndrome in an inaugural way or not. This syndrome is described as primary if it is isolated and secondary if it is satellite of a systemic pathology, lupus most often, justifying the search for its clinical and biological criteria and the evaluation of the probability score [2]. Our observation corresponds to a primary SAPL (atypical thrombotic episode with confirmed presence of antiphospholipids).

Retinal arterial or venous occlusion during primary or secondary SAPL is rare [1]. Its frequency varies according to the studies from 3 to 8% [2]. Various eye charts have been reported in the SAPL: vein occlusion [4] or central retinal artery, diffuse retinal vaso-occlusive vasculopathy (occlusion of the central retinal artery, multifocal retinal arteriolar occlusions or retinal vein occlusion).) sometimes severe [5] as such was the case for this patient. This vasculopathy is all the more severe as it is bilateral. In addition, optic neuropathy or other rarer manifestations such as conjunctival telangiectasia, episcleritis, and limbic keratitis have been reported [2]. The most common sign seems to be a tortuous aspect of the veins. A prospective study in which fluorescein angiography was performed systematically in 17 patients with primary SAPL revealed a tortuous aspect of the veins in 29% of cases [7].

Pathophysiologically, these antiphospholipid antibodies bind in particular to the phospholipids of platelet and endothelial cell membranes, resulting in arterial and venous thrombotic events [6]. The potential mechanisms of thrombosis are explained by two types of interactions [7]: - the interaction with the platelet membrane which is responsible for thrombocytopenia and activation of hemostasis; And the interaction with the endothelial cells resulting in the expression of adhesion molecules, the secretion of inflammatory cytokines and a modulation of arachidonic acid metabolism. Faced with retinal occlusive vasculopathy in a patient with lupus disease, do not hesitate to evoke an SAPL [8] and strive to find antiphospholipid antibodies that we have also been found in this patient. Moreover, a first positive detection is not sufficient to affirm the diagnosis of SAPL: it is necessary to prove the persistence of antiphospholipids at the earliest 6 weeks after the first detection [2].

There are different antiphospholipid antibodies. The most commonly used tests for their detection are the search for an antiprothrombinase (coagulation tests to be done before any heparin) and the search for anticardiolipin antibodies IgG and IgM (Elisa immunological tests). Added to this is the detection of anti- β_2 glycoprotein 1 (Elisa

immunological tests) recognized as one of the cofactors of anticardiolipin antibodies. Finally, the positivity of the VDRL with a negative TPHA is a weak and not very specific test. These tests are positive independently or together. A first positive detection is not sufficient to affirm the diagnosis of SAPL: it is necessary to prove the persistence of antiphospholipids not earlier than 6 weeks after the first detection.

It is essential to search in parallel for associated thrombophilic pathology: antithrombin deficiency, protein C and S, resistance to activated protein C, factor V Leiden mutation, mutation of the prothrombin gene 210 and, in the case of certain authors, fasting hyperhomocysteinemia. The frequency of these disorders of hemostasis, excluding the search for a factor V Leiden mutation and prothrombin gene is of the order of 7 to 21% in occlusions of the central retinal vein [7]. The coexistence of any of these coagulation abnormalities may increase the risk of thrombotic recurrence in patients with ASL. The combination of, for example, an SAPL and a factor V Leiden mutation increases the risk of recurrence compared to an isolated SAPL as probably in the observation.

CONCLUSION

In front of an array of multiple and severe retinal vascular occlusions of the young subject, it is necessary to look for an antiphospholipid syndrome even if there are vascular risk factors. Its diagnosis is important because the treatment will in certain cases involve anticoagulation or antiaggregation, sometimes in the long term, significantly reducing the risk of new thrombotic episodes.

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