LETTER TO THE EDITOR

Combined central retinal vein and cilioretinal artery occlusions in HELLP Syndrome: A case report

Occlusion combinée de l’artère ciliorétinienne et de la veine centrale de la rétine au cours du HELLP syndrome : à propos d’un cas

We report an unusual case of combined central retinal vein (CRVO) and cilioretinal artery occlusions in a patient with hemolysis, elevated liver enzymes, low platelets (HELLP) syndrome.

A 27-year-old primigravida patient at 37 weeks gestation presented with 2 days history of blurred vision and para-central scotoma of the right eye. The best-corrected visual acuity was 20/25 in the right eye and 20/20 in the left eye. Funduscopic examination revealed an area of ischemic retinal whitening in the papillomacular bundle (Fig. 1a, black arrows). There were also some peripapillary cotton wool spots with intraretinal hemorrhages. Examination of the left eye was unremarkable. Fluorescein angiography (Fig. 1b) confirmed delayed filling of the right cilioretinal artery (black arrowhead) with a hypoperfusion of an area (white arrowheads) corresponding to the retinal whitening seen clinically. OCT showed an increased thickness and hyper-reflectivity of the inner retinal layers corresponding to the ischemic area (Fig. 1c, white arrows).

General examination revealed cutaneous and conjunctival pallor, a blood pressure of 145/80 mmHg. Laboratory testing revealed: low hemoglobin (9.77 g/100 mL) with decreased serum haptoglobin levels and low platelets (96,000/mm³); liver enzymes were elevated with lactate dehydrogenase at 466 U/L, aspartate aminotransferase at 325 U/L and alanine aminotransferase at 202 U/L. The prothrombin time, partial thromboplastin time and fibrinogen levels were normal, however d-dimer was slightly elevated (42.2 g/mL). Otherwise the renal function was normal.

HELLP syndrome was diagnosed. In order to exclude another cause for this condition, a workup was carried and normal results were obtained for: antiphospholipid antibody, protein C, protein S, antithrombin III, antinuclear antibodies. A carotid ultrasound duplex scan and an echocardiography were also performed and were unremarkable. The patient was then referred to an obstetrical department. After initial stabilization, obstetric ultrasound revealed mild fetal hypotrophy with partial placental detachment. Urgent cesarean section was then, carried out and resulted in the birth of a viable hypotrophic baby, indicating a chronic fetal distress.

By 21 days postpartum, there was a decrease in visual acuity to 20/50. At funduscopy, the clinical picture had evolved into a right ischemic CRVO, confirmed by a second fluorescein angiography (Fig. 2). A prompt laser retinal photocoagulation was then started.

HELLP syndrome is a life-threatening obstetric complication usually seen in the third trimester, either as part of severe pre eclampsia or as a separate disease entity. It is characterized by three abnormalities: hemolysis, elevated liver enzyme levels and a low platelet count. The pathogenesis remains incompletely understood, it could be related to an aberrant placental development [1], causing a release of

Figure 1. (a) Funduscopy of the right eye revealing an area of ischemic retinal whitening involving the papillomacular bundle (black arrows). (b) Fluorescein angiography (30 seconds after injection) showing delayed filling of the right cilioretinal artery (black arrowhead) with a hypoperfusion of an area (white arrowheads) corresponding to the retinal whitening seen clinically. (c) OCT of the right eye showing increased thickness and hyper-reflectivity of the inner retinal layers corresponding to the ischemic area (white arrows).

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circulating agents, which result in general activation of the coagulation cascade and diffuse endothelial injuries with thrombotic complications.

Several organs have been described to be potential target sites [2], especially liver, kidneys, central nervous system and eyes.

Many reports have previously described ocular involvement in HELLP syndrome, including choroidal ischemia with serous retinal detachment [3], central retinal vein occlusion [4], isolated retinal hemorrhages and edema, purtscher-like retinopathy [5], intravitreal hemorrhages, sinus cavernous thrombosis and cortical blindness. To our knowledge a combined central retinal vein and cilioretinal artery occlusions have never been reported as a complication in this syndrome.

In fact this form of retinal occlusive vasculopathy is generally rare and represents only 5% of all retinal artery occlusions, and occurs in approximately 5% of all cases of central retinal vein occlusion. This could be related to the low prevalence of cilioretinal artery that is present in only 15% to 20% of people [6].

In our case central retinal vein occlusion may have preceded cilioretinal artery occlusion. In fact, after CRVO there is an increase in resistance to retinal venous outflow that cause elevated intraluminal capillary pressure; since the perfusion pressure of the cilioretinal artery is lower than that of the central retinal artery, the cilioretinal artery circulation may be impaired in the presence of CRVO [7].

Disclosure of interest

The authors declare that they have no conflict of interest concerning this article.

References