A CASE OF HELLP SYNDROME: THE IMPORTANCE OF ANGIOGRAPHY WITH INDOCIANINE GREEN FOR DIAGNOSIS OF RETINIC AND CHOROIDAL VASCULAR OCCLUSION.

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Purpose. To present a case of retinal and choroidal vascular occlusion in conjunction with HELLP syndrome.
Methods. A 35 years-old woman presented with HELLP syndrome underwent cesarean delivery for preeclampsia. After 2 months she had a decrease of vision in OD. She had performed an eye examination with FAG and ICG.
Results. The FAG shows at the posterior pole in the perifoveola temporal area different areas of hyperfluorescence. ICG reveals frameworks of hypoperfusion in perimacular areas, for the probable presence of ischemic foci. After about 5 mounths she presented an improvement in visual acuity and a complete resolution of Ophtalmoscopic findings.
Conclusions. Ophtalmic complications are possible during this syndrome. This is the first description of a patient suffering a retinal and choroidal vascolar occlusion after HELLP syndrome, and we report the important of ICG for diagnosis of choroidal ischemia.
Key words: HELLP syndrome, preeclampsia, retinal and choroidal vascular occlusion, thrombotic microangiopathic vasculopathy

CASE REPORT

A 35-year-old woman presented with HELLP syndrome underwent cesarean delivery for preeclampsia, which she has been treated for the previous months. She was admitted to hospital complaining of severe headache, oliguria and non-specific visual disturbance. Her medical and obstetric history was negative. Blood pressure was found to be 190/110 mmHg. Clinical laboratory tests showed elevated white blood cells (16,700 mm3 with 80 % of neutrophils), elevated erythrocyte sedimentation rate (76 mm in the first hour), abnormal liver function with elevation of aspartate aminotransferase (AST / SGOT =166 IU/l) and lactate dehydrogenase (LDH =1,112 IU/l) and severe proteinuria (50 g/l). Analysis of the coagulation system revealed thrombocytopenia (platelets 40,000/mm3), decreased plasma fibrinogen (1,88 g/l) and a short activated partial thromboplastin time (27 seconds). After 2 months from the delivery, she had a decrease of vision, discovered accidentally turning a blind eye, and she has noticed it after an ICU admission for pancreatitis, so advice she was sought from ophthalmology.
The visit found a significative cystoid macular oedema in OD and was prescribed a treatment with Acetazolamide and systemic Steroid. Blood pressure was normal (110/70 mmHg) and the intraocular pressure was normal in both eyes.
After 5 month of previous hospitalization, the patient has performed an eye examination that has found a visual acuity of 20/25, FAG and a Fundus Fluorescein Angiography with Indocianina Green (ICG) .
The FAG shows at the posterior pole in the perifoveola temporal area different areas of hyperfluorescence, which increase in extension in late times with an attitude of cystoid oedema. ICG reveals frameworks of hypoperfusion in perimacular areas, for the probable presence of ischemic foci .
The best corrected visual acuity (BCVA) is 20/25 in the right eye. Systemic steroid and oral Acetazolamide therapy was continuated, but visual acuity did not improve.
After 2 months the best corrected visual acuity(BCVA) is 20/20 in both eyes, and there was a reduction of oedema.
But after 5 months the BCVA in the right eye is 20/32 and 20/20 in the left eye. Optical coherence tomography (OCT) of the right macula detected a foveal elevation (FT 267 µm) and thickening of the temporal foveal section with diffuse hyper-reflectivity of the neuroepithelium.
After a week visual acuity improves, 20/20 in both eyes, and the patient was discharged with home therapy (systemic Steroid, Acetazolamide).

DISCUSSION

During normal pregnancy and immediate puerperium there is an increase in the level of clotting factors and in clotting activity and there are also a number of pathologic sources of thrombosis and of emboli. This increased risk may also shows itself by occurrence of retinal and choroidal vascular occlusion. The earlier literature reports that retinal vein occlusions occur during or immediately after pregnancy.
The HELLP syndrome is a thrombotic microangiopathic vasculopathy that may be present in pregnancy and puerperium. The etiology and pathogenesis of this syndrome has not been elucidated, but it has been forwarded that dysequilibrium in prostanoid metabolism exists. Frequently, the natural evolution of HELLP syndrome is one disseminated intravascular coagulation. Blood pressure values, the plasmatic blood-cloatting factors and platled count in our patient were found in the normal range at the time of retinal vein occlusion, but HELLP syndrome is characterized by unpredictable occurrence of severe maternal complications during and soon after the syndrome (3). The retinal vein occlusion and the choroidal damage are reported causes of visual disturbance observed in HELLP syndrome (4). Choroidal vascular changes, choroidal vasoconstriction, and the ischemia are responsible for the most retinal damage seen in the pregnancy-induced hypertension. FAG shows the presence of macular oedema and Indocyanene Green Angiography suggest that in the hypertensive choroidopathy endogenous vasoconstrictor agents leak freely from the choriocapillaries and act on the walls of the choroidal vessels resulting in choroidal vasoconstriction and ischaemia.
The Indocyanene Green Angiography is an important tool that assist the determination of choroidal Ischemia.

REFERENCES

1. Gonzalvo F. J. Abecia E. Pinilla I. Izaguirre L.B. (2000) : Central retinal vein occlusion and HELLP syndrome. Acta Ophtalmol.Scand. 78: 596-598
2. Murphy M.A. Ayazifar M. (2005): Permanent visual deficities secondary to the HELLP syndrome. J.Neuroophtalmology Jun; 25 (2) : 122-7
3. Tranos P.G. Wickremasinghe S.S. Hundal K.S. (2002) : Bilateral serous retinal detachment as a complication of HELLP syndrome. Eye 16: 491-492
4. Taskapili M. Kocabora S. Gulkilik G. (2007) : Unusual ocular complication of the HELLP syndrome : persistent macular elevation and localized tractional retinal detachment. Ann.Opthalmol. 39 (3): 261-263

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