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Diagnosis and Treatment of Hemoglobinopathy and Retinopathy of Sickle Cells

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INTRODUCTION

We can consider sickle hemoglobinopathiesare due the presence of one or several abnormal hemoglobins with abnormal red blood cell formation under conditions of hypoxia and acidosis.

The deformed red cells are rigid, being able to obstruct the small blood vessels and cause hypoxia.

In a normal erythrocyte, hemoglobin. Which is composed of four alpha chains and two beta chains. At the position of glutamic acid in the beta chain is precisely where we can find a point of mutation. If it was replaced by the lysine, we will have the formation of a hemoglobin. If we had valine, we will have a hemoglobin.

I can also find an AS (carrier of the sickle stroke), an AC association (carrier of hemoglobin C), the SS form (sickle-cell anemia), the SC form (sickle cell disease of the Hb C carrier).

There are also other abnormalities in the synthesis of the blood chain alpha or beta, this will cause the so-called thalassemia, when combined with a sickle hemoglobin, then the sickle or Sthal thalassemia.

OCULAR MANIFESTATIONS

The most serious forms of retinopathy are those that appear in the diseases SC (sickle cell disease with hemoglobin C) and Sthal (Thalassemia).

Sometimes milder hemoglobinopathies can also cause retinopathy.

Proliferative Falciform Retinopathy can be divided into five stages:

- Stage 1: It is characterized by peripheral arteriolar occlusions.
- Stage 2: Peripheral arteriovenous anastomoses begin to appear as existing dilated capillary channels. After vascular occlusion, the peripheral retina appears for the most part avascular and with no capillary perfusion.
- Stage 3: Presents the beginning of neovascularization from the anastomoses. In the beginning, the new vessels remain flat in the retina and show a fan-shaped configuration (neovascularization in "sea fan", this name resembles a marine invertebrate).
- Stage 4: It is characterized by the presence of vitreous hemorrhage of variable degree. Hemorrhage can be caused by ocular trauma, which is sometimes unimportant.
- Stage 5: There is a traction of the vitreous and retinal detachment.

Rhegmatogenous retinal detachment may also appear as a result of a tear formation adjacent to the areas of fibrovascular tissue.

We can find other non-retinal manifestations:

- Conjunctiva: it is common to see conjunctival isolated dark red vascular segments with a "coma" or "corkscrew" shape, affecting the vessels of small caliber, being the most common location in the lower part of the conjunctiva.
- Uvea: in the iris we can find a circumscribed area of ischemic atrophy, usually going from the pupillary edge to the sphincter and sometimes with rubeosis.

In nonproliferative retinopathy there may be asymptomatic lesions such as tortuous veins, "sun figure" pigmentations (black sunbursts, peripheral chorioretinal atrophy), "salmon

plaque" hemorrhages (pink superficial superficial hemorrhages), bright spots (deposits of peripheral hemosiderin, located inside the cystic cavities), peripheral arterioles in "silver thread", retinal breaks specially in the equator andangioidstreaks in rare cases.

Symptomatic lesions may be various occlusions of the central artery of the retina, macular arteriole, central vein of the retina and vascular occlusions of the choroid.

In the optic disk, it is common to see small transient red spots, caused by accumulation of sickle cells.

Figure1. *Retinography of a patient with sickle cell retinopathy, stage 4 with hemovitreous.*

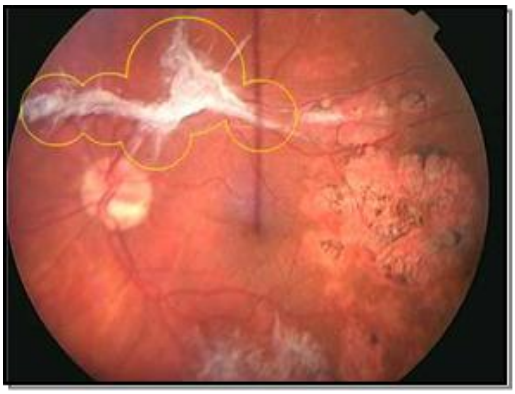

Figure2. *Image showing fibroglial tissue with vitreoretinal traction in Thalassemia*

FLOURESCEIN ANGIOGRAPHY

Generally in the posterior pole, the great vessels of the retina are normal, and may be tortuous, caused by peripheral arteriovenous anastomoses, sometimes encountering multiple obstructions of the perifoveal retinal capillary network.

In the first stage, we will observe dark areas of hypofluorescence due to the absence of vessels, especially in the extreme periphery of the retina and around these areas will be the amputated vessels in their path in whose obstructed limb we will see an accumulation of contrast. There will also be peripheral retinal hemorrhages that dissect the subretinal space, causing an accumulation of pigment that was already mentioned, the so-called "black sunburst sign". Stage two with the presentation of the anastomoses, in an attempt to revascularize the damaged regions, angiography will show hyperfluorescent vessels at the edge of the ischemic areas, unlike neovessels, they do not extravasate contrast. These anastomoses are obstructed in hypoxia crises and they will develop neovessels, which do provoke extravasation of contrast.

We can observe in stage three the neovascularization that was previously described and that we compare with the shape of a "fan" or a marine invertebrate, called Gorgonia Flabellum "be fan".

In the angiography we will see these neovessels constituted by a nourishing arteriole, a capillary network and a drainage venule. The process will become increasingly extensive.

The damaged retina is usually the superior temporal, followed by the superior nasal, inferior temporal and inferior nasal.

In the fourth stage, there will begin to be an adhesion of the neovascular vitreous complex with the provocation of hemorrhage by vascular traction, this being the first sign of disease with the subsequent formation of fibroglial tissue, with greater adherence of the neoformed vascular tissue.

In 60% of cases spontaneous occlusion of the neovessels occurs, unfortunately this is accompanied by neovascularization complicating the process.

The slow blood flow predisposes the formation of clots. And finally stage 5, is the traction of the vitreous adhered with the formation of tears and retinal detachment as already described above.

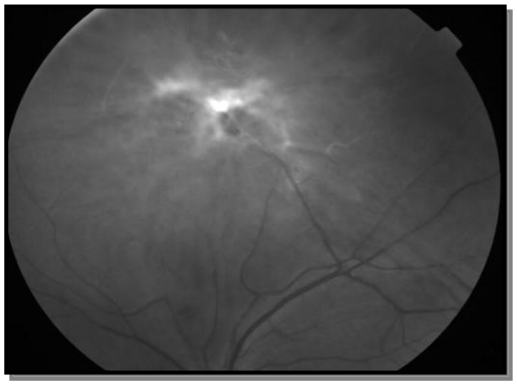

Figure3. *Fluorescein angiography of a patient with thalassemia, presenting stage 3 sickle cell retinopathy, where we can observe a peripheral neovascularization with the "Sea Fan" image.*

Figura4.*Retinographyshowinganhemorragestageprovokedbyretinopatysicklecells*

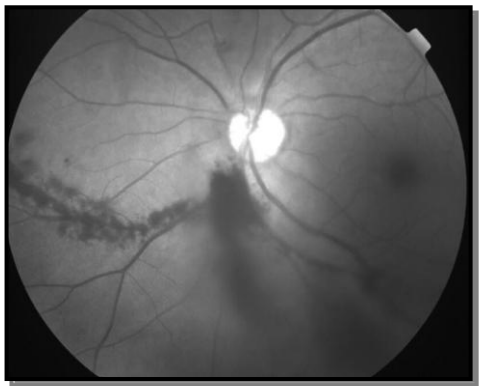

Figure5. *Fluorescein angiography of the previous image.*

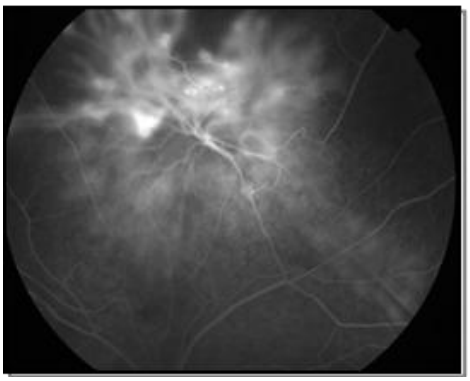

Figure6. *Another case of thalassemia, showingin this angiography a neovascular tuft,fed by a nutritious arteriole (feeder),giving an image "Sea Fan".*

TREATMENT

Argon laser photocoagulation with peripheral circular pattern in the retina, to treat the areas of absence of capillary perfusion, causes the regression of the lesions in a high percentage of cases.

The treatment must be divided, no more than 2 hours in each session. We must be careful during the treatment of non-photocoagulation directly the neovessels to avoid hemorrhage, and monitor the pulse of the central artery of the retina and branches because of the high risk of occlusion that exists during photocoagulation.

Photocoagulate all ischemic areas.

The power of the laser should be slight to avoid occlusion of the choroidal vessels and rupture of Bruch's membrane, when the retina is hyperpigmented.

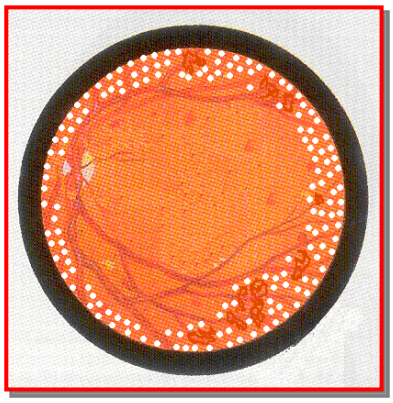

Figure7. *Sickle cell Retinopathy scheme treated with peripheral circular photocoagulation.*

Spot: 200-500um,Time: 0.2 s, Periphery: 360º

Figure8. *Fluorescein angiography, showing the peripheral scars by photocoagulation, following the treatment scheme*

It would be very important to close the neovascular membrane through the occlusion of the arterial feeder (afferent vessel), this can be associated with the destruction of the ischemic retina and never as an isolated procedure, since we would not be treating the cause and the consequence. Another option is peripheral cryotherapy, this being more aggressive we should leave this technique for cases in which the vitreous hemorrhage prevents us from visualizing the fundus.

The pars planavitrectomy in the detachment of the retina by traction and / or in persistent vitreous hemorrhage, sometimes the post-surgical result is not what we expect.

Sometimes the association of vitrectomy with the placement of a scleral buckle is necessary when there is an associated rhegmatogenous retinal detachment. In these cases always perform an ultrasound evaluation.

These patients are prone to have sickling crisis caused by the surgical act, general anesthesia, which can cause ischemia of the optic nerve and the macula with low vision as a consequence.

Despite all the treatments, the underlying disease persists and with the appearance of new neovessels, this is why our treatment must be carried out by areas and not in a localized way.

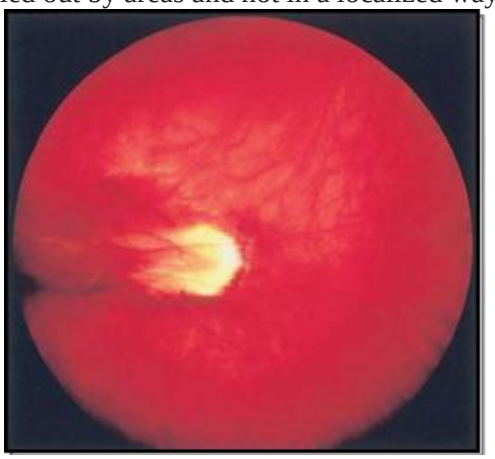

Figure9. *Retinography presenting traction vitreous in the Falloid Cell Retinopathy*

We will evaluate the patient with a fluorescein angiography in short times, to investigate if there is activity or not of the neovascular tuft and appearance of new ischemic areas.

REFERENCES

[1] Welch AB, Goldberg MF. Sickle-cell hemoglobin and its relation to fundus abnormality. Arch Ophthalmol.1966; 75:353-362.

[2] Moschandreou M, Galinos S, Valenzuela LR, Constantaras AA, Goldberg MF, Adams J. Retinopathy in hemoglobin C trait (AC hemoglobinopathy). Am J Ophthalmol 1974; 77:465-471.

[3] Condon PI, Serjeant GR. Behaviour of untreated proliferative sickle retinopathy. Br J Ophthalmol 1980, 64:404-411.

[4] ScottMcLeod D, Goldberg MF, Lutty GA. Dual perspective analysis of vascular formations in sickle cell retinopathy. Arch Ophthalmol 1993; 111:1234-1245.

[5] Condon, P., I.; Marsh, R., J.; Maude, G., H. *et al.*(1983). *Alpha Thalasemia and macular vasculature in homozygous sickle cell disease*. British Journal of Ophthalmology, 67, 779-781

[6] Jampol, L., E.; Condon, P., I.; Faver, M. *et al.*(1983). *A randomized clinical trial of feeder vessel photocoagulation of proliferative sickle cell retinopathy*, 90, 540-545.

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