Case Report: Application of adjuvant anti-VEGF in the management of Coats' Disease

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Introduction:

Coats' disease is an idiopathic retinal telangiectasia described by George Coats in 1908. It is a vascular disease with epidemiological predominance in men and in the first decade of life. It presents as telangiectasia associated with subretinal and intraretinal exudation, and may present with exudative retinal detachment. It is not associated with systemic diseases and does not present heredity, despite the possibility of genetic predisposition, since some patients have a somatic mutation in the NDP gene.

It presents clinically with low unilateral visual acuity, leukocoria and strabismus and infrequently with pain, heterochromia and nystagmus. In the evaluation of the posterior segment, aneurysmal arteriolar dilations and telangiectasias are observed, associated with exudate and extensive serous retinal detachment. Diagnosis is essentially clinical, however multimodal analysis with fluorescein angiography with initial hyperfluorescence of dilations and late uptake, as well as Optical Colorance Tomography (OCT) if possible, contributes to the diagnosis and management of Coats disease.

Methodology:

The information contained in this case report was obtained by reviewing the medical records of two patients, multimodal evaluation, documentary records, and literature review regarding the clinical picture, epidemiology, and clinical and surgical management of Coats' Disease.

Case report:

Patient 1 SM, 14 years old, student, from São Paulo sought ophthalmological assistance with complaint of low visual acuity (BAV) in left eye (OE) for 15 days. He had no relevant personal, ophthalmologic, or family history. On examination, visual acuity with correction (AVCC) was 20/25 in the right eye (RAS) and finger count at 50cm in the LAS. Anterior biomicroscopy without alterations and intraocular pressure of 14mmHg in both eyes. Fundoscopy of the FO, without alterations in the OD and in the OE showed peripheral telangiectasias, aneurysmal dilatations in the middle temporal periphery, diffuse exudation in the posterior pole and subretinal fluid in the foveal area, with a clinical diagnosis of Coats disease.

Patient 2 ASF, 9 years old, from São Paulo, was taken by her parents to the ophthalmologic service complaining of low visual acuity and alteration of the red reflex in the EO for 1 month. She had no personal or ophthalmologic history, and her father had a history of low visual acuity under clinical investigation. In the ophthalmological examination, he presented a CVA of 20/25 in the right eye and hand movement in the left eye. Anterior biomioscropy without changes and intraocular pressure of 12 mmHg in OD and 14mmHg in OE. Fundoscopy showed no changes in the right eye and in the left eye, peripheral telangiectasias, diffuse aneurysmal dilatations, exudation in the posterior pole and optic nerve, and the presence of a large amount of subfoveal subretinal fluid.

The retinography documented the findings of the FO of both patients (Figure 1 and 2). Angiofluoresceinography was performed with hyperfluorescence of dilations and leakage with late impregnation, associated with areas of hypofluorescence due to non-perfusion between the telangiectasias (Figure 2 and 3). OCT of the OE showed loss of foveal contour, exudate in external plexiform layer and extensive serous retinal detachment (RD) in macular region in both cases (Figure 4 and 5).

Faced with the diagnosis of Coats' disease, treatment was performed with argon laser photocoagulation and cryotherapy in the region of the peripheral lesions, together with intra-vitreous injection of vascular endothelial growth factor inhibitor (anti-VEGF) at the same time. During follow-up after the procedure, both patients show significant improvement of macular exudation and serous detachment, as well as showed improvement of OE AVCC from CD at 50cm to 20/30 in patient 1 SM and of hand movement to 20/400 after 1 month of approach.

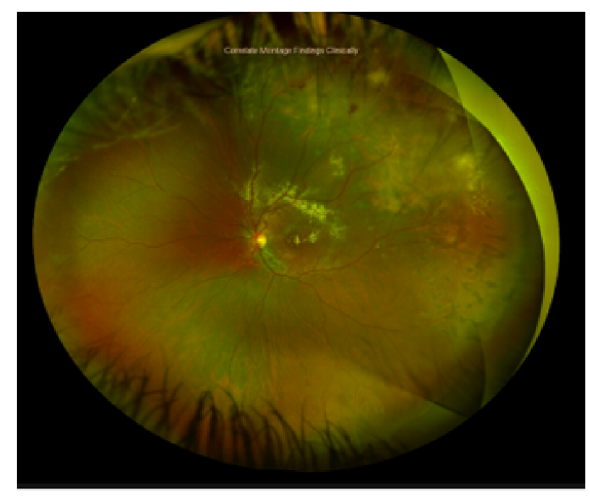

Figura 1. Retinografia OE - Paciente 1 SM

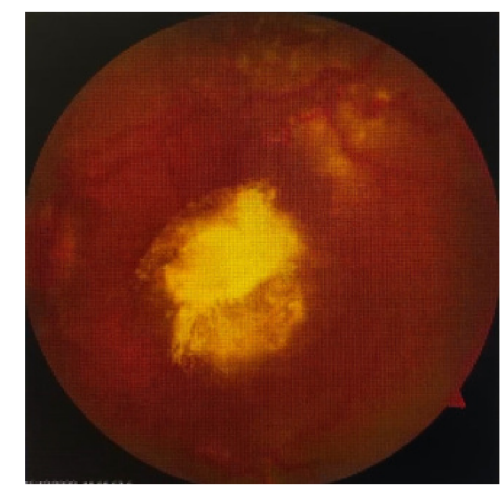

Figura 2. Retinopgrafia OE - Paciente 2 ASF

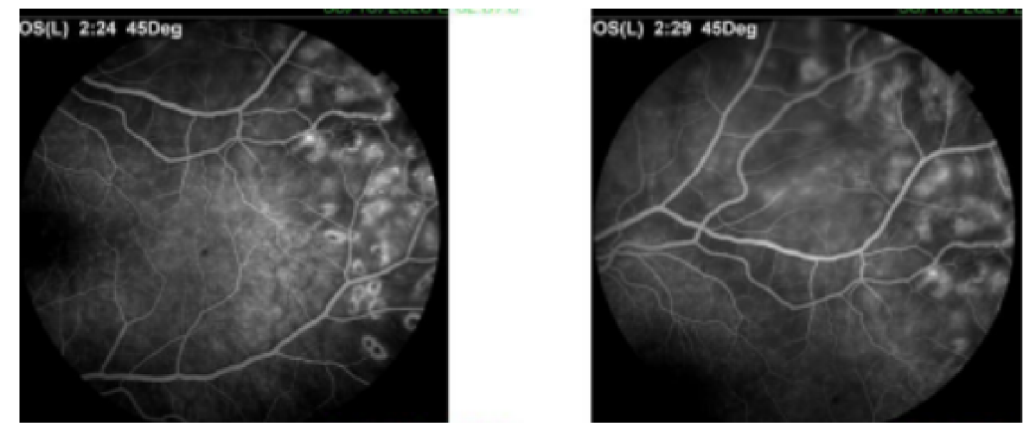

Figura 3. Angiofluoresceinografia- Paciente 1 SM

Discussion:

Coats disease is a disorder of retinal vasculature with peripheral retinal non-perfusion and high permeability of telangiectasias, resulting in macular edema, serous detachment and areas of non-perfusion. Thus, its pathophysiology involves retinal hypoxia and, consequently, stimulation of neovascularization, with a pathological role of elevated vascular endothelial growth factors.

The classic and well documented treatment involves observation in cases with mild disease without threat to the macula; photocoagulation and cryotherapy in stage 1 and 2 for treatment of telangiectasias. Few studies have been performed regarding treatment with anti-VEGF therapy, but initial results are promising regarding safety and efficacy, however long-term safety remains undetermined and needs further study. In stage 3 in which there is serous DR, the association of cryotherapy, photocoagulation and anti-VEGF appears to be effective and safe, resulting in improvement of retinal lesions and final visual acuity.

References:

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