WEGENER'S GRANULOMATOSIS ASSOCIATED TO UNILATERAL RETINAL VASCULITIS WITH OCCLUSIVE VASCULOPATHY AND SECONDARY MACULAR EDEMA: A CASE REPORT

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INTRODUCTION

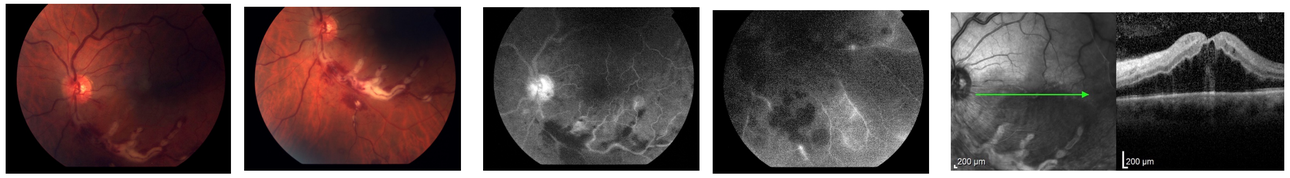
Wegener's granulomatosis is a multisystem inflammatory disease whose histopathology includes necrosis, granuloma formation and vasculitis. Vasculitis mainly affects small to medium-sized vessels. The reported incidence of ocular involvement varies from 50-60% of patients. Any structure of the eye can be affected, with orbital disease being the most common - secondary to nasal and paranasal sinus disease.

METHODS

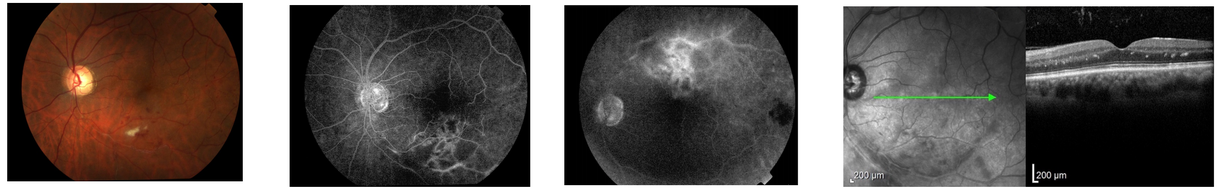
Clinical case description of a patient with unilateral retinal vasculitis associated with occlusive vasculopathy and secondary macular edema. During a complementary investigation of ocular disease, Wegener's granulomatosis was diagnosed.

CASE REPORT

JCSB, male, 55 years old, complaint of low visual acuity of the left eye for a week. He presented with a marginal corneal infiltrate associated with episcleritis for two months. Joint symptoms for four months, recurrent urinary infections and recent hospitalization for severe bilateral otitis. Denies skin lesions, dyspnoea or neurological symptoms. Ophthalmic examination: 20/20 visual acuity in the right eye and figures in the left eye. Normal biomicroscopy in the right eye, left eye: conjunctival hyperemia, endothelial fibrin, anterior chamber reaction 2 + / 4 +, absence of synechiae. Normal fundoscopy in the right eye, left eye: vascular sheath, retinal hemorrhages, extensive periflebitis in the lower vascular arch, alteration of the macular luster. Normal angiofluoresceinography in the right eye, left eye shows contrast leakage in the optic disc, staining and vascular leakage - perivasculitis - and areas of poor peripheral retinal perfusion. Macular optical coherence tomography: diffuse increase in retinal thickness, intra retinal cystoid spaces and subretinal fluid in the left eye. Laboratory tests: PCR, VHS and FR increased, HLA B27 negative. C-ANCA (1:80) and Anti-PR3 positive. EAS with significant proteinuria and hematuria. Infectious causes ruled out (herpes, cytomegalovirus, syphilis, toxoplasmosis, tuberculosis). Chest tomography showed a cavitated nodule in the right lower lobe and irregular solid nodules in the left lower lobe. Nuclear magnetic resonance of orbits with no anormalities. Ophthalmological diagnostic hypothesis: severe retinal vasculitis in the left eye associated with lower venous thrombosis and secondary macular edema. Systemic diagnostic hypothesis: ANCA vasculitis with ocular and renal involvement.



Pre-treatment. Retinography: periflebite and retinal hemorrhages in the lower vascular arch. Angiofluoresceinography: extravasation of contrast in an optic disc, hypofluorescence by blockage in areas of hemorrhage, hypofluorescence by vascular filling defect in areas of peripheral retinal ischemia. Macular optical coherence tomography: severe macular edema. Referred to hospital. Pulse therapy was performed with methylprednisolone 1 g / EV for 3 days and cyclophosphamide (Cyclops scheme). Oral prednisone (1mg / kg / day) in gradual remission. Intra-vitreous injections of anti-angiogenic - ranibizumab - three injections with monthly interval in the left eye. After intravenous pulse therapy, oral corticosteroids and injections of intravitreal angiogenic, he showed in visual acuity (20/50) in the affected eye, significant reduction in the retinal vascular inflammatory aspect and improvement of cystoid macular edema. improvement



After treatment. Retinography: vascular thinning, reduction of retinal hemorrhages and vascular inflammatory signs. Angiofluoresceinography: decreased contrast leakage in the optic disc and vessels, beginning of collateral formation. Macular optical coherence tomography: decreased foveal thickness and macular edema. Currently, he presents with retinal vasculitis in resolution and under control - using oral azathioprine, without the use of corticosteroids. He is being treated only for macular edema secondary to occlusive vasculopathy, maintaining the use of anti-angiogenic, when necessary, and treating ischemic retinal areas with photocoagulation with argon laser.

DISCUSSION

Wegener's granulomatosis can affect any system in the body, with involvement of the upper respiratory tract, lungs, kidneys and eyes being frequent. Retinal vasculitis with retinal artery or vein occlusion, chorioretinitis, macular edema, retinal detachment and retinal necrosis are rare and have poor visual prognosis. Combined treatment of cyclophosphamide and corticosteroids, has obtained good results in the remission of most patients, being recommended for more severe situations.

In the reported case, the patient had unilateral retinal vasculitis, retinal venous thrombosis and secondary macular edema. Therefore, he was in a severe stage of ocular and systemic disease, and a protocol with pulse therapy of cyclophosphamide and methylprednisolone was indicated. In addition, photocoagulation with argon laser and treatment with intra-vitreous anti-angiogenic was performed in the affected eye. After treatment, the patient improved of the systemic disease and retinal vasculitis.

KEY WORDS

Wegener's granulomatosis; Vasculitis; Retinopathy

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