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Posterior scleritis

Esclerite posterior

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Abstract

Posterior Scleritis (PS) is an inflammation of the posterior scleral segment, of low inflammatory or infectious etiology, with a rapid, progressive and irreversible evolution with severe visual impairment, especially if the diagnosis and treatment of (PS) are not performed in time skillful. Ocular pain, eye movement pain, headache and visual haze are the main signs and symptoms. It can be of idiopathic cause or associated with systemic disease in up to 45% of the cases. Rheumatoid arthritis is described as the most common association. Infectious causes may be present such as: Simple Herpes, Ophthalmic Herpes Zoster, Syphilis and Tuberculosis. Bilaterality can occur in up to 35% of cases being more prevalent in women from the fifth decade of life.

Keywords: *Scleritis; Sclera; Fundis oculi; Posterior segment of the eye; Pain; Female*

Resumo

Esclerite Posterior (EP) é uma inflamação do segmento escleral posterior, de etiologia inflamatória ou infecciosa, pouco diagnosticada, com evolução rápida, progressiva e irreversível com severo comprometimento visual, principalmente se o diagnóstico e o tratamento da (EP) não forem realizados a tempo hábil. A dor ocular, dor à movimentação ocular, cefaléia e embaçamento visual são os principais sinais e sintomas. Pode ser de causa Idiopática ou associada a doença sistêmica em até 45% dos caso. A Artrite Reumatóide é descrita como a associação mais comum. Causas infecciosas podem estar presente tais como: Herpes Simples, Herpes Zoster Oftálmico, Sífilis e Tuberculose. A bilateralidade pode ocorrer em até 35% dos casos sendo mais prevalente em mulheres a partir da quinta década de vida.

Descritores: Esclerite; Esclera; Fundo de olho; Segmento posterior do olho; Dor; Feminino

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Introduction

Posterior scleritis (PS) is an inflammation of the posterior scleral segment of inflammatory or infectious etiology which is poorly diagnosed and frequently treated late, and thus can compromise vision and lead to blindness.[1]

Ocular pain, eye movement pain, headache, and blurry vision are the main signs and symptoms. Anterior Scleritis (AS) is present in 43% of cases manifesting as hyperemia, photophobia, and tearing, being an important nosological symptom. It has a rapid, progressive and irreversible progression with severe visual impairment, especially if PS is not diagnosed and treated in a timely manner, causing catastrophic damage to noble eye structures. Bilaterality can occur in up to 35% of cases, and with frequent recurrence. The frequency of PS in childhood and young adults is low, being more prevalent in females aged between 40 and 59.[2]

The association between AS and PS is mainly in the diffuse form, and may be of idiopathic cause or associated with systemic disease in up to 45% of cases.[3,4]

Rheumatoid arthritis is described as the most common association. Infectious causes may be present such as Simple Herpes, Ophthalmic Herpes Zoster, Syphilis and Tuberculosis. Rheumatological diseases such as Systemic Lupus Erythematosus and Systemic Vasculitis as Wagner's Granulomatosis, Polyarteritis Nodosa, Giant Cell Arteritis, and inflammatory bowel diseases such as Ulcerative Colitis and Crohn's Disease.[5]

Differential diagnosis includes conditions such as Closed angle glaucoma, choroidal folds, optic disk edema, serous detachment of the retina, choroidal detachment, subretinal exudate, intraocular tumor, and uveal effusion.[6]

A multidisciplinary approach is recommended with the evaluation of the general practice and a good questionnaire of the systems and laboratory evaluation, in addition to the rheumatologist follow-up, which is very advisable for diagnostic cooperation and treatment.

Our objective is to describe a case of a 59-year-old female patient with posterior scleritis without systemic disease associated.

Case Report

IAR, female, 59 years old, Caucasian, started ocular pain in the right eye (RE) 1 month ago, and was examined in two different ophthalmological services by specialists, without precise diagnosis and being treated as chronic conjunctivitis and anterior uveitis.

At the ophthalmologic examination she had visual acuity of the RE LogMar 1.4, photomotor reflex preserved, ocular motility pain, aplanation tonometry at 16mmHg (BE). Normal biomicroscopy. The fundoscopy of the RE showed papillary hyperemia with justa-papillary haemorrhages, serous posterior pole detachment, presence of pigmentary alterations, and yellowish spots (Figure 1). Completely normal RE examination.

Ultrasound examination revealed a discrete posterior pole detachment; however, no alterations were observed as the T signal (Figure 2).

Optical Coherence Tomography (OCT) (Figure 3) revealed detachment of the sensorineural retina. In Angiofluoresceinography (Figure 4) papilledema and block hypofluorescence were observed suggesting edema at the level of the EPR/choriocapillar complex, and multiple points of dye extravasation from the choriocapillar.

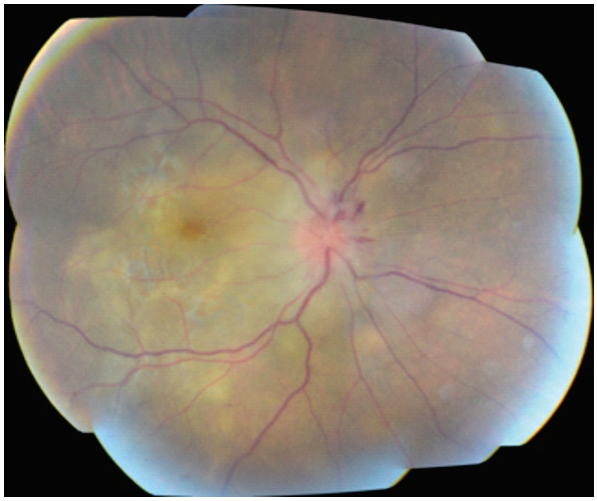

Figure 1: Initial aspect of the condition.

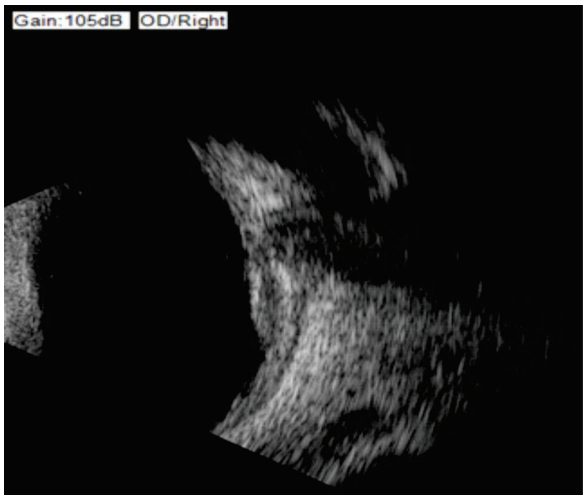

Figure 2: Evidence of a discrete detachment of a justa-papillary neuro-sensory retina without the T-Signal.

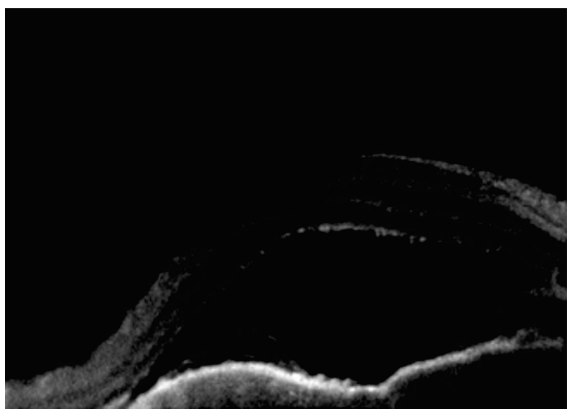

Figure 3: OCT revealing detachment of sensorineural retina.

Laboratory tests were requested, but they did not present conclusive alterations. Thus, treatment with corticosteroid therapy such as pulse therapy was started, with methylprednisolone 500 mg as the initial dose, and 125 mg every 48 hours for 10 days, with

a gradual reduction of the corticoid dose until its withdrawal with 20 days. After 1 month of treatment onset, the patient presented improvement of the clinical condition. The visual acuity was of LogMar 0.4 with treatment of the initial inflammatory process, both in the posterior pole and papilledema, with some posterior pole pigment alterations persisting (Figures 5, 6 and 7).

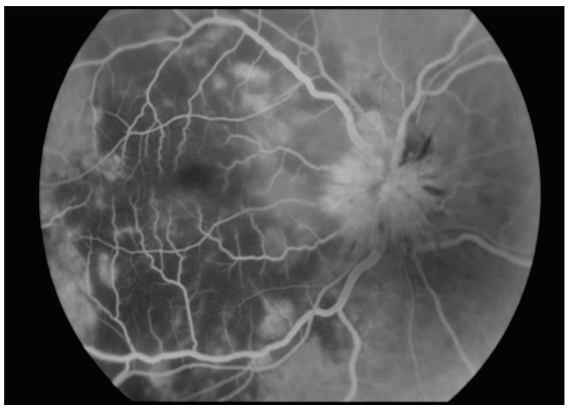

Figure 4: Initial fluorescence angiography. Note a large edema of deep layers.

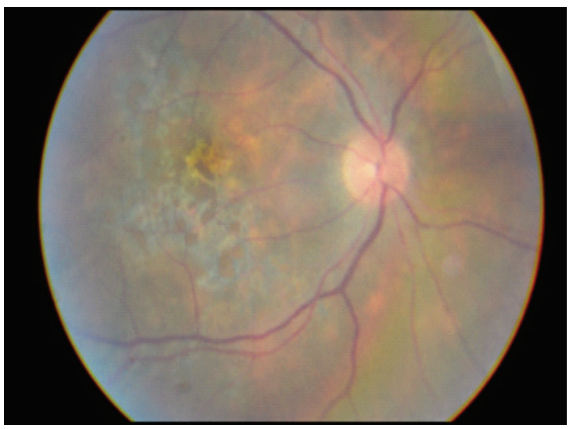

Figure 5: Note the absence of papilledema and resolution of inflammatory process.

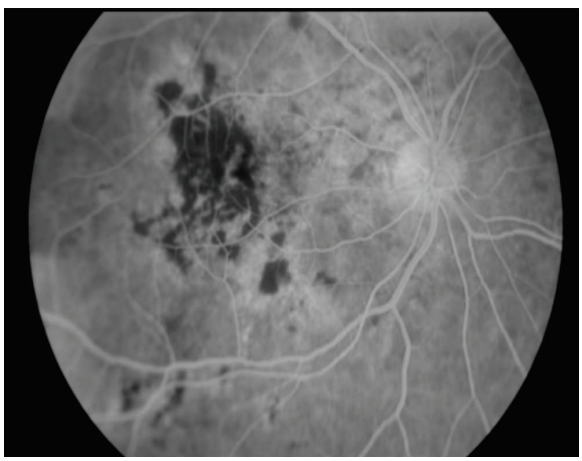

Figure 6: Fluorescence angiography showing pigmentary alterations in the posterior pole and absence of papilledema.

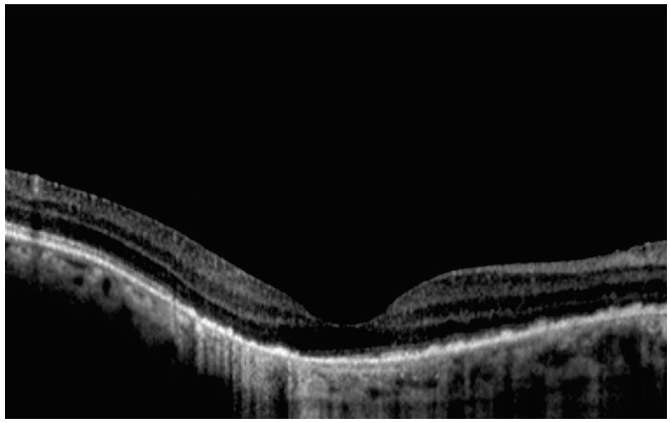

Figure 7: OCT showing total resolution of neuro-sensory retinal detachment.

Discussion

Regarding the classification of Posterior Scleritis according to Watson (1976), it is possible to divide it into five types: anterior diffuse, anterior nodular, anterior necrotizing without inflammation, anterior necrotizing with inflammation, and posterior. Nodular forms, when associated with thickening of the retina and choroid, may mimic intraocular neoplasms such as choroidal melanoma.[7]

In fundoscopy, PS may present with varied clinical signs, with serous retinal detachment, optic disc edema, and subretinal infiltrates being more frequent. Other findings are vitreitis, annular choroidal detachment, choroidal folds, subretinal mass, and whitish intra retinal deposits which are similar to hard exudates, besides subretinal exudation.

Ultrasonography (US) usually provides important data for diagnosis, especially in the absence of signs in the eye fundus. A very common alteration in this examination is sclerocoroid thickening (thickening of the posterior scleral wall) and the T signal (fluid inside the tenon capsule and optic nerve sheath determined by the dark T-like appearance on ultrasound.[8] However, the patient (US) did not reveal the classic "T" signal.

Treatment is based on the correct diagnosis, with oral corticosteroids (prednisone - 1mg/kg/day) being the main drug. In more severe cases and nonresponsive to conventional therapy, immunosuppressants (azathioprine or cyclosporine) are used. Long-term suppression is often necessary, in order to prevent relapses, aiming at better control of the ocular condition. Despite the aggressive condition, the correct treatment can avoid large visual losses, reducing the alterations of the ocular structures.[9,10]

Conclusion

Early diagnosis and prompt treatment of (PS), as well as knowledge about its clinical characteristics and forms of presentation are extremely important for a rapid diagnosis. As it is a disease with many differential diagnoses, it should not be forgotten as a cause of ocular inflammation, mainly because it is a potentially destructive disease.

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