

Simultaneous bilateral and unilateral recurrent posterior scleritis

Esclerite posterior bilateral simultânea e unilateral recorrente

Ana Paula da Silva Maganhoto¹, Sara Correia¹, Letícia Oliveira Squillace¹, Roberto Ivo Pasquarelli Neto¹

ABSTRACT

Posterior scleritis is an ocular inflammatory disease that affects the sclera e whose diagnosis is difficult. The main clinical manifestations are worsening of visual quality, ocular pain and finding in the posterior segment, such as serous retinal detachment, choroidal folds, optic disc edema and sclera thickening. We should award for systemic findings, because it is often accompanied by rheumatoid arthritis, systemic lupus erythematosus, inflammatory bowel disease and infections, such as syphilis and tuberculosis. This paper aims to describe two cases: one patient with simultaneous bilateral posterior scleritis without systemic alterations correlated with the disease. Bilateral presentation is uncommon in this pathology; and another case of recurrent unilateral anterior and posterior scleritis.

Keywords: Scleritis; Sclera; Posterior eye segment; Funds oculi; Retinal detachment; Case reports

RESUMO

Esclerite posterior é uma doença inflamatória ocular que acomete a esclera e cujo diagnóstico é difícil. A clínica da doença envolve a piora da qualidade visual com dor ocular e achados do segmento posterior, como descolamento de retina seroso, dobras de coroide, edema de disco óptico e espessamento escleral. Deve-se ficar atento aos achados sistêmicos, pois, muitas vezes, está associada a doenças como artrite reumatoide, lúpus eritematoso sistêmico, doença inflamatória intestinal e doenças infecciosas, como sífilis e tuberculose. Este trabalho objetiva descrever dois casos: uma paciente com quadro de esclerite posterior bilateral simultânea, sem alterações sistêmicas correlacionadas com a doença. A apresentação bilateral é incomum nesta patologia; e outro caso de esclerite anterior e posterior unilateral recorrente.

Descritores: Esclerite; Esclera; Segmento posterior do olho; Fundo de olho; Descolamento de retina; Relatos de casos

¹ Department of Ophthalmology, Santa Casa de Misericórdia de Santos, Santos, SP, Brazil.

The authors declare no conflict of interests.

Received for publication 07/12/2016 - Accepted for publication 22/10/2017.

INTRODUCTION

Posterior scleritis is an inflammatory ocular disease affecting the sclera and which is often underdiagnosed,⁽¹⁾ which may compromise the patients' final visual outcome.

This disease may occur alone or along with rheumatologic diseases, such as rheumatoid arthritis and systemic lupus erythematosus; infectious diseases such as tuberculosis and syphilis; and inflammatory bowel diseases.⁽²⁾

It is most commonly found unilaterally but can occur in both eyes simultaneously.⁽³⁻⁵⁾ It usually occurs with ocular pain, low visual acuity, serous retinal detachment, and optic disc edema.⁽³⁻⁶⁾

Its diagnosis is difficult to perform because the presentation and findings are not always classic. Currently, the clinical findings associated to findings of complementary exams, mainly ocular ultrasonography, are used. Other tests may be used to assist in the diagnosis of associated systemic diseases, as well as to exclude differential diagnoses.

Our objective is to describe the case of a 32-year-old patient with simultaneous bilateral posterior scleritis with no association so far to systemic diseases; and a 34-year-old patient with recurrent unilateral scleritis, also without associated systemic diseases.

CASE REPORT 1

A 35-year-old female patient, black, was admitted to the emergency service of ophthalmology at Irmandade Santa Casa de Misericórdia de Santos, Brazil, with headache and nausea for a month and low visual acuity (VA) in the left eye (LE) for a week ago, with discomfort in the ocular region. With no other systemic symptoms, joint pain, dermatological changes nor previous ocular surgeries. She presented systemic arterial hypertension as comorbidity, controlled with losartan 50 mg.

At the ophthalmologic exam, she presented VA in the right eye of 20/30, and in the LE of 20/200 according to the Snellen table. Biomicroscopy, direct and consensual pupil reflexes and swing light test, extrinsic ocular movement and normal ectoscopy in both eyes (BE). The intraocular pressure was 14 mmHg in BE.

Funduscopy showed papillary edema with hyperemia, 360° blurring and optic disk elevation, with peripapillary hemorrhages and choroidal folds in the macular region in the RE; LE with 360° papillary edema with hyperemia and border blurring, peripapillary haemorrhages and serous retinal detachment in the lower and temporal regions, partially affecting the macula.

The patient was hospitalized at the ophthalmology care for etiological investigation. We asked for laboratory tests, chest X-ray, opinion of the infectious team (to rule out infectious diseases) and neurology (for CEREBROSPINAL FLUID collection), as well as imaging tests.

Our main diagnostic hypotheses at the initial stage were malignant hypertension (personal history of hypertension), intracranial hypertension (due to bilateral papillary edema), Vogt-Koyanagi-Harada syndrome (bilateral papillary edema and serous retinal detachment) and posterior scleritis (papillary edema, serous retinal detachment and ocular discomfort).

Laboratory tests revealed normal blood count; negative serologies; positive inflammatory activity tests (PCR 13.01 mg/L and ESR 63 mm 1st hour); anti-ENA-SM antibodies, anti-JO-1, anti-RNP, anti-nuclear SSA (Ro), P-ANCA and C-ANCA negative;

computed tomography of the skull within the limits of normality and of orbits with internal contouring thickening of the posterior chamber of the left ocular globe in the posteromost quadrant and lateral to the left (Figure 1). Infectology ruled out infectious causes. CEREBROSPINAL FLUID was normal (clear, 0.3/mm³ leukocytes, glucose 60mg/dL, protein 38mg/dL, bacterioscopy and culture negative, opening pressure 56mmHg). Nuclear magnetic resonance of the skull was within normality, and of the orbits evidenced a hyperintense laminar area in T2 with 0.2 cm thickness in the posterior region of the vitreous chamber of the LE.

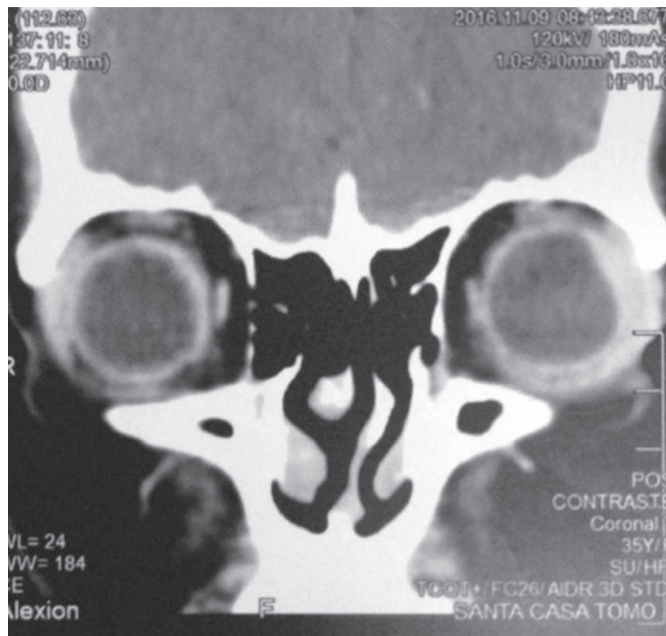


Figure 1: Tomography of the skull and orbits with thickening of the posterior chamber of the left eye in the posteromost quadrant and lateral to the left.

The first exams ruled out malignant hypertension, since the patient's blood pressure control ranged from 120x80 to 140x90 mmHg throughout the hospitalization; intracranial hypertension, because tomography was normal and CEREBROSPINAL FLUID presented opening pressure within normality; and Vogt-Koyanagi-Harada syndrome, because although the ocular findings were congruent with the syndrome, we did not have systemic findings present, such as pleocytosis in cerebrospinal fluid, dermatological and neurological changes (headache alone is not considered enough to fit the change criteria in the central nervous system). Thus, we only had bilateral posterior scleritis as the main hypothesis for the case.

Ultrasonography showed a choroidal thickening of the RE (Figure 2) in the lower wall of the nasal cavity, and serous retinal detachment of inferior wall with temporal wall choroidal thickening in the LE (Figure 3). These findings were congruent with our main diagnostic hypothesis. We did not perform angiofluoresceinography due to the unavailability of the exam during the follow-up period. We also disregard the hypothesis of Harada's disease (isolated ocular form), as it usually presents as a panuveitis, which was not the initial manifestation described in the case, as well as the sclerocoroid thickening of the ultrasound exam being more characteristic of scleritis (localized and greater than 2 mm) than that of Harada (where thickening is usually diffuse).

Thus, we reached the diagnosis of simultaneous bilateral posterior scleritis and promptly started treatment with methylprednisolone pulse therapy 1g/day for five days, followed by gradual reduction of oral corticosteroid. The patient showed reduction of the retinal detachment and improvement of final visual acuity, with 20/20 in RE and 20/40 in LE at hospital discharge. The patient is being followed up in an outpatient ambulatory with oral corticosteroid regression, and was referred for rheumatologic follow-up in order to investigate systemic diseases.

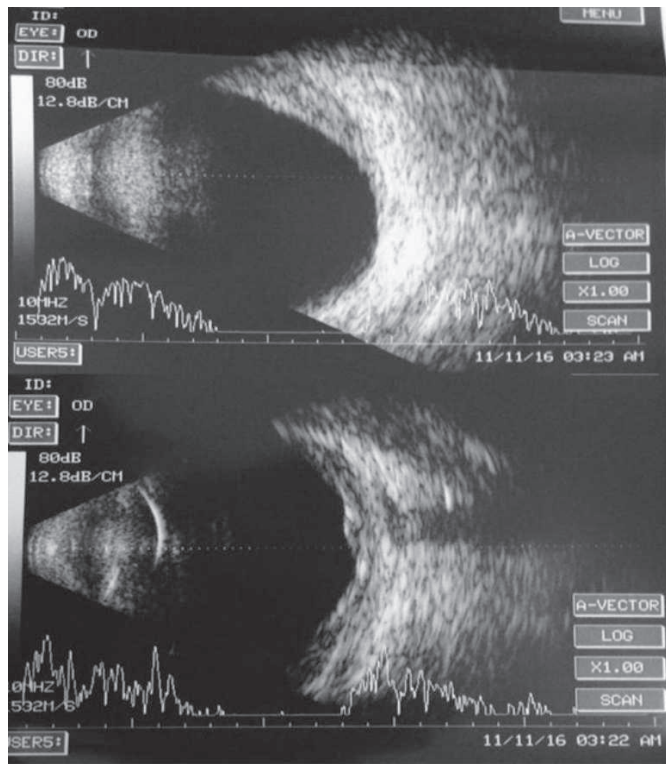


Figure 2: Ultrasonography of the right eye showing choroidal thickening in the nasal portion of the inferior wall.

CASE REPORT 2

AFF, female, brown, 34 years old, was admitted to the ophthalmology emergency department of Irmandade Santa Casa de Misericórdia de Santos complaining of pain and hyperemia in the right eye (RE) for 8 days, followed by low visual acuity (VA). The patient had no co-morbidities nor continuous-use medication.

She had a history of similar ocular episodes since 2007. She had previously been treated for central serosal chorioretinopathy in 2007, 2008 and 2013, and in 2014 she was diagnosed with anterior and posterior scleritis, being treated with indomethacin 50 mg every 8 hours, prednisone 20 mg/day topical ecorticoid.

The ophthalmological exam presented VA in the RE of finger counts (FC) at 1 meter, and of 20/20 in the left eye (LE) in the Snellen table. Biomicroscopy presented diffuse hyperemia without improvement after the test of phenylephrine in the RE; LE without changes. Intraocular pressure (IOP) of 10 mmHg in both eyes. Fundoscopy of the RE revealed diffuse retinal edema

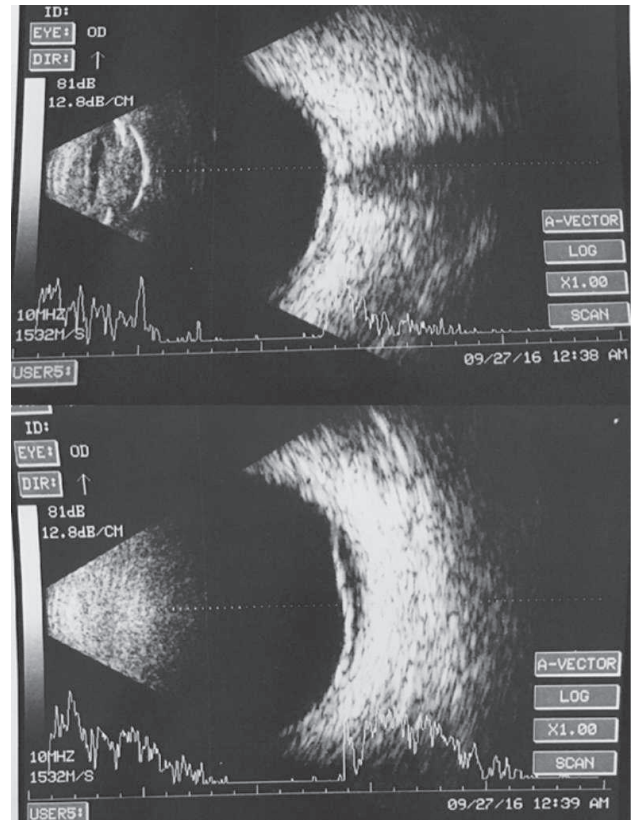


Figure 3: Ultrasonography of left eye showing serous retinal detachment in the inferior wall and choroidal thickening in temporal wall.

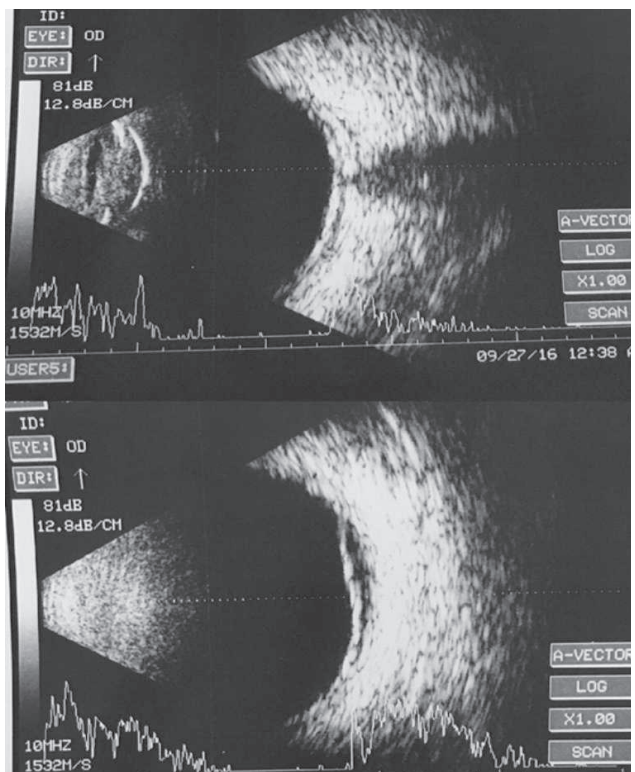


Figure 4: Ultrasonography of the right eye with the “T” sign and retinal detachment.

in the posterior pole, optic disc edema, choroidal folds, serous retinal detachment (RD) in the posterior pole; LE within the normal range.

We can infer the diagnosis of scleritis in the RE from the ophthalmologic findings and the patient's history. The patient was hospitalized at the ophthalmology care for complementary exams and therapy onset.

Laboratory tests and serologies were within the normal range (hemoglobin 13.6 mg/dL, leucocytes 12880 mg/dL, platelets 224000 mg/dL, C-reactive protein 2.51, VDRL and HIV 1 and 2 negative, toxoplasmosis IgG positive and IgM negative, cytomegalovirus IgG positive and IgM negative, FAN negative, rheumatoid factor negative), except for the erythrocyte sedimentation rate, which was above the reference value (40 mm in the 1st hour). Chest X-ray showed no changes. Computed tomography (CT) of the skull showed no abnormalities, and of orbits revealed parietal thickening of the right eyeball on the posterior projection, with apparent thickening of the choroid and retina and extension to the ipsilateral optic nerve. Ultrasonography (US) of the RE revealed the classic "T" sign, which indicates fluid in Tenon's capsule and optic nerve sheath, and inferior serous RD (Figure 4).

The patient received pulse therapy with methylprednisolone 1mg/day for five days, followed by regression with oral prednisone and indomethacin 50mg every 8 hours. Currently, we are undergoing ambulatory follow-up at our service, with VA of 20/35 in the RE and 20/20 in the LE, and funduscopy with decreased macular glow in the RE. She was referred for rheumatologic follow-up in order to investigate systemic diseases.

DISCUSSION

Scleritis is a scleral inflammatory disease, and can be divided according to Watson's 1976 classification into five types: anterior diffuse, anterior nodular, anterior necrotizing without inflammation, anterior necrotizing with inflammation, and posterior.^(1,2,5)

Posterior scleritis is the most uncommon form of this disease, found in 2 to 12% of cases.^(1,4,6) The average age of the patients is the fourth decade, and is more common in females.^(2,3-7) Involvement may be unilateral or bilateral, with unilateral being the most common form.⁽³⁻⁵⁾ In the study of Machado et al.,⁽³⁾ 73.9% of cases were unilateral and 26.1% of cases were bilateral.

Both cases reported in this study show posterior scleritis in middle-aged women, which correlates with the literature data. The patient in case 1 presented a simultaneous bilateral condition, which is not the most usual presentation of the disease; in case 2, the condition was anterior and posterior unilateral.

The most frequent symptoms are low visual acuity (VA), periocular / orbital pain, headache, and pain at ocular movement.^(1,3-6) Signs found on ophthalmologic exam include ciliary injection, anterior uveitis, optic disc edema, serous retinal detachment, retinal striae, subretinal infiltrates, choroidal folds, proptosis, limitation of extraocular movement.^(1,3-6)

The condition may occur alone or be associated with systemic or infectious diseases such as tuberculosis, herpes simplex or zoster, syphilis, rheumatoid arthritis, systemic lupus erythematosus, polyarteritis nodosa, Wegener's granulomatosis,

giant cell arteritis, and inflammatory bowel disease (ulcerative colitis and Crohn's disease).^(2-4,7) Therefore, there is a need to perform complementary tests to confirm or exclude such pathologies, as well as appointments with experts in the area. Among the exams, we counted CBC, X-ray of the chest, and FTA-ABS and VDRL as the initial screening.⁽⁵⁾ Further examinations and investigations should be requested according to the table presented by the patient.

Patients in our clinical cases had low VA, optic disc edema, and serous retinal detachment, which are commonly found in this disease. To date, there are no other systemic diseases related to this ocular condition. However, they were referred for rheumatologic ambulatory follow-up, as there are reports in the literature of a subsequent development of the diseases mentioned above.

The diagnosis of posterior scleritis is still a challenge. Clinical findings (intense ocular pain, serous retinal detachment, optic disc edema) are combined with complementary exams for the diagnosis to be confirmed. Ultrasonography showed the scleral thickening (classic, but not pathognomonic, "T" sign representing liquid in the Tenon's capsule and in the optic nerve sheath)^(1,2,4), sclerocoroid thickening (greater than 2 mm)^(3,4,6), and serous retinal detachment.

In case 1, the patient had clinical scleritis, which was confirmed after ocular ultrasonography, showing scleral thickening in the right eye and serous retinal detachment in the left eye; as well as exclusion of other differential diagnoses after the normal range of exams, such as computed tomography of the skull and cerebrospinal fluid. In case 2, the patient already had a history of the disease, a clinical characteristic, and an ultrasound exam with the classic T sign, making the diagnosis more evident.

Treatment includes the use of non-steroidal anti-inflammatory drugs and systemic corticosteroids from 60 to 80 mg / day, with gradual reduction of medication.^(1,2) In some cases, pulse therapy with methylprednisolone 1g / day for three days is necessary.⁽³⁾ In the case of recurrence of the disease with reduction of the oral medication, the use of immunosuppressive drugs may be of great value, such as cyclosporine and azathioprine.⁽³⁾

Scleritis may have severe complications such as permanent low visual acuity.⁽³⁾ Early diagnosis as well as the onset of therapy can promptly decrease visual sequelae.

Patients were treated with methylprednisolone pulse therapy 1 g / day for five days, followed by gradual reduction of oral corticosteroids. Both showed improvement of the ocular changes, as well as clinical improvement. Currently, they are being followed up in the ophthalmologic and rheumatologic ambulatory, without evidence so far of relapses or complications.

REFERENCES

1. Kellar JZ, Taylor, BT. Posterior scleritis with inflammatory retinal detachment. *West J Emerg Med.* 2015; 16(7):1175-6.
2. Machado DO, Curi ALL, Fernandes RS, Bessa TF, Campos WR, Oréfice F. Esclerite: características clínicas, associação sistêmica, tratamento e evolução de 100 pacientes. *Arq Bras Oftalmol.* 2009; 72(2): 231-5.
3. Machado DO, Curi AL, Bessa TF, Campos WR, Oréfice F. Esclerite posterior: características clínicas, associação sistêmica, tratamento e evolução de 23 pacientes. *Arq Bras Oftalmol.* 2009; 72(3):321-6.
4. Lavric A, Gonzalez-Lopez JJ, Majumder PD, Bansal N, Biswas J, Pavesio C, et al. Posterior scleritis: analysis of epidemiology, clinical factors, and risk of recurrence in a cohort of 114 patients. *Ocul Immunol Inflamm.* 2016;24(1):6-15.

5. Watson PG, Hayreh SS. Scleritis and episcleritis. *Br J Ophthalmol.* 1976;60(3):163-91.
6. Biswas J, Mittal S, Ganesh SK, Shetty NS, Gopal L. Posterior scleritis: clinical profile and imaging characteristics. *Indian J Ophthalmol.* 1998;46(4):195-202.
7. Watson P, Romano A. The impact of new methods of investigation and treatment on the understanding of the pathology of sclera inflammation. *Eye (Lond).* 2014;28(8):915-30.

Corresponding author:

Ana Paula da Silva Maganhoto
Dr. Claudio Luis da Costa, 50, Jabaquara - Department of
Ophthalmology, Hospital Irmandade de Santa Casa de
Misericórdia de Santos, Santos, SP, Brazil. ZIP Code: 11075-900
Phone N^o.: +55 13 3202-0600
E-mail: anamaganhoto@hotmail.com