



## Case report

## Central serous chorioretinopathy as a cause of vision loss in chronic relapsing inflammatory optic neuropathy

Miguel Raimundo<sup>a</sup>, Cristina Fonseca<sup>a</sup>, João Lemos<sup>b</sup>, Pedro Fonseca<sup>a,\*</sup><sup>a</sup> Department of Ophthalmology, Centro Hospitalar e Universitário de Coimbra, Praceta Mota Pinto, 3000-075, Coimbra, Portugal<sup>b</sup> Department of Neurology, Centro Hospitalar e Universitário de Coimbra, Praceta Mota Pinto, 3000-075, Coimbra, Portugal

## ARTICLE INFO

## Keywords:

Chronic relapsing inflammatory optic neuropathy  
Optic neuritis  
Central serous chorioretinopathy  
Steroids

## ABSTRACT

**Purpose:** Chronic relapsing inflammatory optic neuropathy (CRION) is a type of idiopathic recurrent optic neuritis that responds to systemic corticosteroids and relapses on steroid withdrawal or dose reduction. Central serous chorioretinopathy (CSCR) is often associated with glucocorticoid therapy. This paper aims to highlight CSCR as a cause of visual loss in patients being treated with corticosteroids for optic neuritis.

**Observations:** We describe the case of a 42-year-old woman with a history of CRION in her left eye who presented with painful vision loss in the right eye and diffuse right optic disc edema. Steroid therapy was initiated, leading to visual and perimetric improvement. Two months later however, the patient returned with painless visual loss, now related to CSCR. Despite oral steroids being continued, there was spontaneous tomographic and visual recovery after four months.

**Conclusions and importance:** We believe this is the first report of CSCR causing vision loss in a patient with CRION treated with oral corticosteroids. CSCR should be suspected in patients with optic neuritis of any cause who develop vision loss while on treatment with steroids.

## 1. Introduction

Chronic relapsing inflammatory optic neuropathy (CRION) is a form of idiopathic recurrent optic neuritis that responds promptly to treatment with systemic corticosteroids and relapses upon withdrawal or reduction of the dose of steroids.<sup>1–3</sup> Central serous chorioretinopathy (CSCR) is an underdiagnosed condition characterized by macular neurosensory retinal detachment and/or detachment of retinal pigment epithelium.<sup>4</sup> Traditionally, it has been associated with hypercortisolism, most commonly arising from glucocorticoid therapy.<sup>5,6</sup> We describe a previously unreported case of a CRION patient who presented with CSCR while she was being treated with steroids.

## 2. Case report

A 39-year-old woman was first admitted to our hospital with recurrent episodes of subacute deterioration of vision in her left eye (OS), accompanied by pain on eye movement, left relative afferent pupillary defect and left optic disc edema. Her visual acuity was 20/16 in the right eye (OD) and 20/70 OS. Standard automated perimetry showed a left inferior altitudinal defect.

A complete workup for optic neuritis was undertaken. Her complete blood count, hepatic, renal, and thyroid function, erythrocyte sedimentation rate, C-reactive protein, serum angiotensin-converting enzyme, antinuclear antibodies, anti-neutrophil cytoplasmic antibodies, toxoplasmosis screen, and cytomegalovirus, Epstein-Barr virus, herpes simplex virus, varicella zoster virus, Brucella, *Borrelia burgdorferi*, *Treponema pallidum*, QuantiFERON®-TB Gold assay, human immunodeficiency virus, and hepatitis B and C virus serology results were normal. NMO-IgG testing was negative and there were no oligoclonal bands on analysis of cerebrospinal fluid. Magnetic resonance imaging of the brain and orbits showed enlargement of the left optic nerve, consistent with left optic neuritis, but was otherwise normal. There were no pulmonary or interstitial lesions on chest computed tomography (CT) or gallium scans.

The patient was treated with intravenous and then oral methylprednisolone, showing prompt pain resolution and visual improvement. However, her vision deteriorated and her ocular pain recurred whenever the oral steroids were tapered, and a diagnosis of CRION was made.

The patient was lost to follow-up at that time but re-presented to the emergency department three years later, at the age of 42 years, with a

\* Corresponding author. Department of Ophthalmology, Centro Hospitalar e Universitário de Coimbra, Praceta Mota Pinto, 3000-075, Coimbra, Portugal.

E-mail addresses: [mglraimundo@gmail.com](mailto:mglraimundo@gmail.com) (M. Raimundo), [cristina\\_ffonseca@hotmail.com](mailto:cristina_ffonseca@hotmail.com) (C. Fonseca), [merrin72@hotmail.com](mailto:merrin72@hotmail.com) (J. Lemos), [pedroluisfonseca@gmail.com](mailto:pedroluisfonseca@gmail.com) (P. Fonseca).

<https://doi.org/10.1016/j.ajoc.2018.06.013>

Received 25 February 2018; Received in revised form 6 May 2018; Accepted 18 June 2018

Available online 20 June 2018

2451-9936/ © 2018 The Authors. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license

(<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

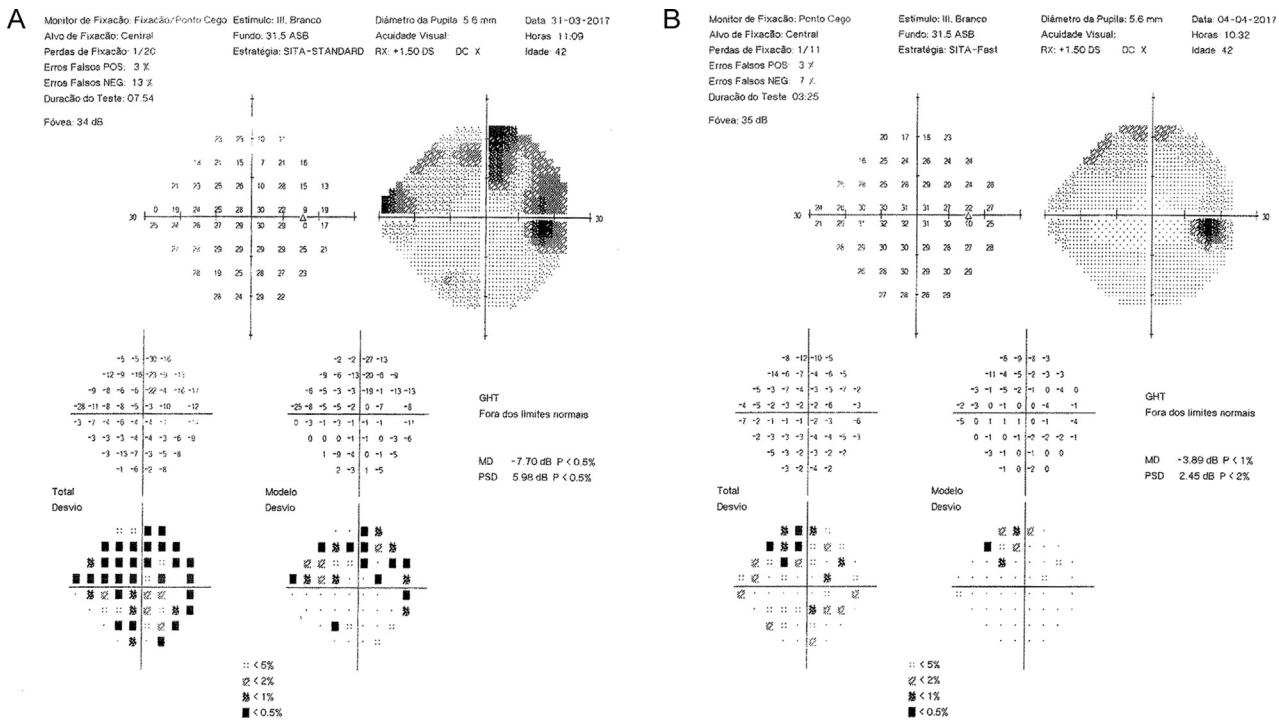


Fig. 1. Standard automated perimetry (24-2) during a relapse of CRION in the right eye, showing a superior arcuate defect(A), and after 3 days of intravenous methylprednisolone, showing marked perimetric improvement (B).

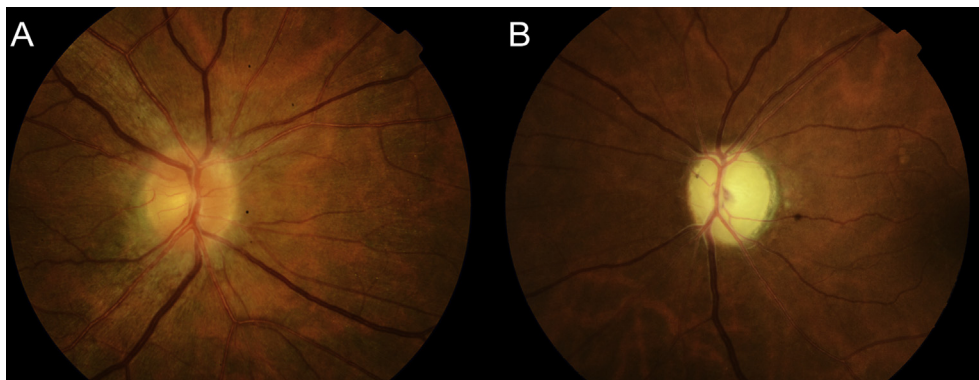


Fig. 2. Fundus photography during a relapse of CRION in the right eye, showing diffuse optic disc edema OD (A) and optic disc pallor OS (B).

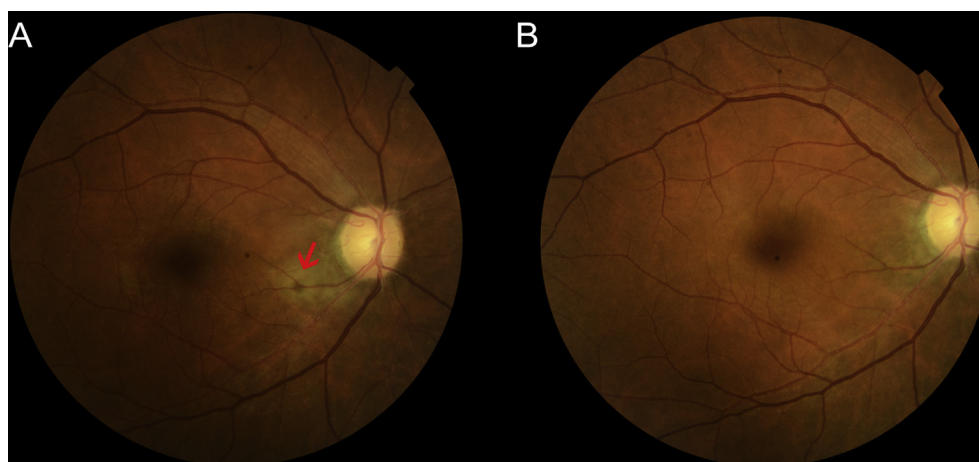


Fig. 3. T2-weighted magnetic resonance image of the orbits (coronal view). There is marked homogeneous hyperintensity of the right optic nerve (solid arrow) and an increased volume of cerebrospinal fluid surrounding the left optic nerve, consistent with optic neuritis OD and optic nerve atrophy OS (dashed arrow). R, right; L, left.

3-day history of painful vision loss in the OD. On examination, her visual acuity was 20/20 OD and her color vision was 17/17 Ishihara plates. Standard automated perimetry showed a superior arcuate defect

(Fig. 1A). She had no light perception OS and there was a relative afferent pupillary defect OS. Fundoscopy showed diffuse optic disc edema OD and optic disc pallor OS (Fig. 2). Neurologic examination and systems review were normal. Magnetic resonance imaging of the brain and orbits revealed thickening and enhancement of the right optic nerve and significant atrophy of the left optic nerve, but was otherwise normal (Fig. 3). The patient was admitted and treated with intravenous methylprednisolone. Within 3 days, she noticed prompt resolution of the pain and there was improvement on sequential standard automated perimetry (Fig. 1B). Optic disc edema improved after treatment with intravenous steroids and she was discharged and maintained on oral prednisone 60 mg/day on a slow tapering regimen, oral methotrexate 15 mg/week, folic acid 1 mg/day, and omeprazole 40 mg/day.

Two months later, the patient reported painless worsening of vision in her right eye. Visual acuity was 20/25 OD and there was no dyschromatopsia or optic disc edema OD. A shallow neurosensory macular detachment was suspected on fundoscopy (Fig. 4A) and confirmed by spectral-domain optical coherence tomography (SD-OCT, Fig. 5A). There were no other fundus abnormalities. Thus, a diagnosis of CSCR



**Fig. 4.** Fundus photography of the right eye showing a shallow neurosensory macular detachment (arrow) corresponding to acute CSCR (A), with spontaneous resolution 4 months later (B).

secondary to steroid therapy was made. As mentioned above, previous workup had been negative for underlying autoimmune or infectious diseases that might be complicated by optic neuritis and neurosensory macular detachment. Considering the previous history of CRION, treatment with high-dose oral prednisone was maintained after discussion of the therapeutic options with the patient. At the 2-month follow-up, her visual acuity OD improved to 20/16, there was no dyschromatopsia, but there was still residual subretinal fluid on SD-OCT imaging (Fig. 5B).

Four months following the onset of CSCR, the patient was asymptomatic. Visual acuity OD was 20/16, color vision was 17/17 and there were no fundoscopic abnormalities (Fig. 4B) or subretinal fluid on SD-OCT imaging (Fig. 5C).

### 3. Discussion

CRION is a severe form of recurrent optic neuritis that was first described in 2003.<sup>1–3</sup> A recent systematic literature review of 122 cases proposed five diagnostic criteria for this entity: a history of optic neuritis and at least one relapse; evidence of visual loss; seronegativity for NMO-IgG; contrast enhancement of the inflamed optic nerves on orbital imaging; and response to systemic immunosuppressive treatment and relapse upon withdrawal or dose reduction.<sup>2</sup> A diagnosis of CRION also requires exclusion of other causes of optic neuritis, such as optic neuritis due to multiple sclerosis, granulomatous optic neuropathy (sarcoidosis and TB) or infectious optic neuropathies. Therefore, careful history taking, examination and ancillary workup are warranted.<sup>2</sup>

Our report is unique in that our patient suffered CSCR-related visual loss during follow-up. We believe that changes in visual acuity should be investigated promptly because most are likely related to a relapse of optic neuritis. However, it is important to highlight CSCR as a potential cause of visual acuity loss in patients with any form of optic neuritis being treated with corticosteroids. Although the etiology of CSCR and the factors that trigger it are not clearly understood, this condition is frequently associated with the use of steroids.<sup>4,5</sup> Indeed, steroids ability to change the function of the ion pump in the retinal pigment epithelium and to modify the permeability of the blood-ocular barriers, may play a role in CSCR.<sup>6</sup> In addition, steroids increase platelet aggregation and thus increase blood viscosity which may potentially damage the choroidal circulation.<sup>6</sup> This and the fact that CSCR has never been associated with CRION, leads us to believe that in our patient the CSCR was secondary to corticosteroid use.

Acute CSCR is usually a self-limiting process and the standard initial approach is observation and modification of risk factors.<sup>6</sup> Given the association between CSCR and the use of corticosteroids, withdrawal of steroids is generally recommended.<sup>6</sup> Nevertheless, as shown in our patient, the need to treat CRION versus the need to taper corticosteroids in patients with CSCR can lead to a therapeutic dilemma. In the majority of typical acute cases, we believe it is advisable to allow for spontaneous resolution, but other therapeutic approaches may be considered to expedite reabsorption of fluid, such as laser photocoagulation and photodynamic therapy.<sup>6</sup> After discussing the benefits and risks of available therapeutic options with the patient, she agreed to continue oral prednisone and to be carefully monitored. Oral prednisone was maintained and spontaneous visual recovery and subretinal fluid resolution was observed after four months.

In conclusion, we report a case of CRION who presented with CSCR while under steroid treatment. Furthermore, we believe central serous chorioretinopathy may be underdiagnosed in the context of optic neuritis of any cause under steroid treatment and should be suspected in such patients who develop painless vision loss during follow-up.

### 4. Patient consent

The patient provided written consent allowing for anonymized clinical data collection and publication of the present case report.

### Acknowledgments and disclosures

#### Funding

No funding or grant support.

#### Conflicts of interest

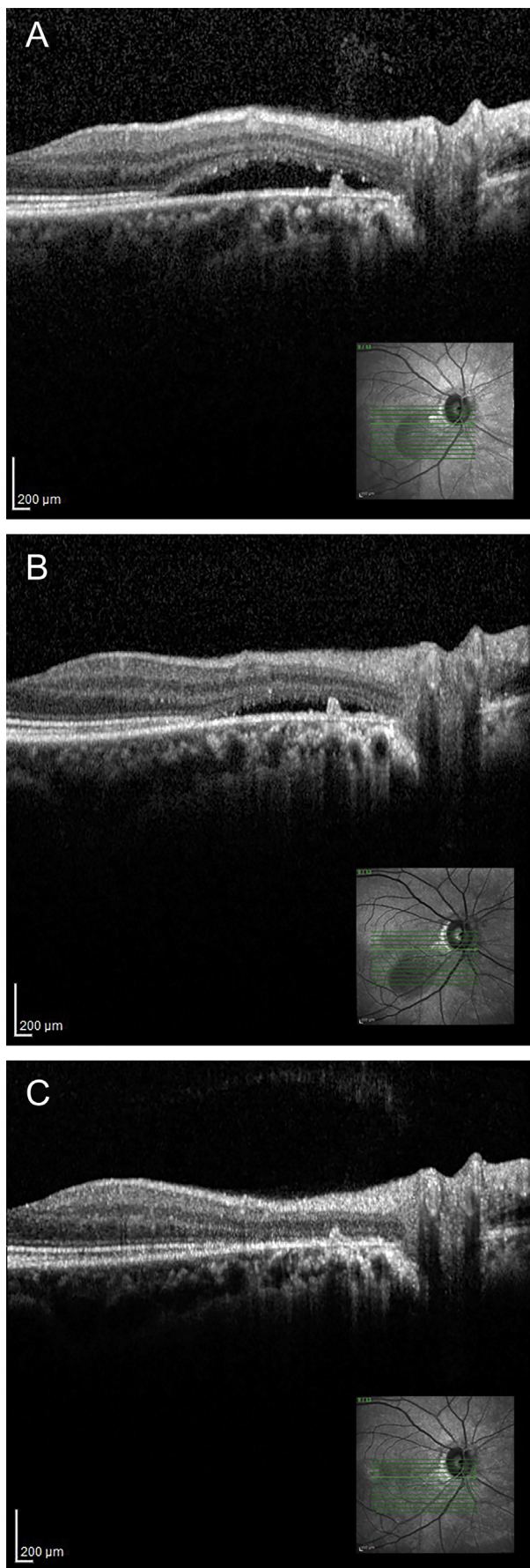
The following authors have no financial disclosures: MR, CF, JL, PF.

#### Authorship

All authors attest that they meet the current ICMJE criteria for authorship.

### Acknowledgments

None.



**Fig. 5.** Sequential spectral-domain optical coherence tomographic images of the periphery showing central serous chorioretinopathy. (A) Neurosensory retinal detachment nasal to the fovea. (B) Partial reabsorption of subretinal fluid after two months. (C) Complete resolution of central serous chorioretinopathy after four months.

**Appendix A. Supplementary data**

Supplementary data related to this article can be found at <http://dx.doi.org/10.1016/j.ajoc.2018.06.013>.

**References**

1. Kidd D, Burton B, Plant G, Graham E. Chronic relapsing inflammatory optic neuropathy (CRION). *Brain*. 2003;126(2):278–284.
2. Petzold A, Plant G. Chronic relapsing inflammatory optic neuropathy: a systematic review of 122 cases reported. *J Neurol*. 2014;261(1):17–26.
3. Petzold A, Plant G. Diagnosis and classification of autoimmune optic neuropathy. *Autoimmun Rev*. 2014;13(4–5):539–545.
4. Daruich A, Matet A, Dirani A, et al. Central serous chorioretinopathy: recent findings and new pathophysiology hypothesis. *Prog Retin Eye Res*. 2015;48:82–118.
5. Carvalho-Recchia C, Yannuzzi L, Negrão S, et al. Corticosteroids and central serous chorioretinopathy. *Ophthalmology*. 2002;109(10):1834–1837.
6. Nicholson B, Noble J, Forooghian F, Meyerle C. Central serous chorioretinopathy: update on pathophysiology and treatment. *Surv Ophthalmol*. 2013;58(2):103–126.