CASE REPORT

Papilloedema and Autoimmune Retinopathy from Systemic Lupus Erythematosus

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ABSTRACT

A 33-year-old female presented with bilateral papilloedema and constricted visual fields from autoimmune retinopathy. She then developed a painful peripheral neuropathy that led to further work-up and the diagnosis of systemic lupus erythematosus. Papilloedema and autoimmune retinopathy from systemic lupus erythematosus is a unique presentation.

ARTICLE HISTORY

Received 9 June 2017
Revised 11 July 2017
Accepted 11 July 2017

KEYWORDS

Autoimmune retinopathy;
idiopathic intracranial
hypertension; papilloedema;
pseudotumor cerebri;
systemic lupus
erythematosus

Introduction

Systemic lupus erythematosus (SLE) has various ocular manifestations. It is occasionally associated with papilloedema.^{1–7} Retinal ischaemic changes and vasculitis from SLE can occur as well,⁸ but only a single case of autoimmune retinopathy has been reported in the past.⁹ We report a patient who presented with both papilloedema and autoimmune retinopathy leading to a diagnosis of SLE.

Case report

A 33-year-old female presented with bilateral disc oedema and decreased peripheral vision. Her past medical history was significant for end-stage renal disease secondary to pre-eclampsia in 2002. She underwent a cadaveric kidney transplant in 2005. Rejection in 2012 resulted in removal of the transplanted kidney, and she has been on chronic haemodialysis since then. She has a history of depression and anaemia.

A few months before presentation, she reported “blurred vision.” She was found to have disc oedema and constricted visual fields. Magnetic resonance imaging/angiography/venography (MRI/MRA/

MRV) of the brain without contrast was unremarkable. She underwent a lumbar puncture with an opening pressure of 58 cm H₂O. She had a body mass index (BMI) of 28 kg/m². She was diagnosed locally with idiopathic intracranial hypertension (IIH) and started on acetazolamide 500 mg twice daily.

At presentation to the neuro-ophthalmology clinic, best-corrected visual acuity was 20/25 in the right eye and 20/40 in the left eye. There was no relative afferent pupillary defect. Dilated fundoscopic examination showed trace disc oedema in the right eye and grade II disc oedema in the left eye (Figure 1A). The arterioles were mildly attenuated. There was no bone spicule-like pigmentation in the periphery. The retinal nerve fibre layer thickness was elevated, left greater than right, on optical coherence tomography (OCT) (Figure 1B). Humphrey visual fields showed severe generalised constriction in both eyes (Figure 1C).

Goldmann visual fields (GVFs) revealed a ring scotoma in both eyes (Figure 2A). OCT of the macula showed loss of the ellipsoid zone in the parafoveal region, which corresponded to the ring scotoma (Figure 2B). Electroretinography (ERG)

Figure 1. Fundus photos demonstrate bilateral disc oedema, left greater than right (A). Optical coherence tomography (OCT) shows thickening of the retinal nerve fibre layer, left greater than right (B). 24-2 Humphrey visual fields show severe constriction of both eyes (C).

showed almost complete loss of the scotopic response and severe blunting of the photopic response (Figure 2C). Anti-retinal antibodies analyzed at the Ocular Immunology Laboratory at Casey Eye Institute were positive against 23 kDa that did not react to recoverin. They were also positive to 30-kDa (carbonic anhydrase II), 36-kDa (glyceraldehyde 3-phosphate dehydrogenase [GAPDH]), and 62-kDa proteins.

Two months later, she developed pain in her feet. This pain continued to worsen, and she then developed numbness and weakness in her feet and lower legs. An electromyogram revealed severe, subacute, length-dependent, axonal peripheral

neuropathy. A biopsy of the sural nerve showed a severely decreased density of myelinated fibres and several small mononuclear inflammatory cells, indicating an inflammatory or immune process. Work-up revealed an anti-nuclear antibody (ANA) of 3.4 U, anti-double-stranded deoxyribonucleic acid (DNA) of 104 IU/ml, Sjögren syndrome type A antigen (SSA) of 6.6 U, and ribonucleoprotein (RNP) of 1.9 U. Repeat lumbar puncture showed an opening pressure of 26 cm H₂O with a protein of 68 mg/dL but otherwise normal cerebrospinal fluid (CSF) constituents. Paraneoplastic antibody panel was negative. Vitamin B₁₂ and folate were normal at 536 ng/L

Figure 2. Manual visual fields demonstrate bilateral ring scotomas (A), which correspond to areas of ellipsoid photoreceptor loss in the parafoveal region on OCT (B). ERG shows bilateral severe reduction of both rod and cone function (C).

and $>20 \mu\text{g/L}$, respectively. Lyme, QuantiFERON-TB, syphilis, and human immunodeficiency virus were all negative. Serum electrophoresis was normal.

The work-up was consistent with SLE, which, in retrospect, may have been responsible for her prior kidney failure. She was started on intravenous (IV) methylprednisolone 1 g daily for 5 days followed by IV cyclophosphamide. Five-month follow-up on cyclophosphamide showed a dramatic improvement in the papilloedema (Figure 3). The visual fields remained severely constricted from the retinopathy.

Discussion

We describe a patient presenting with papilloedema and autoimmune retinopathy from SLE, which is a

unique combination. The improvement in the papilloedema in response to immunosuppressive treatment indicated that the raised intracranial pressure was due to the underlying SLE rather than idiopathic intracranial hypertension. There are multiple case reports and series demonstrating the association between SLE and intracranial hypertension, some due to venous sinus thrombosis or anaemia, whereas others are independent of these factors.^{1–7} It has been speculated that raised intracranial pressure from SLE is from thrombotic obliteration of the vascular bed of the arachnoid villi leading to decreased CSF absorption. Immune complex deposition or direct antibody injury to the arachnoid villi may also be playing a role.⁴

Interestingly, the majority of her visual loss was from the underlying autoimmune retinopathy. She

Figure 3. Five-month follow-up later demonstrates significant improvement in the disc oedema (A). Humphrey visual fields show severe constriction in both eyes, which is unchanged (B).

tested positive for multiple auto-retinal antibodies, including antibodies against carbonic anhydrase II and GADPH. Although these antibodies can be seen in normal patients,¹⁰ the clinical picture with subacute worsening constriction of visual fields, photoreceptor disruption, significant attenuated ERG, and stabilization with immunosuppression was consistent with autoimmune retinopathy.

The patient initially was referred for papilloedema and constricted visual fields, which were seen on automated perimetry. Tunnel vision from non-organic vision loss is not an uncommon referring complaint in patients with IIH.¹¹ However, Goldmann visual fields showed a ring scotoma, indicating true organic disease, and pointed toward retinal pathology, which was confirmed on OCT and ERG. Although retinopathy causing retinal haemorrhages and cotton wool spots is common from SLE,⁸ there is only a single prior case report of SLE-associated autoimmune retinopathy, in which the patient passed away from progressive multifocal leukoencephalopathy and had histopathological analysis of the eyes.⁹ This showed diffuse loss of photoreceptors, including in areas with normal retina and choroidal vasculature, which supports autoimmune damage to the retina that is independent of SLE-associated occlusive disease. They also demonstrated immunofluorescence staining of normal human retina when incubated with the patient's serum but did not characterise the

molecular weights, so we are unable to elucidate if the same auto-retinal antibodies were shared with our patient.

The combination of papilloedema and autoimmune retinopathy from SLE is rare and unique. They can possibly occur simultaneously in SLE because of the autoimmune nature of SLE, which may predispose a patient to autoimmune retinopathy, in conjunction with SLE's thrombotic or antibody-mediated compromise of the arachnoid villi that can lead to papilloedema.

Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the article.

Funding

This work was supported, in part, by an unrestricted grant to the Department of Ophthalmology by Research to Prevent Blindness, Inc., New York, New York, USA.

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