CASE REPORT

Visual loss from scleritis in C-ANCA-positive microscopic polyangiitis

Salah Alrashidi, Yacoub A Yousef, Hatem Krema

Princess Margaret Cancer Centre, Department of Ophthalmology and Vision Sciences, University of Toronto, Toronto, Canada

Correspondence to
Dr Hatem Krema,
htmkrm19@yahoo.com

SUMMARY

Scleritis is an inflammation of the outer eye coating that manifests with redness and ocular pain, and tends to be more severe when associated with a systemic collagen disease. Antineutrophil cytoplasmic antibody (ANCA)-positive microscopic polyangiitis is an autoimmune, multisystem, chronic disease characterised by damage to the endothelial lining of small vessels, and can be associated with severe scleritis that necessitates early aggressive therapy to prevent severe ocular complications.

BACKGROUND

We present a case of necrotising scleritis and loss of vision in a patient who has been misdiagnosed for 7 years as recurrent conjunctivitis, and was previously diagnosed with cytoplasmic antineutrophil cytoplasmic antibody (C-ANCA)-positive microscopic polyangiitis.^{1–3}

CASE PRESENTATION

A 77-year-old female patient was referred for ophthalmic evaluation of an intraocular mass, with significant loss of vision in the right eye over the previous 6 months. She suffered from intermittent pain and redness in the right eye for 7 years that used to be temporarily relieved with a course of prednisolone eye drops, on a presumptive diagnosis of conjunctivitis. On examination, best-corrected visual acuity was 20/300 in the right eye and 20/20 in the left. Intraocular pressure was normal in both the eyes. The right eye demonstrated tenderness on palpation without limitation of ocular motility. A slit-lamp examination showed diffuse conjunctival and episcleral vascular injection. Interestingly, there was a localised area of conjunctival and episcleral tissue loss of 1×1 mm in base diameter at the inferior conjunctival fornix, with erosion of the exposed sclera (figure 1A). An ocular fundus examination revealed dense vitreous cells associated with inferior extensive retinal detachment (figure 1B). Underneath the detachment, there was an ill-defined non-pigmented subretinal diffuse lesion involving the lower half of the fundus. The left eye examination was unremarkable.

INVESTIGATIONS

B-scan ultrasonography of the right eye revealed that the subretinal lesion had a marked thickening of the sclerochoroidal complex that extended between the 3 and 10 o'clock meridians, associated with retinal detachment and vitreous opacities (figure 1C).

Figure 1 (A) Marked conjunctival inflammation with an area of conjunctival and episcleral tissue necrosis (arrow) with erosion of sclera. (B) The fundus image showing hazy fundus view from associated dense vitreous cells with inferior retinal detachment (arrow). (C) Ultrasonography image showing marked thickening involving sclera and choroid (arrows) with an overlying retinal detachment and vitreous cells.

DIFFERENTIAL DIAGNOSIS

The clinical and imaging findings suggested necrotising scleritis, inducing exudative retinal detachment,

To cite: Alrashidi S, Yousef YA, Krema H. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-009583

which accounts for her vision loss. On further questioning about possible systemic associations, we obtained a history from her referring physician of C-ANCA-positive microscopic polyangiitis, which manifested as severe anaemia, recurrent acute renal impairment and polyarthropathy that used to be controlled by short courses of systemic steroids and cyclophosphamide. Possible opportunistic infections, such as tuberculosis, secondary to immunosuppressants' use were ruled out.

TREATMENT

The patient was treated with a combination of 20 mg/day of oral prednisolone and 50 mg/day of azathioprine.

OUTCOME AND FOLLOW-UP

After 4 months, the redness and pain resolved and the conjunctiva healed over the necrotic sclera (figure 2A). However, there was no improvement in vision despite subtotal resolution of

retinal detachment due to persistence of macular oedema (figure 2B). Scleral thickness showed no change with ultrasonography (figure 2C).

DISCUSSION

The three major ANCA-associated vasculitides are granulomatosis with polyangiitis, previously known as Wegener's granulomatosis, Churg-Strauss syndrome and microscopic polyangiitis, which can be widespread resulting in multiorgan failure.^{3–5} Our patient has been diagnosed with ANCA-associated microscopic polyangiitis that affected the eye with recurrent episodes of pain and redness for 7 years, without being correctly diagnosed as scleritis, until she presented recently with severe ocular inflammation and vision loss.

ANCA-positive vasculitis has been reported to produce scleritis in 75% of patients, in addition to recurrent inflammation in other ocular and periocular structures including the orbit, nasolacrimal system, conjunctiva, cornea, uvea and the retina.^{2–3} The non-necrotising anterior scleritis is the most frequent subtype that can be clinically misdiagnosed as conjunctivitis, if the associated systemic vasculitis is not considered for pathogenesis of the ocular presentation. The more aggressive necrotising subtype, which manifested in our patient, affects approximately one-quarter of the vasculitis-induced scleritis patients and can lead to loss of the eye.⁶

The visual loss in our patient is due to the development of longstanding extensive exudative retinal detachment secondary to marked choroidal inflammation associated with the recurrent episodes of necrotising scleritis. The tissue loss observed at the conjunctival fornix is a consequence of the systemic small vascular inflammatory process, which causes focal ischaemic necrosis.^{2–6} The scleritis in this patient used to be previously misdiagnosed as simple conjunctivitis, which was insufficiently treated by repeated courses of topical steroids that caused temporary resolution of the ocular symptoms.

The use of systemic immunosuppressive therapy is needed in such cases to control the disease and the associated necrotising scleritis at an early stage,⁶ and usually a combination of steroids and immunosuppressants is needed for better disease control and survival improvement.⁷

Figure 2 (A) Resolution of conjunctival inflammation and healing of conjunctiva over an area of scleral thinning. (B) Subtotal resolution of retinal detachment with persistence of vitreous cells and macular oedema. (C) Scleral thickness showing no changes after treatment.

Learning points

- ▶ Early ocular assessment in patients with systemic vasculitis should be undertaken to rule out the presence of associated scleritis.
- ▶ A slit-lamp examination with the aid of ocular ultrasonographic imaging can readily differentiate conjunctivitis from scleritis.
- ▶ Early treatment of cytoplasmic antineutrophil cytoplasmic antibody-positive polyangiitis patients with systemic steroids and immunosuppressants could prevent eventual vision loss.

Contributors HK and SA contributed to the concept and design, and interpretation of the results. HK, YAY and SA were involved in writing up the manuscript and approved the final draft.

Competing interests None.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Okhravi N, Oduguwa B, McCluskey P, *et al.* Scleritis. *Surv Ophthalmol* 2005;50:351–63.
- 2 Hoang LT, Lim LL, Vaillant B, *et al.* Antineutrophil cytoplasmic antibody-associated active scleritis. *Arch Ophthalmol* 2008;126:651–5
- 3 Watkins AS, Kempen JH, Choi D, *et al.* Ocular disease in patients with ANCA-positive vasculitis. *J Ocul Biol Dis Inform* 2009;3:12–19.
- 4 Harman LE, Margo CE. Wegener's granulomatosis. *Surv Ophthalmol* 1998;42:458–80.
- 5 Straatsma BR. Ocular manifestations of Wegener's granulomatosis. *Am J Ophthalmol* 1957;44:789–99.
- 6 Watson PG, Hayreh SS. Scleritis and episcleritis. *Br J Ophthalmol* 1976;60:163–91
- 7 Sainz de la Maza M, Foster CS, Jabbur NS, *et al.* Scleritis associated with systemic vasculitic diseases. *Ophthalmology* 1995;102:687–92.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow