Behçet's disease presenting as bilateral occlusive retinal vasculitis in a young woman

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DESCRIPTION

A 41-year-old white European woman presented with a 4-week history of painless floaters in her left eye and best-corrected visual acuity (BCVA) of 6/6 and 6/30 in right and left eyes, respectively. She had no relevant medical history and was a non-smoker. She reported orogenital ulceration, erythema nodosum, left hand weakness and arthralgia.

Examination showed no anterior segment inflammation or rubeosis. Intraocular pressures were 16 mm Hg bilaterally. A left relative afferent pupillary defect was present with +2 vitritis, retinal periphlebitis and inferotemporal branch retinal vein occlusion with macular oedema (figure 1A). Widefield fluorescein angiography (figure 1B) confirmed extensive retinal ischaemia with neovascularisation and collateral blood vessels. Optical coherence tomography confirmed macular oedema (figure 1C).

Serology for rheumatoid factor, anticyclic citrullinated peptide antibodies, antinuclear factor and infectious aetiologies (*Mycobacterium tuberculosis*, *Borrelia burgdorferi*, *Treponema pallidum* and *Toxoplasma gondii*) was negative. MRI brain revealed cerebral vasculitis. Biopsy of the genital ulceration showed leukocytoclastic vasculitis. Human leucocyte antigen (HLA)-B51 was negative but a diagnosis of Behçet's disease (BD) was made in accordance with International Study Group for Behçet's Disease criteria (ISGBD). She was started on oral prednisolone (50 mg daily) and tacrolimus (2 mg two times per day); however, 11 months later, she had a flare comprising arthritis, cerebral vasculitis and right eye uveitis (figure 2A). Systemic therapy was changed to mycophenolate mofetil and adalimumab with intravitreal antivascular endothelial growth factor and sectoral pan-retinal photocoagulation (PRP) to both eyes. With this immunosuppression regimen, she achieved remission and her BCVA remained 6/6 in both eyes. However, further PRP was required for progressive retinal ischaemia.

BD is an idiopathic multisystem inflammatory disorder, prevalent along the Silk Road (15–420:100 000), primarily affecting eyes, skin, joints and central nervous system vasculature.[1] Peak onset is in the third decade; with no gender predilection.[2] There is ocular involvement in 70%, with 16.6% of all-cause uveitis attributed to BD.[2 3] Hallmark features include oral ulceration (98%), genital ulceration (80%–87%), erythema nodosum (50%), maculopapular rash (45%–90%), positive pathergy test (8.6%–70%) and ocular inflammation, three of which (particularly recurrent oral ulceration) are required by ISGBD to make a BD diagnosis.[4 5] HLA-B51 is positive in only 60%.[6] Classic ophthalmic features are acute anterior uveitis in a white eye with shifting hypopyon, vitritis, occlusive retinal vasculitis, optic disc oedema and retinal neovascularisation.[7] Sequelae include cataract, glaucoma, optic atrophy and vascular attenuation in end-stage disease.[8] Treatment aims to induce remission, preventing progressive occlusive retinal vasculitis. Acute sight-threatening uveitis is amenable to high-dose glucocorticoids and/or biological agents (eg, infliximab or interferon alpha).[9] Remission of posterior uveitis in BD should be maintained with systemic immunosuppression and may require antimetabolites (eg, methotrexate, azathioprine, mycophenolate), calcineurin inhibitors (eg, ciclosporin,

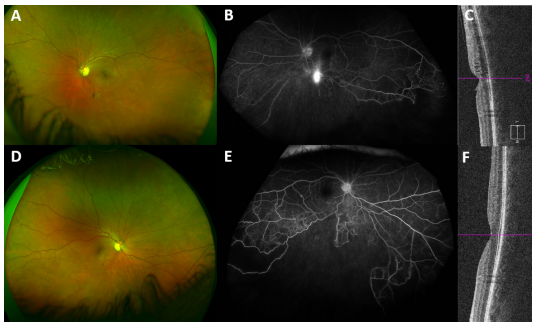

Figure 1 (A) Widefield fundus photograph (Optos 'California', Optos plc, Scotland) of the left eye showing occlusive vasculitis of the inferortemporal retinal vessels. (B) Widefield late phase fluorescein angiogram (FFA) of the left eye confirming extensive ischaemia in the inferior retina with focal leakage from neovascularisation. (C) Macular optical coherence tomography (OCT, Cirrus 5000, Carl Zeiss Meditec, Dublin, USA) scan showing intraretinal oedema. (D) Widefield fundus photograph of the right eye showing similar ischaemia 11 months later. (E) Widefield FFA showing marked ischaemia of the inferior retina. (F) Resolution of left eye oedema on OCT following treatment.

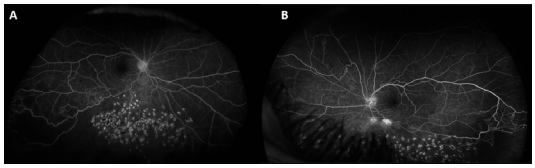

Figure 2 Follow-up widefield FFAs of the right (A) and left (B) eyes showing photocoagulation burns in the inferior retina with progression of retinal ischaemia secondary to occlusive vasculitis and prominent peripheral collateral vessels. FFAs, fluorescein angiography.

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tacrolimus) and/or biological agents (eg, adalimumab, infliximab).[9] Rituximab has been used for refractory ocular BD with promising short-term results; however, longer follow-up studies are required.[10] Unilateral exacerbations may benefit from intravitreal corticosteroid therapy adjunctive to systemic treatment.[11] Despite treatment, legal blindness (ie, BCVA ≤6/60) occurs in 13% of eyes of patients with BD, typically due to ischaemic maculopathy.[12]

Learning points

- Behçet's disease (BD) is an important diagnosis with a 5% mortality rate by 5–10 years, typically due to vascular events (cardiac or cerebral). Ophthalmic presentation may allow early diagnosis of BD, facilitating treatment and preventing debilitating systemic sequelae.
- Ophthalmic involvement in BD may lead to serious visual morbidity in young patients. Early diagnosis may prevent bilateral blindness and the accompanying loss of quality of life and economic contribution.
- BD should always be considered in the differential diagnosis of any ophthalmic or systemic vasculitis even in areas of low prevalence.

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