Cilioretinal artery vasculitis as the initial ocular manifestation of Behçet's disease

Dear Editors,

Behçet's disease (BD) is a multisystem inflammatory disorder characterised by obliterative vasculitis involving vessels of all calibres. Ocular involvement, most commonly presenting as non-granulomatous uveitis and retinal vasculitis, represents a major cause of visual morbidity (1). Retinal venous occlusions are well recognised in BD, whereas primary involvement of the cilioretinal artery has rarely been reported as an initial ocular manifestation (2). We report a patient with cilioretinal artery vasculitis as the first ocular sign of BD, with complete visual recovery after early immunosuppressive therapy and stable long-term follow-up. Written informed consent was obtained from the patient for publication of anonymised clinical data and images.

A 37-year-old man presented with sudden, painless visual loss in the left eye for one day. He reported recurrent oral aphthous ulcers and papulopustular skin lesions for two years. Best-corrected visual acuity was 20/20 in the right eye and counting fingers at 1 meter in the left eye. Slit-lamp examination revealed mild anterior chamber inflammation (1+ cells) with vitreous cells bilaterally. Fundus examination of the left eye showed retinal haemorrhages and whitening along the distribution of the cilioretinal artery, while the right eye was normal. Fundus fluorescein angiography demonstrated focal vascular leakage strictly confined to the cilioretinal artery territory; there was no evidence of peripheral fern-like leakage or other vascular abnormalities, with the peripheral retina remaining completely normal. Optical coherence tomography revealed inner retinal oedema consistent with acute arterial ischaemia (3) (Fig. 1). Systemic evaluation confirmed Behçet's disease according to both the 2014 International Criteria for Behçet's Disease (ICBD) and the 1990 International Study Group (ISG) criteria (4, 5).

Treatment was initiated promptly with oral prednisone (48 mg/day, tapered over three months), azathioprine, and colchicine. Visual acuity improved to 20/20 within three months, accompanied by complete anatomical and angiographic resolution. No ocular relapses occurred during follow-up (Fig. 1). The patient was followed regularly for four years after the initial presentation, with scheduled ophthalmic evaluations and no ocular relapses during this period. At the most recent examination, visual acuity remained stable at 20/20, and there was no clinical or imaging evidence of recurrent inflammation. The patient continues azathioprine therapy (150 mg/day) without ocular complications.

However, bilateral femoral head avascular necrosis was diagnosed by magnetic resonance imaging 18 months after the initiation of treatment. Total hip arthroplasty was performed on one side, and surgical intervention is planned for the contralateral hip. The development of avascular necrosis is likely related to cumulative corticosteroid exposure and/or BD-associated microvascular pathology (6). This complication underscores the systemic consequences of BD and the need for close rheumatologic monitoring alongside ophthalmic follow-up.

This case expands the spectrum of arterial involvement in BD, demonstrating that cilioretinal artery vasculitis may represent the initial ocular manifestation. Early and aggressive immunosuppressive therapy can result in complete and sustained visual recovery (3). Nevertheless, the occurrence of avascular necrosis underscores the importance of minimising corticosteroid exposure and implementing steroid-sparing strategies in BD management (7). Prompt recognition of atypical ocular presentations, combined with early systemic treatment, is essential to preserve vision and limit long-term morbidity in BD.

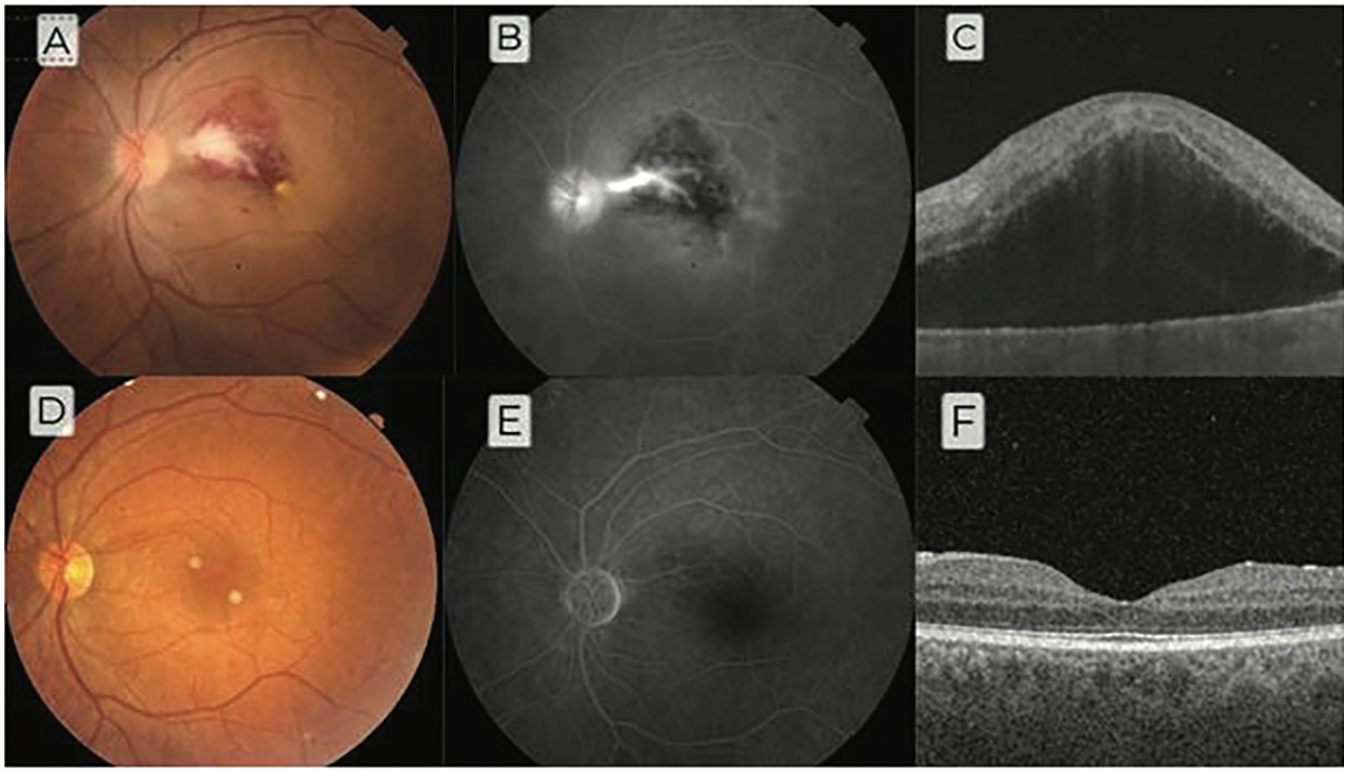


Fig. 1. Multimodal retinal imaging at presentation and at 4-year follow-up.
A. Colour fundus photograph at presentation showing retinal whitening and intraretinal haemorrhage along the distribution of the cilioretinal artery in the left eye. **B.** Fundus fluorescein angiography at presentation demonstrating focal vascular leakage confined to the cilioretinal artery territory without peripheral vasculitis. **C.** Optical coherence tomography at presentation revealing hyperreflective thickening of the inner retinal layers consistent with acute arterial ischaemia. **D.** Colour fundus photograph at 4-year follow-up demonstrating complete resolution of haemorrhage and retinal whitening. **E.** Fundus fluorescein angiography at 4-year follow-up showing absence of vascular leakage. **F.** Optical coherence tomography at 4-year follow-up demonstrating restoration of foveal contour without residual oedema.

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