Unilateral Idiopathic Retinal Venous Beading

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Dear Editor,

Retinal venous beading is a rare retinal vascular anomaly characterized by segmental dilatation of the retinal veins. It is typically associated with systemic or ocular disorders such as hypertension, diabetes or central retinal vein occlusion (CRVO). Idiopathic retinal venous beading is exceedingly uncommon, especially unilateral presentations, and may serve as a clinical indicator of systemic disorders, particularly renal or metabolic diseases such as Fabry and Alport syndromes.[1]

This report presents a rare case of unilateral idiopathic retinal venous beading, outlines its differential diagnosis, and discusses potential pathophysiological mechanisms.

A 45-year-old woman presented for a routine ophthalmological evaluation with no specific complaints. Her medical history was notable for a stable, non-progressive pituitary microadenoma that had been under regular surveillance for several years.

On examination, best-corrected visual acuities were 20/20 in both eyes. Anterior segment examination was unremarkable, and intraocular pressures were measured as 16 mmHg in the right eye and 18 mmHg in the left eye.

Macular optical coherence tomography (OCT), optic nerve head OCT, and Humphrey 30-2 visual field testing were all within normal limits. Fundoscopic examination of the right eye revealed marked venous beading involving the superior and inferior temporal veins, with no associated hemorrhages, exudates, or macular edema but with additional vascular sheathing of the nasal vessels (Figure 1A). Ghost vessels and shunt vessels were observed nasal to the optic disc in the right eye, indicating a previous nasal branch retinal vein occlusion (BRVO) (Figure 1B). Fundus fluorescein angiography of the right eye revealed no leakage from the optic disc or retinal vessels but confirmed venous beading and retinal shunt vessels located nasally (Figure 1C). The left eye was entirely normal (Figure 1D, E).

Systemic evaluation, including blood pressure monitoring, complete blood count, and metabolic panel, was unremarkable. Family history was notable for paternal hypertension and fibromuscular dysplasia (FMD), with death from stroke at age 65. The patient's mother and sibling had a history of varicose veins.

Rheumatology reported borderline antinuclear antibody positivity with anti-polymyositis/scleroderma antibody, which was deemed clinically insignificant. Neurological assessment, including brain magnetic resonance imaging and magnetic resonance venography, was normal. Carotid Doppler ultrasound revealed a minimal fibrofatty plaque without clinical significance.

Multidisciplinary evaluation (internal medicine, neurology, rheumatology) showed no significant abnormalities. A diagnosis of unilateral idiopathic retinal venous beading with nasal BRVO was established, and the patient was advised regular follow-up and education on warning symptoms.

Keywords: Retinal venous beading, fibromuscular dysplasia, nasal branch retinal vein occlusion

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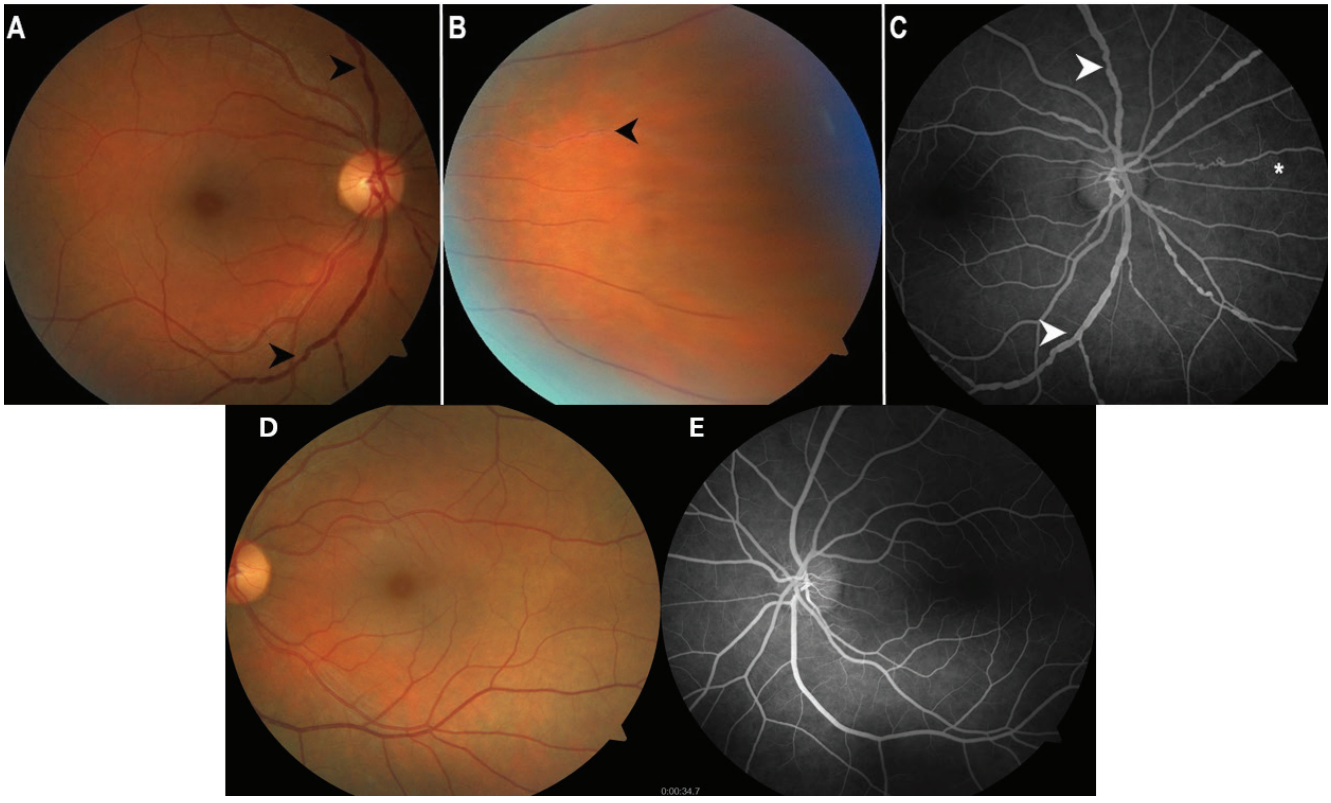

Figure 1. Fundus photographs of the right eye showed venous tortuosity and beading (A, black arrow head) and vascular sheathing in the nasal periphery (B, black arrow head). Fundus fluorescein angiography (C) demonstrated venous beading (white arrowheads) and retinal shunt vessels (white asterisk) indicating a previous venous occlusion. In the left eye, the color fundus photograph (D) and fluorescein angiography (E) appeared normal

Many causes of retinal venous beading have been described in the literature. As most cases are associated with systemic diseases, presentation is bilateral and characterized by flame-shaped retinal hemorrhages, retinal venous congestion and tortuosity, optic disc swelling, cotton wool spots, and macular edema.[1]

Previously, Meredith[1] reported inherited retinal venous beading. Prominent segmental beading of the retinal veins was observed in two male and three female family members across two affected generations. Additionally, various individuals exhibited focal retinal infarctions, surface retinal neovascularization, vitreous hemorrhages, microaneurysm formation, altered vascular permeability with lipid exudation, and localized edema. Renal disease was identified in two affected individuals during the fourth and fifth decades of life, respectively. Meredith[1] hypothesized that these retinal vascular abnormalities might be associated with Alport syndrome or Fabry disease.

Fonseca and Dantas[2] and Keyser and Ferguson[3] reported isolated instances of retinal venous tortuosity and beading in their patients, with no other affected family members. They referred to these cases as an idiopathic form of retinal venous tortuosity.

In a 24-year-old man with no known pathological history, Konaté and Mariko[4] described an isolated instance of bilateral retinal venous tortuosity of unknown origin and without visual sequelae that was detected incidentally during a routine fundus examination.

A woman with retinal venous beading and conjunctival vascular aneurysms but no systemic anomalies or family history was the subject of an isolated case reported by Ehongo and Rasquin.[5]

A similar beading morphology is also observed in FMD, an arterial occlusive disorder that may also contribute to retinal venous pathology. The characteristic "string of beads" angiography sign in FMD occurs due to the development of muscle fibers and connective tissue inside arterial vessel walls, usually in the medial layer.[6] It typically affects young individuals and has been reported in multiple siblings across several families.

FMD most commonly involves small- and medium-sized arteries, including the carotid, renal, and vertebral

arteries.[7] Astrike-Davis et al.[8] described a patient with CRVO associated with FMD and hypertension, suggesting a synergistic effect on retinal vasculature. The central retinal vein shares a common adventitial sheath with the central retinal artery. Therefore, arterial pathology such as FMD may compress the vein, leading to occlusion.[7] Additionally, FMD may induce turbulent flow, predisposing to thrombus formation and CRVO.

Other reports of familial cases of retinal venous beading across successive generations support a hereditary pattern in some patients.[9,10] However, unilateral sporadic cases have also been described. Abdel-Hay and Raman[11] reported a case of sporadic unilateral retinal venous beading in 2018. Similar to our case, no comparable clinical findings were identified among the patient's family members. In the absence of associated ocular or systemic vascular disease, the authors suggested that this condition represents a sporadic vascular defect. Our findings are consistent with this interpretation, further supporting the notion that unilateral retinal venous beading may occur as an isolated, non-familial entity.

The venous changes observed in the nasal quadrant of the right eye suggested a previous nasal BRVO. However, rather than attributing the generalized venous beading pattern to a prior BRVO, we believe that this occlusive event may represent a secondary consequence of the underlying venous beading observed in our case. Furthermore, a history of retinal venous occlusion in the absence of systemic hypertension or metabolic disease also suggests that idiopathic retinal venous beading could serve as an early warning sign.

This case emphasizes the importance of comprehensive diagnostic evaluation and long-term follow-up in patients with idiopathic retinal venous beading. Regular follow-up is essential both for early detection of complications and to contribute to the long-term understanding of this rare entity. Given the potential risk of future vascular occlusions in critical organs, cardiology and neurology consultations and appropriate antiplatelet or anticoagulant therapy may be warranted.

Ethics

Informed Consent: Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Declarations

Authorship Contributions

Surgical and Medical Practices: E.K., Concept: E.K., A.S.S., G.Y., Design: E.K., A.S.S., G.Y., Data Collection or Processing: E.K., Analysis or Interpretation: E.K., A.S.S., G.Y., Literature Search: E.K., A.S.S., Writing: E.K., A.S.S.

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