

Racemose hemangioma is an arteriovenous vascular malformation in which the retinal vessels appear abnormally dilated. The arteries are connected with the veins directly, without the interposition of a capillary network (Fig. 1A), forming a fragile vascular mass with turbulent flow that is predisposed to thrombosis phenomena. The fluorescein angiography (Fig. 1B) shows filling of the malformation without exudation. The optical coherence tomography and the optical coherence tomography-angiography en face (Figs. 1C and 1D) show the dilated vessels in the inner layers of the retina. A systemic study in these patients is essential because similar, potentially fatal malformations can be found elsewhere in the body.

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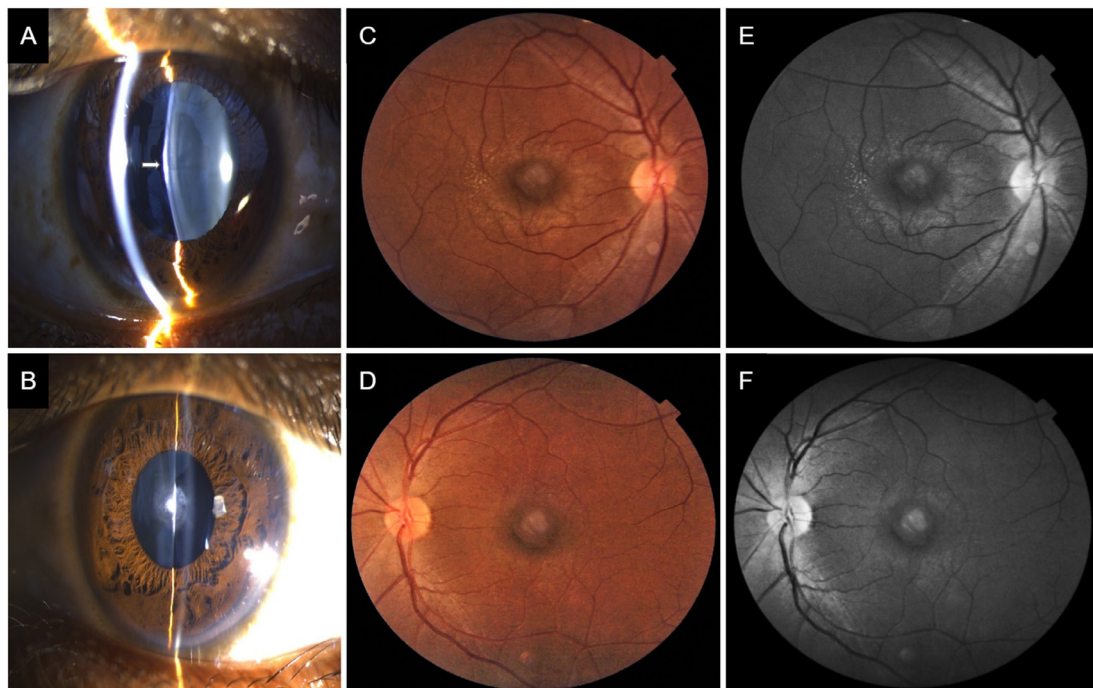
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Footnotes and Disclosure

The authors have no proprietary or commercial interest in any materials discussed in this article.

Role of multimodal ocular imaging in Alport syndrome



A 19-year-old man with Alport syndrome presented with diminution of vision in his left eye. Slit-lamp biomicroscopy showed an early anterior lenticonus with an anterior subcapsular cataract in his right eye (Fig. 1A) and a dense anterior polar and anterior subcapsular cataract in his left eye (Fig. 1B). Fundus photography revealed central perimacular dot-and-fleck retinopathy with a dull macular reflex or lozenge in both eyes (Fig. 1C, 1D). The flecks were better visualized on red-free imaging as a ring of fine white stippling around the macula (Fig. 1E, 1F). Bilateral temporal retinal thinning was present on optical coherence tomography (Fig. 2A, 2B) with distortion and temporal enlargement of the foveal avascular zone of the superficial retinal plexus on

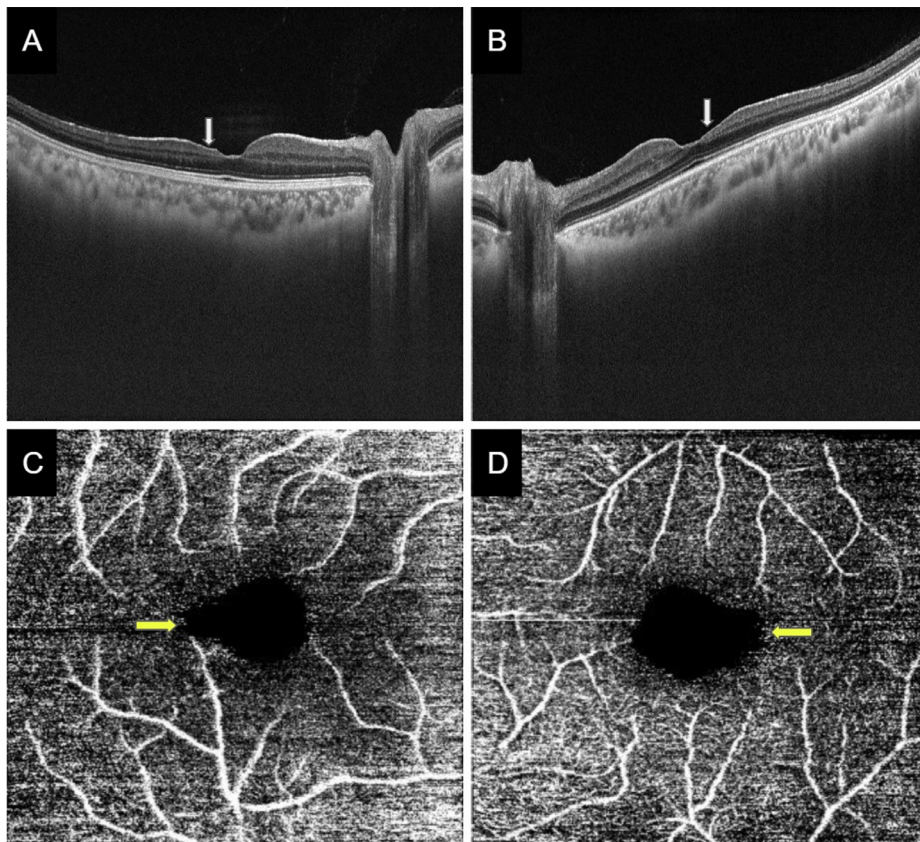
optical coherence tomography angiography (Fig. 2C, 2D). Ophthalmic imaging can provide a rapid, noninvasive, and relatively inexpensive tool for diagnosis of Alport syndrome and also can help predict progression of renal disease and mode of inheritance in certain situations.

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IgG₄-related orbital disease

