

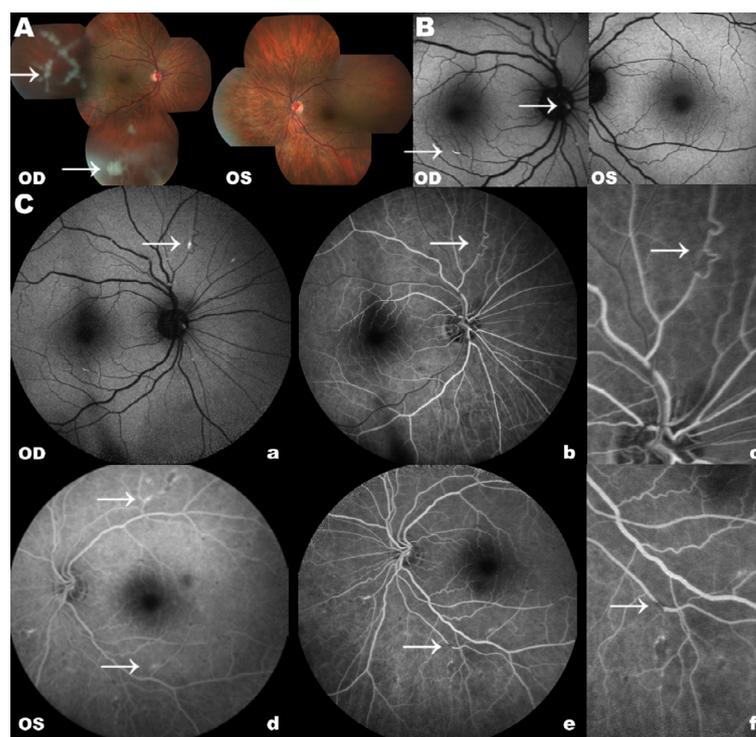
Retinal amyloid angiopathy

Angiopatia amiloide retiniana

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A 57-year-old male with previous myocardiopathy, polyneuropathy, bilateral cataract, and autonomic dysfunction had a family history of Familial Amyloid Polyneuropathy (FAP)^{1,2}. When he was 55 years old, he underwent a genetic testing, which detected a deleterious heterozygous mutation c.325G>A, Glu109Lys, on exon 3 of the Transthyretin gene, for the diagnosis

of FAP³. He was treated with Vyndaqel® (tafamidis meglumine) and amiodarone. He developed reduction of visual acuity in the right eye and floaters. Physical examination found that he had visual acuity of 20/40 (OD) and 20/25 (OS), with altered campimetry only in OD. The neuro-ophthalmological evaluation is provided in Figures 1-4. Laboratory testing excluded other



OD: right eye; OS: left eye; FAF: fundus autofluorescence; arrow: alterations.

Figure 1. A: Retinography showing vitreous opacity in temporal periphery in OD. It was normal in the OS. **B:** FAF images show hyper autofluorescence whitish deposit above the optic disk and in the superior and inferior vascular arcades of the OD. The FAF was normal in the OS. **C:** Fluorescein angiography: **a** – whitish deposit in the nasal superior retina; **b** – superior peripheral vascular tortuosity, collateral secondary to arterial occlusion; **c** – detail of figure **b**; **d** – whitish deposit in the superior and inferior (hyper autofluorescence) temporal vascular arcades; **e** – arterial retinal occlusion in the inferior periphery in the OS; and **f** – detail of figure **e**.

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hematological abnormalities. The final diagnosis was retinal amyloid angiopathy secondary to FAP. Ocular manifestations in FAP are rare; nevertheless, neurologists should investigate visual symptoms in patients with FAP. Retinal amyloid angiopathy is even rarer, but is a sight-threatening complication. Neurologists should assess the visual acuity, the intraocular pressure, vessel tortuosity, collaterals, or scalloped pupils,

promptly indicating an ophthalmological evaluation⁴. Recent progress in the neuro-ophthalmological evaluation indicated that retinal amyloid angiopathy is more frequent than previously reported⁴. Therefore, neurologists should be aware of this complication in patients with FAP, especially in those presenting vitreous amyloidosis or longer duration of the disease⁵.

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