

Idiopathic retinal vasculitis, aneurysms, and neuroretinitis (IRVAN) – Case Study

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Introduction

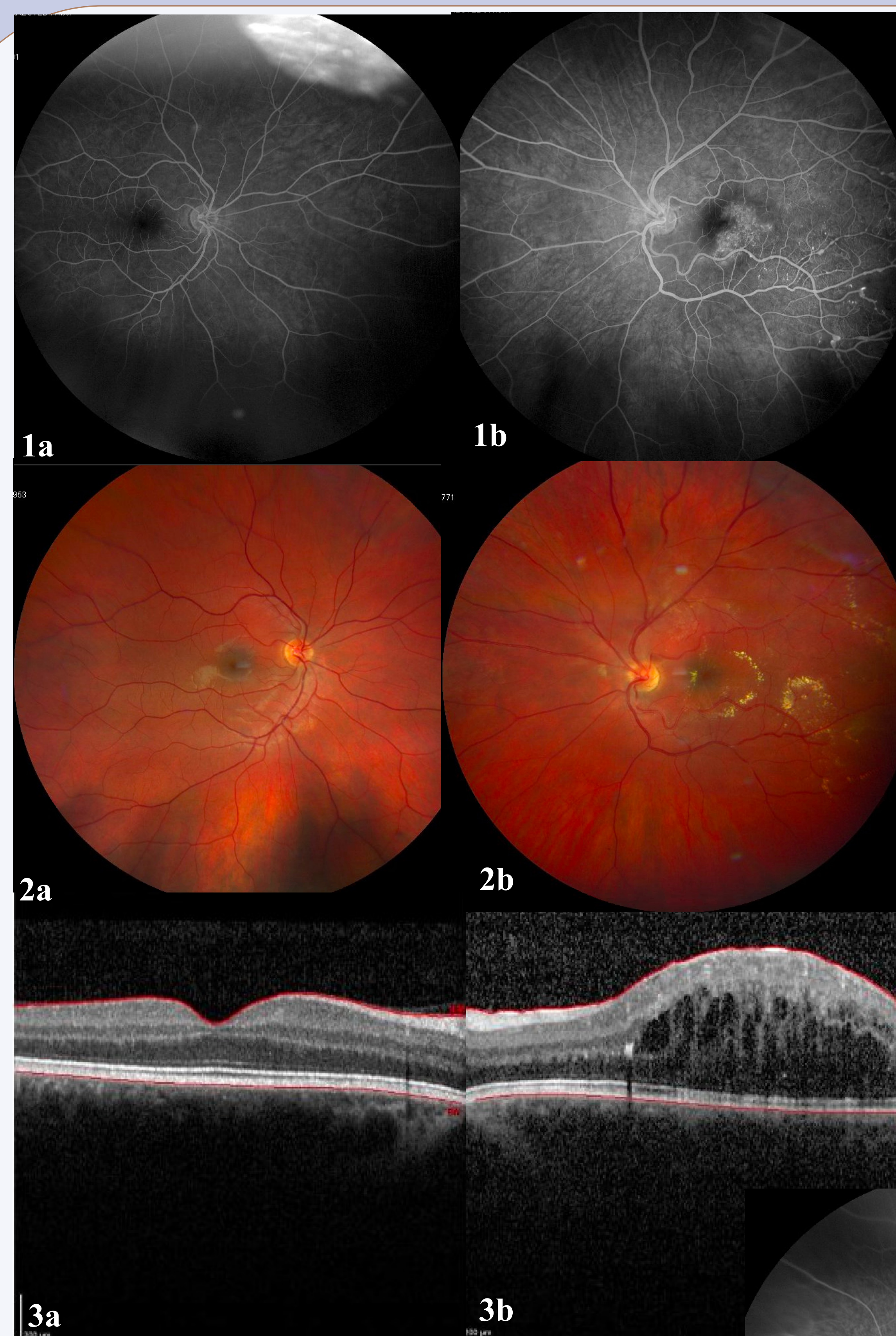
- ❖ Idiopathic retinal vasculitis, aneurysms, and neuroretinitis (IRVAN), while rare, most commonly presents in the third or fourth decade of life.¹
- ❖ Can be preceded by anterior uveitis and/or vitritis, and is associated with multiple leaking aneurysmal dilations, irregular venous dilation, and vascular sheathing.¹
- ❖ The three major criteria are retinal vasculitis, aneurysmal dilations at arterial bifurcations, and neuroretinitis, while minor criteria are peripheral capillary nonperfusion, retinal neovascularization, and macular exudation.²
- ❖ Clinical course varies with some presenting with self-limiting course to patients with severe cases progressing to vitreous hemorrhage and neovascular glaucoma.¹

Case Presentation

- ❖ 31yo M w/ blurry vision. Prior hx of head trauma, vitamin D deficiency, and melanoma
- ❖ Fluorescein angiogram was obtained as showed in the next section. OCT macula was obtained
- ❖ Labs within normal limits
 - ❖ Lysozyme, PR3-ANCA, MPO, HLA- B27, B51, A29, ESR, CRP, C3, Toxoplasma, Bartonella, Treponema pallidum, QuantiFERON-TB Gold, Toxocara, ACE, Lyme titers

Initial Presentation 1/2020

- ❖ 1.25 mg/0.05cc intravitreal Avastin injection
- ❖ Prednisone 60mg PO



Initial Presentation 1/2020

Fluorescein angiography Fig. 1a OD, Fig. 1b OS from initial presentation. IVFA OS shows extensive hyperfluorescence of the optic disc, macula, and aneurysmal bulbs. There is also peripheral nonperfusion.

Color Fundus Photography Fig. 1a OD, Fig. 1b OS from initial presentation. OS shows nasal optic disc edema, exudates, an ERM in the macula, and extensive perivascular sheathing of the arterioles and veins.

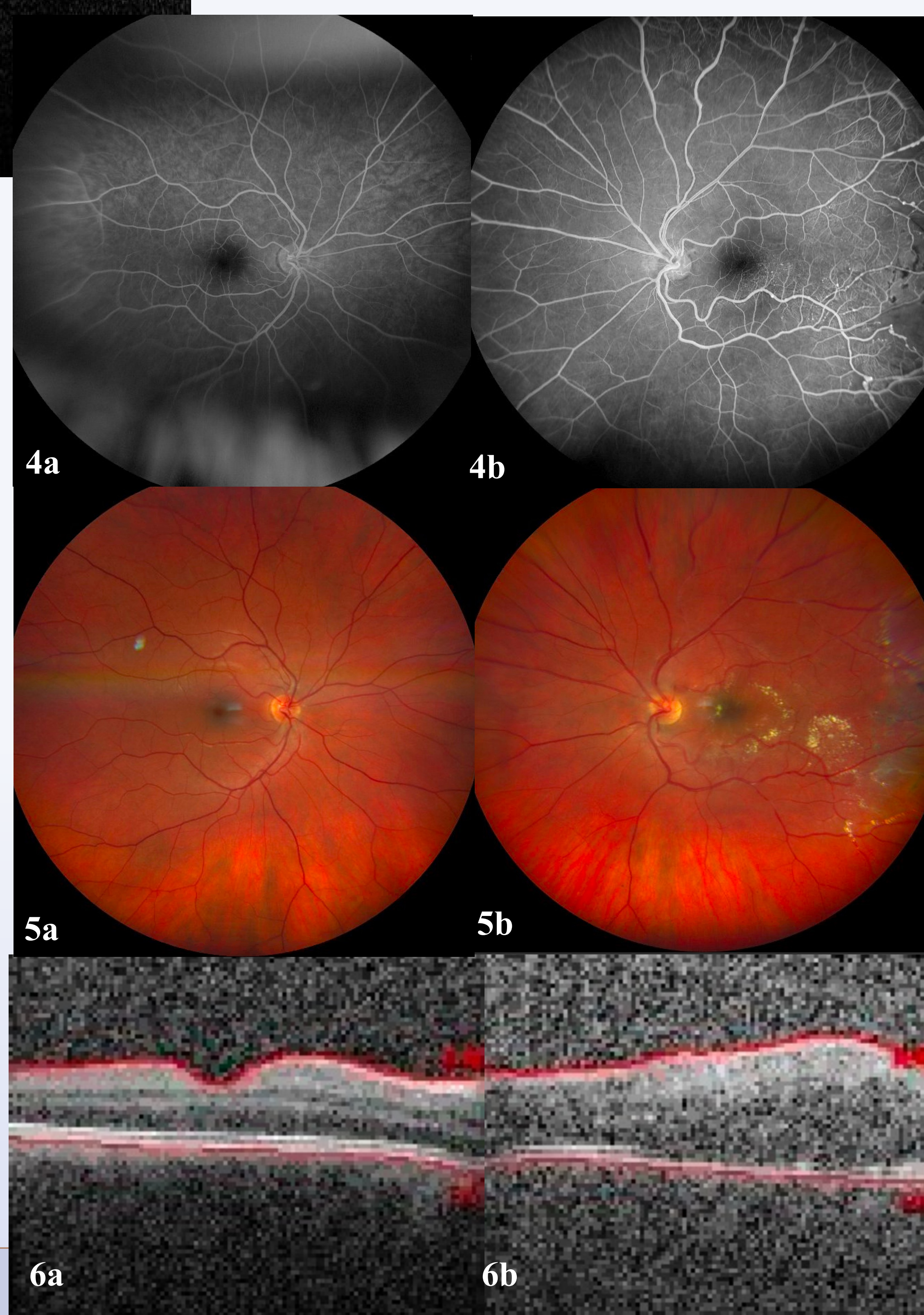
OCT Retina Fig. 3a OD, Fig. 3b OS from initial presentation; Normal OD. OS shows loss of foveal contour with significant intraretinal edema.

Follow Up Presentation 2/2021

Fluorescein angiography Fig. 4a OD, Fig. 4b OS; Normal perfusion, OD Improved compared to prior, OS after Avastin and oral prednisone treatment.

Color Fundus Photography Fig. 5a OD, Fig. 5b OS; stable from prior presentation

OCT Retina Fig. 6a OD, Fig. 6b OS; Interval improved intraretinal fluid



Case (cont.)

Two week follow up

- ❖ Sectoral panretinal laser photocoagulation (PRP)

Follow up 2/2021

- ❖ Vision gradually improving
- ❖ Exam stable with retinal thickening partially secondary to ERM and exudates in macula OCT with interval decreased intraretinal fluid
- ❖ IVFA with decreased leakage in the macula and with decreased leakage peripherally

Discussion

- ❖ IRVAN is a diagnosis of exclusion. An extensive evaluation to rule out other etiologies was performed.⁴
- ❖ Early panretinal laser photocoagulation should be considered shortly after (or before) development of neovascularization and when angiographic evidence of widespread retinal nonperfusion is present.³
- ❖ Patient was treated with Avastin, oral prednisone, PRP, and showed visual improvement.
- ❖ This patient presents with a rare retinal vascular entity. If IRVAN is left untreated, it may lead to severe bilateral visual loss.

References

1. Pichi, Francesco, Ciardella, Antonio. Imaging in the Diagnosis and Management of Idiopathic Retinal Vasculitis, Aneurysms, and Neuroretinitis (IRVAN). *Int Ophthalmol Clin.* 2012;52(4):275-282. doi:10.1097/IIO.0b013e318265d418.
2. Bajgai P, Katoch D, Dogra MR, Singh R. Idiopathic retinal vasculitis, aneurysms, and neuroretinitis (IRVAN) syndrome: clinical perspectives. *Clin Ophthalmol.* 2017;11:1805-1817. Published 2017 Oct 6. doi:10.2147/OPTH.S128506
3. Samuel M.A., Equi R.A., Chang T.S. vol. 114. 2007. pp. 1526–1529. (Idiopathic Retinitis, Vasculitis, Aneurysms, and Neuroretinitis (IRVAN): New Observations and a Proposed Staging System).
4. Moosavi M, Hosseini SM, Shoeibi N, Ansari-Astaneh MR. Unilateral idiopathic retinal vasculitis, aneurysms, and neuroretinitis syndrome (IRVAN) in a young female. *J Curr Ophthalmol.* 2015;27(1-2):63-66. Published 2015 Nov 17. doi:10.1016/j.joco.2015.10.006



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