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# *Diagnostic and Therapeutic Challenges*

Edited by H. Richard McDonald

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This case is submitted by Drs. Yi Xuan, Wei Liu, Gezhi Xu, and Yongjin Zhang, Department of Ophthalmology, Eye and ENT Hospital of Fudan University, Shanghai Key Laboratory of Visual Impairment and Restoration, Shanghai, China; commented by Dr. Arun D. Singh (Cleveland, Ohio).

## Case Report

A 42-year-old man presented with a 1-month history of decreased vision in the left eye in March 2016. His best-corrected visual acuity (BCVA) was 20/20 in the right eye and 20/400 in the left eye. He denied any history of hypertension, diabetes, hyperlipidemia, or other systemic disease. The anterior segment and intraocular pressures were normal. Dilated fundus examination of the left eye revealed the presence of an orange-red tumor superonasal to the fovea in the macular region surrounded by massive subretinal and intraretinal hemorrhages and retinal exudation. This tumor was fed by a pair of dilated retinal vessels, with coexisting anteriovenous nicking (Figure 1A). No other lesions were noted in the peripheral fundus.

Fundus fluorescein and indocyanine green angiography showed the presence of hyperfluorescent dilated feeder arterioles and fine capillary filling of the tumor in the arterial phase. The engorged draining vein becomes prominent in the venous phase. The hyperfluorescent tumor was surrounded by hypofluorescence corresponding to hemorrhage (Figure 1B). The tumor demonstrated progressive hyperfluorescence with late leakage of dye into the surrounding structures. Scattered microaneurysms were also found around the tumor (Figure 1C). Optical coherence tomography (Spectralis HRA-OCT; Heidelberg

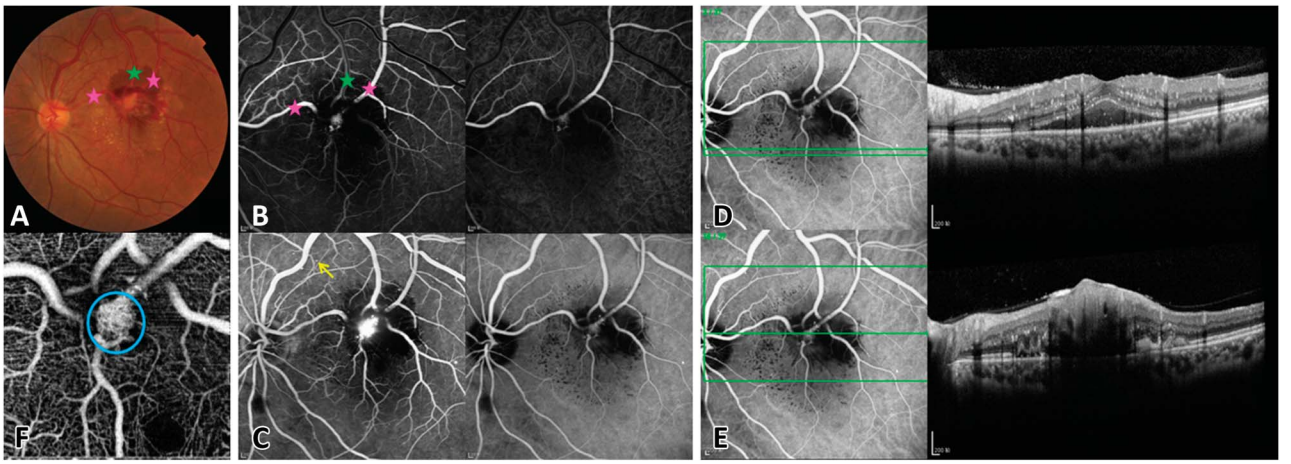


**Fig. 1.** Fundus presentation of the patient at baseline. **A.** Fundus photograph shows an RCH superonasal to the fovea in the macular region, supplied by dilated, tortuous feeder arteriole (pink asterisk) and draining vein (green asterisk). **B.** Fundus fluorescein and indocyanine green angiography depicts the feeder arteriole (pink asterisk) and draining venule (green asterisk), supplying the bright hyperfluorescent lesion at the early stage. **C.** Fundus fluorescein and indocyanine green angiography shows progressive hyperfluorescence of the tumor with late leakage of dye, surrounded by scattered hyperfluorescent microaneurysms (yellow arrow). **D.** Optical coherence tomography shows the presence of intraretinal and subretinal fluid at the macula. **E.** Optical coherence tomography through the tumor scan shows hyperreflective lesion within the inner retinal layer. **F.** Optical coherence tomography angiography revealed that the tumor was composed of capillary network at the superficial slab (blue circle).

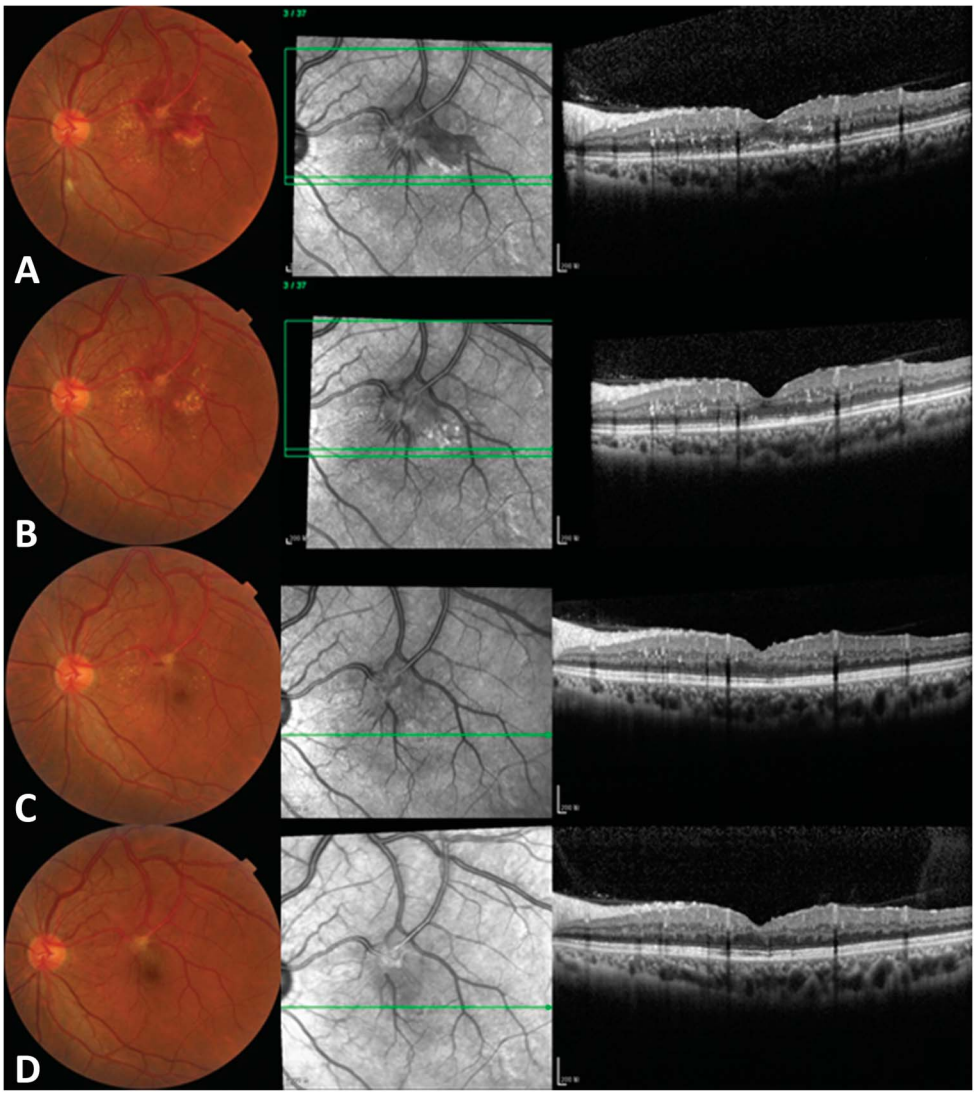


**Fig. 2.** Fundus changes during follow-up period. **A.** At 2 weeks after first injection, fundus photograph showed that the hemorrhage was largely absorbed and the tumor was decreased in size. Optical coherence tomography revealed that the subretinal fluid was remarkably resolved. **B.** At 1 month after first injection, fundus photograph showed the complete absorption of hemorrhage. The optical coherence tomography displayed that the subretinal fluid was further resolved. **C.** At 1 month after third injection, fundus photograph demonstrated that the tumor was further decreased in size and changed to pale white in color. Retinal exudation was remarkably regressed. The optical coherence tomography exhibited that the subretinal fluid was completely resolved. **D.** At 1-year follow-up, no retinal exudation and edema were found.

Engineering, Heidelberg, Germany) demonstrated the subretinal and intraretinal fluid accumulation in the macula (Figure 1D) and a hyperreflective lesion within the inner retinal layer, with intraretinal fluid through the tumor scan (Figure 1E). Optical coherence tomography angiography (Topcon Corporation, Japan) showed that the tumor was composed of a capillary network (Figure 1F) at the level of the superficial slab. The fundus of the right eye was normal.

Systemic examination including magnetic resonance angiography of the brain and ultrasonography of the abdomen ruled out the existence of any abnormalities. The patient underwent an injection of intravitreal ranibizumab. Two weeks later, BCVA improved to 20/66 with the hemorrhage largely absorbed. The intraretinal and subretinal fluid had significantly resolved, and the tumor was remarkably decreased in size (Figure 2A). One month after the injection, BCVA stabilized at 20/66, although retinal edema remained (Figure 2B). He then received a second dosage of ranibizumab and his BCVA increased to 20/50, one month later. The size of the tumor progressively decreased. Fundus fluorescein and indocyanine green angiography demonstrated tumor shrinkage with narrowing of vessels, remaining mild leakage from the tumor; microaneurysms were still scattered around the tumor (Figure 3, A and B). One month after the third injection of ranibizumab, his BCVA improved to 20/33 and the retinal edema had completely resolved in the macula. The color of the tumor had changed from red to pale white (Figure 2C). The optical coherence tomography angiography revealed that the abnormal capillary network had collapsed (Figure 3C). This result remained stable throughout 1 year of follow-up (Figure 2D).

We believe this lesion to represent a retinal capillary hemangioma (RCH) and present this case for discussion of diagnosis and treatment.

## Dr. Arun D. Singh (Cleveland, Ohio):

Retinal capillary hemangioma is a benign angiomatous hamartoma of the retina. It may occur sporadically or as a manifestation of von Hippel–Lindau disease.[1–3] Sporadic RCH tends to occur later in life as compared with RCH of the von Hippel–Lindau type.[1] Sporadic RCH typically manifest as a solitary, unilateral tumor located in the temporal periphery, whereas those associated with von Hippel–Lindau disease are multifocal, bilateral, or juxtapapillary in location.[1] The visual loss is generally caused by

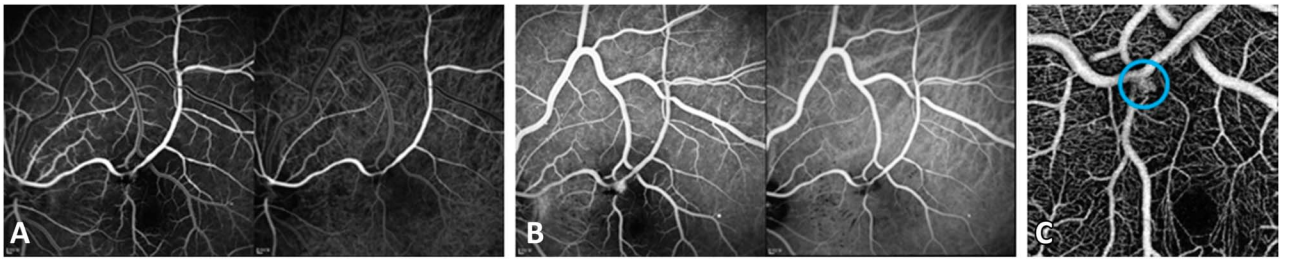


**Fig. 3.** Fundus fluorescein and indocyanine green angiography at one month after second injection shows tumor shrinkage with narrowing of vessels (**A**), remaining mild leakage of the tumor at the late stage (**B**); and microaneurysms still scattered around the tumor. The abnormal capillary network revealed on optical coherence tomography angiography at the superficial slab becomes collapsed (**C**) (blue circle).

exudation from the retinal tumor. The intraretinal and subretinal fluid may progress to exudative retinal detachment and glial proliferation that can lead to traction retinal detachment.[1,4]

The diagnosis is based on clinical findings of young age at presentation, extramacular location, and presence of prominent feeder vessels, which may not be evident with small RCH. Retinal hemorrhage is only exceptionally observed in RCH (less than 3%) and almost always absent in smaller lesion (less than 1 mm).[5]

None of the features outlined above were noted in the lesion reported herein. More importantly, careful review of the fundus photograph, fluorescein angiogram, and optical coherence tomography angiogram reveals that the lesion is centered on the retinal arteriole. Hence, the findings reported herein support a diagnosis of a retinal macroaneurysm rather than RCH.[6] The clinical course of resolution, attributed to intravitreal ranibizumab, also supports the diagnosis of macroaneurysm, which are known to respond to the anti–vascular endothelial growth factor,[7] unlike RCH, which do not[8,9]

**Editor's Note:**

Drs. Xuan, Liu, Xu, and Zhang have presented a man with decreased vision associated with a vascular tumor. Dr. Arun Singh discusses this case for us.

Dr. Singh takes issue with the diagnosis of RCH. He reviews RCH, a benign angiomatosis hamartoma of the retina, and discusses the differences between sporadically occurring RCH and those occurring as part of the VHL syndrome. Specifically, sporadic RCH are usually unilateral, solitary tumors in the temporal periphery, and those that occur as part of von Hippel–Lindau disease are usually multifocal, bilateral, or juxtapapillary. Vision is lost because of exudation, glial proliferation, or traction. Hemorrhage is rarely seen in smaller lesions. Dr. Singh does not feel that this lesion represents an RCH, but rather a retinal arteriolar macroaneurysm. He states that the age of presentation, location, and presence of hemorrhage are not typical of an RCH. Moreover, the lesion is centered on a retinal arteriole, and the clinical course of resolution after anti-VEGF injection is more consistent with this lesion being a retinal aneurysm. He ends by declaring that macroaneurysms respond to anti–vascular endothelial growth factor, RCHs do not.

We thank Drs. Xuan, Liu, Xu, and Zhang for their case, and Dr. Arun Singh for his analysis.

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