

# Recurrent retinal and choroidal ischemia in a case of ocular ischemic syndrome

Koushik Tripathy<sup>1</sup> and Shahana Mazumdar

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**Abstract:** A 62-year-old male patient presented with fluctuating vision in both eyes. On fundus fluorescein angiogram, there was an area of choroidal and retinal ischemia in the left eye. The carotid evaluation revealed complete stenosis of the right internal carotid artery and 90% stenosis of the left internal carotid artery for which he underwent left modified endarterectomy. The vision and angiographic features improved after the procedure. Eleven months after the procedure, he again presented with choroidal ischemia due to recurrent left internal carotid artery blockage which improved gradually after carotid stenting.

**Keywords:** atherosclerosis, choroidal filling, giant cell arteritis, short posterior ciliary artery, triangle of Amalric

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## Introduction

Choroidal ischemia is an uncommon finding which may be caused by systemic diseases and/or local vascular disorders.<sup>1</sup> Common systemic disorders include giant cell arteritis (GCA), malignant hypertension, pregnancy-induced hypertension, renal failure, collagen vascular diseases, and hematological diseases including disseminated intravascular coagulation and thrombotic thrombocytopenic purpura. Local vascular diseases causing choroidal ischemia include carotid artery disease, trauma, and laser.<sup>1</sup> In this case report, we describe a patient with recurrent choroidal and retinal ischemia who required carotid endarterectomy and later carotid stenting. A written consent to publish the medical data and images was obtained from the patient.

## Case description

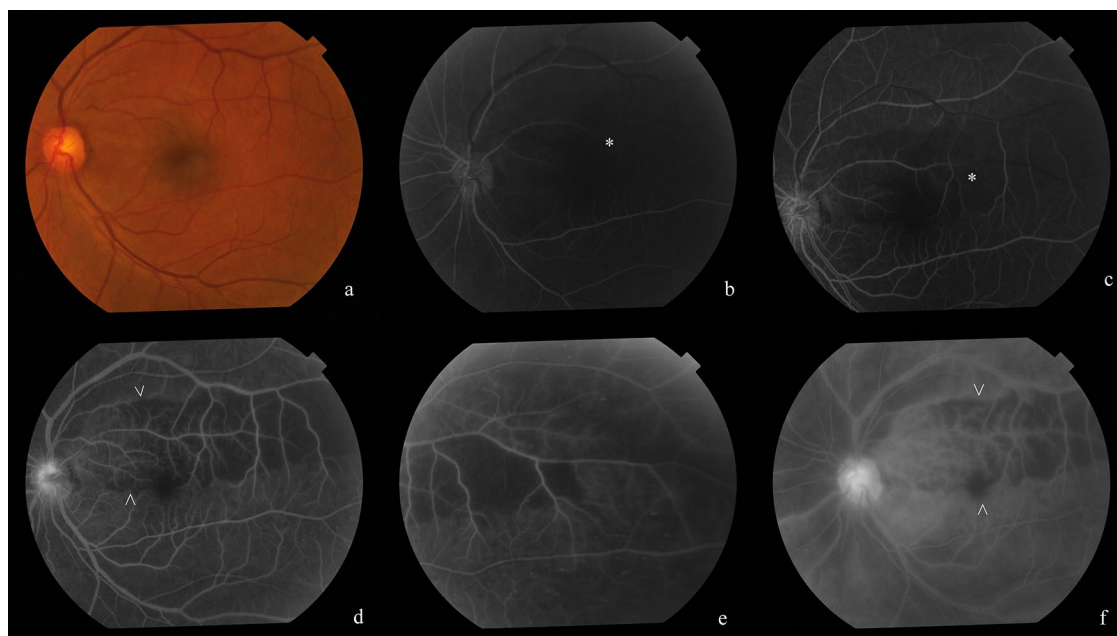
A 62-year-old apparently healthy male presented on 17th April 2015 with fluctuating vision in both eyes for 1 year. He had no history of diabetes, hypertension, coronary artery disease, hyperlipidemia, or smoking. At presentation, the best-corrected visual acuity (BCVA) was 6/6p, N6 in the right (RE) and 6/18, N18 in the left eye (LE). The intraocular pressure (IOP) was 12 mm Hg in both eyes. Each eye had a clear lens and no anterior chamber cells. There was no new vessel in the

iris (NVI) or in the angle of the anterior chamber (NVA). The vitreous was clear bilaterally. The fundus showed dilated veins in both eyes (Figure 1(a) and 2(a)). On fundus fluorescein angiogram (FFA), there was delayed arm to retina time. There was delayed filling of the superotemporal artery with visible leading edge in the left eye (Figure 1(b), \*). The filling of the superotemporal vein was also delayed in the LE (Figure 1(b)). A horizontal band of hypofluorescence was noted at the level of the choroid in the LE (Figure 1(c), (d), (e), (f)). Staining of the optic nerve head was more prominent in the LE (Figure 1(e) and (f)). Peripheral capillary nonperfusion (CNP) and microaneurysms were seen in both eyes (Figure 1(d)). Paravascular staining was noted in the LE in late phases of FFA (Figure 1(d) and (f)). The RE showed perifoveal capillary changes with mild late leak (Figure 2(b) and (c)). Optical coherence tomography (OCT) macula of RE was unremarkable (Figure 2(d)). There was some inner retinal thinning in the superior macula of the LE (Figure 2(e)). Hemogram, erythrocyte sedimentation rate (ESR), and C-reactive protein (CRP) were within normal limits. Carotid studies revealed complete stenosis of the right internal carotid artery and 90% stenosis of the left internal carotid artery. The right carotid was considered inoperable and had extensive anastomoses.

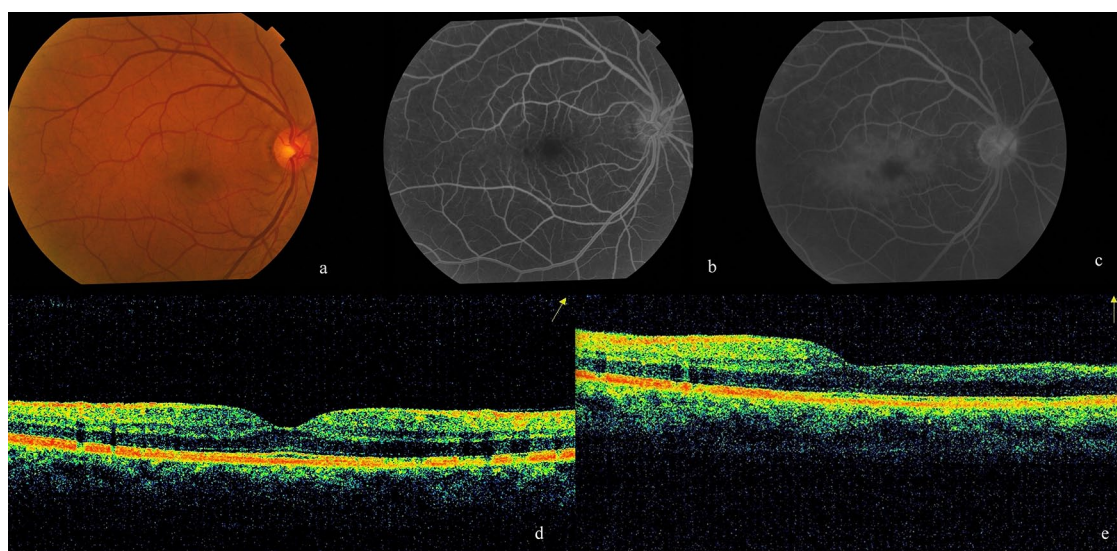
Correspondence to:  
**Koushik Tripathy**  
Department of Vitreoretina  
and Uvea, ICARE Eye  
Hospital & Postgraduate  
Institute, E3A, Sector-26,  
Noida, Uttar Pradesh,  
India - 201301.  
[koushiktripathy@gmail.com](mailto:koushiktripathy@gmail.com)

**Koushik Tripathy**  
Department of Vitreoretina  
and Uvea, ICARE Eye  
Hospital & Postgraduate  
Institute, Noida, India.

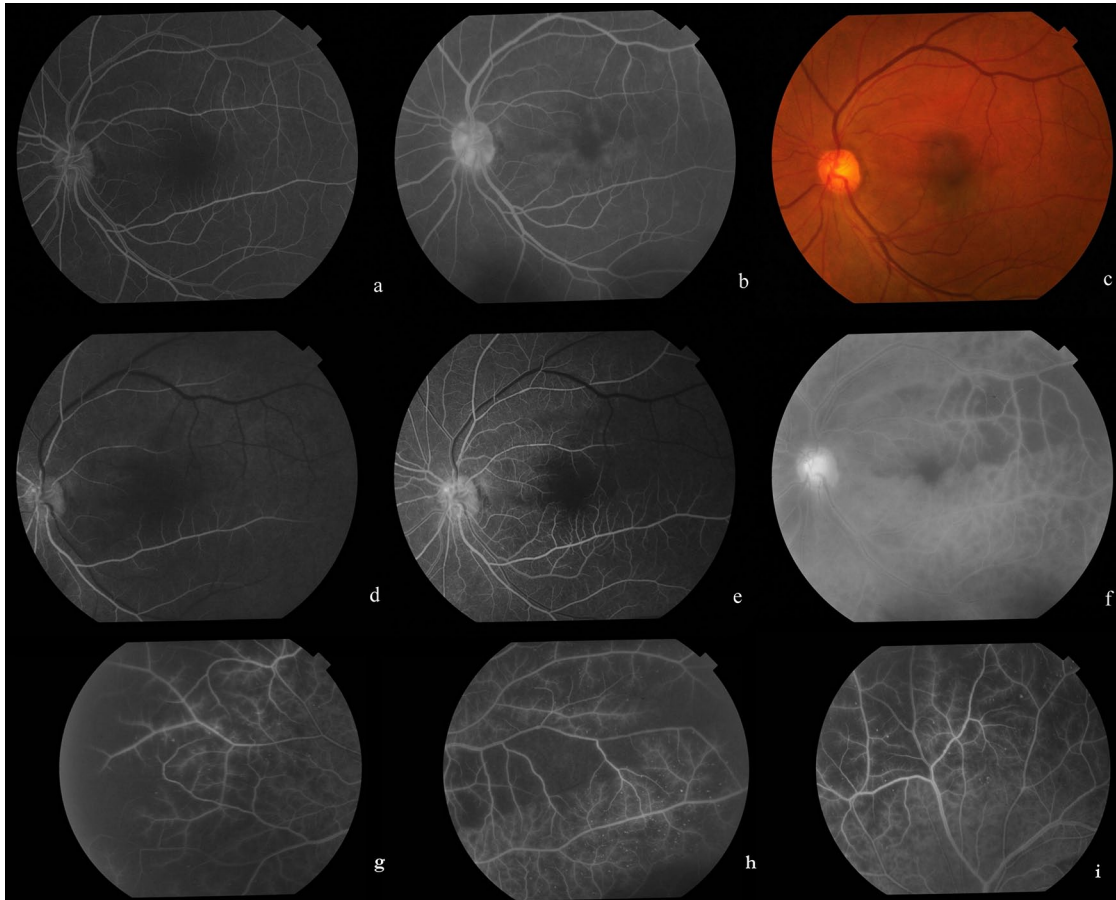
**Shahana Mazumdar**  
Department of Vitreoretina  
and Uvea, ICARE Eye  
Hospital & Postgraduate  
Institute, Noida, India



**Figure 1.** (a) The fundus picture of the left eye at the presentation, showed venous dilation, (b) The early arteriovenous (AV) phase of fundus fluorescein angiogram (FFA) image showed a slowly progressing front edge of the dye (\*) in the superotemporal retinal artery (suspected cilioretinal artery) and delayed filling of the superotemporal retinal vein. The perfusion of whole choroid seems delayed and compromised, (c) Late AV phase, showed the area of choroidal ischemia, and sluggish flow in the superotemporal retinal artery (\*), (d, e) Mid phase showed disk leak and the horizontal area of choroidal ischemia (in between arrowheads ^), (f) Late phase showed obvious disk leak and paravascular staining.



**Figure 2.** (a) The right fundus at presentation, (b) Early phase of FFA revealed irregular caliber of the retinal arterioles and microvascular abnormality around the fovea and temporal microaneurysms, (c) The late FFA image of the right eye showed mild perifoveal capillary leak, (d) The optical coherence tomography (OCT) scan of the right fovea showed normal foveal contour and absence of intraretinal/subretinal fluid, (e) The vertical optical OCT scan (going from inferior to superior) of the left fovea shows mild inner retinal thinning of the superior macula.  
FFA: fundus fluorescein angiogram.



**Figure 3.** (a) Four and half months after carotid endarterectomy, the choroidal and retinal ischemia has markedly improved, but there is still some paravasular capillary leakage (b) in the late phase and disk leak, (c) Eleven month after the procedure, left eye showed neovascularization of the disk. The arterial phase (d) showed impaired choroidal perfusion and arterial dye front in the superotemporal retinal artery. The arteriovenous phase (e) shows sluggish arterial circulation, and evident recurrent choroidal ischemia. Late phase (f) shows severe paravasular and capillary leak and disk leak. Peripheral retinal capillary nonperfusion areas were noted (g, h, i).

He underwent left modified endarterectomy on 27<sup>th</sup> April 2015.

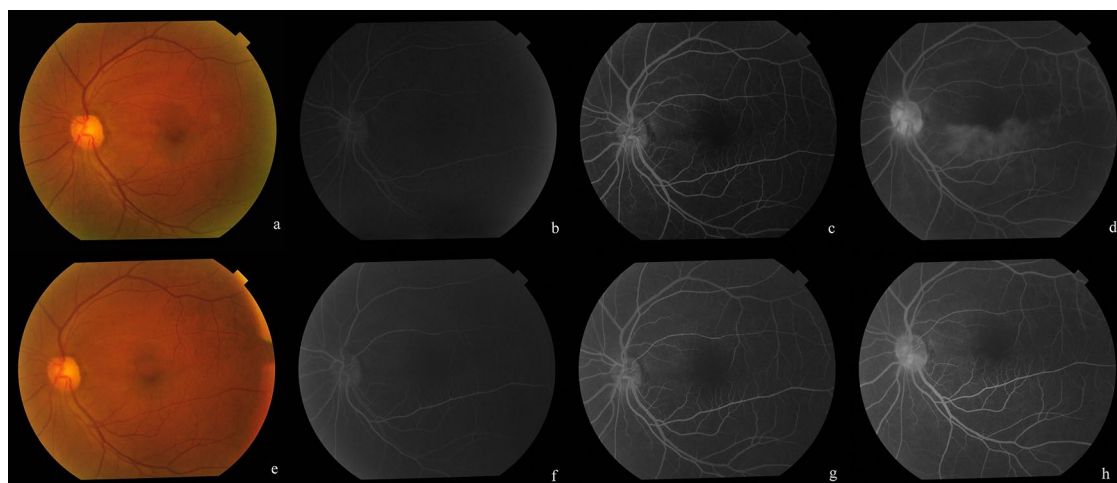
Four and half months after the procedure (on 17<sup>th</sup> September 2015), BCVA was 6/6p, N6 in RE and 6/9, N6 in LE. IOP was 10 mmHg in both eyes. The FFA showed significantly improved perfusion, decreased choroidal ischemia, and disk staining (Figure 3(a) and (b)) in the left eye.

Eleven months after the procedure (on 25<sup>th</sup> March 2016), the vision of the LE dropped again. This time, the BCVA was 6/6, N6 in RE and 6/12P, N18 in LE. IOP was 9 mmHg in both eyes. There were fine new vessels at the disk (NVD) superiorly (Figure 3(c)). FFA showed leak from the NVD

and severe recurrence of the choroidal and retinal ischemia (Figure 3(d)–(f)) in the LE. The OCT showed thinning of the superior macula in LE as was noted before. Peripheral retinal new vessels were not seen. Laser of peripheral capillary non-perfusion areas (Figure 3(g)–(i)) was done in LE.

He underwent computed tomography (CT) angiography of neck vessels which showed recurrent left internal carotid blockage. He underwent carotid stenting on 28<sup>th</sup> March 2016 (11 months after the initial surgery).

On 16<sup>th</sup> June 2016 (2 and ½ months after the carotid stenting), the BCVA was 6/6, N6 in RE and 6/9, N10 in the left eye. IOP was 10 mmHg



**Figure 4.** (a) Fundus photo of the left eye two and a half month after carotid stenting, (b, c, d) Corresponding FFA showed improved retinal and choroidal circulation, (e) Fundus photo after 2 and ½ years of carotid stenting of the left side, (f, g, h) FFA shows subtle residual choroidal ischemia and mild disk staining in the late phase (h).

FFA: fundus fluorescein angiogram.

in either eye. However, the FFA features showed some residual choroidal ischemia and leak in the inferior macula (Figure 4(a)–(d)).

On the recent most visit (21<sup>st</sup> September 2018, 2 and ½ year after the second surgery/carotid stenting), the BCVA was 6/6 (RE) and 6/9p (LE), and there was mild progression of cataract in the left eye. The IOP was 11 mmHg (RE) and 13 mmHg (LE). There was no NVI or NVA in either eye. On FFA, the choroidal ischemia improved much with some remaining late disk hyperfluorescence (Figure 4(e)–(h)).

### Discussion

Our patient had bilateral ocular ischemic syndrome (OIS),<sup>2</sup> due to complete occlusion of the right internal carotid artery and 90% stenosis of left internal carotid artery. The features of OIS in this patient include venous dilation, irregular retinal arterial caliber, peripheral capillary nonperfusion, mid-peripheral microaneurysm, late optic nerve hyperfluorescence in both eyes, perifoveal capillary changes and leakage in the right eye, and marked paravascular staining in the left eye. The entire choroid had delayed and markedly poor filling with non-filling of the short posterior ciliary artery (SPCA) in the superior macular region. Another peculiar finding in the case is the presence of simultaneous recurrent sluggish circulation in the superotemporal retinal artery, which

appeared to arise from the trunk of the central retinal artery at the center of the optic nerve. Also, the superior macular inner retina was thin compared to the inferior macula suggesting a sequel of inner retinal ischemia or branch retinal arterial occlusion. However, the superotemporal retinal artery may have been a cilioretinal artery that arises from the choroidal circulation (personal communication with Dr. Sohan Singh Hayreh) which can explain simultaneous focal retinal and choroidal ischemia.<sup>3</sup> Also, in branch retinal arterial occlusion subsequent vascular refilling after relieving carotid stenosis is unlikely (personal communication with Dr. Mahesh P Shanmugam).

Choroidal ischemia in a triangular pattern as in Amalric triangle may appear similar to our case, but the apex of this triangle is usually toward the fovea and involve mid-peripheral retina<sup>4,5</sup> contrary to our case which had choroidal ischemia in the superior part of the macula which extended a little beyond the posterior pole.

Another explanation of the area of choroidal ischemia could be the area of watershed zone (WSZ), which is the border of supplied areas of 2 end arteries.<sup>3</sup> If the perfusion pressure of one or more end arteries is reduced, the peripheral margin of the territory of that artery/ies (at WSZ) is most prone to ischemia/vascular compromise. When there are 2 posterior ciliary arteries (PCA, medial and lateral), the WSZ is usually vertically

oriented and may pass through the optic disk, or nasal or temporal to the optic disk.<sup>3</sup> When 3 or 4 PCAs are present, the WSZs usually radiate from the disk and usually horizontal WSZ pass through the fovea.<sup>3</sup> Also, most of the WSZs of short posterior ciliary arteries (SPCAs) involve the fovea and almost radiate from the fovea, making it most vulnerable to ischemia.<sup>3</sup> However, the area of choroidal ischemia in our case was superior to the fovea and bisected the fovea which points more toward the vascular compromise of an SPCA supplying that area.

Because of generalized poor filling of the choroid, no watershed zones were seen (personal communication with Dr. Sohan Singh Hayreh). Low IOP in our case was related to OIS. In OIS, the choroidal vascular bed is affected more than retinal vasculature as choroidal circulation has no autoregulation<sup>1</sup> compared to retinal circulation.

Normal ESR, CRP, and absent suggestive history/clinical features excluded giant cell arteritis in our patient. At presentation, the optic disk of the LE had mild edema which was evident on FFA as optic disk staining. This is caused by ischemia of the optic nerve head which is supplied by the posterior ciliary circulation.<sup>6</sup> However, LE later developed neovascularization of the optic disk (Figure 3(c)) which accentuated the disk hyperfluorescence (Figure 3(e) and (f)), but reduced considerably after scatter photocoagulation of the peripheral capillary nonperfusion areas and carotid stenting (Figure 4). The color doppler imaging of the orbital circulation<sup>7</sup> was not performed, which is a limitation of this case report.

A combination of retinal arterial occlusion with choroidal ischemia has been reported in internal carotid artery stenosis,<sup>4</sup> polyarteritis nodosa,<sup>8</sup> and Wegener's granulomatosis.<sup>9</sup> Recurrent retinal and choroidal ischemia may also be noted in sickle cell disease.<sup>10</sup>

Carotid endarterectomy has been noted to improve or stabilize visual acuity in cases with chronic OIS due to internal carotid artery stenosis.<sup>11</sup> Also, there are multiple reports showing improvement or prevention of progression of ocular ischemia after carotid endarterectomy or carotid artery stenting.<sup>12</sup>

In conclusion, we report an interesting case of ocular ischemia which initially improved and later

worsened after carotid endarterectomy, and improved again after carotid artery stenting.

#### Authors' note

Part of the manuscript was presented in a presentation at Noida Ophthalmological Society meeting, 2018, and Retnet India meet 2019.

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#### Conflict of interest statement

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#### ORCID iD

Koushik Tripathy  <https://orcid.org/0000-0002-9756-128X>

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