Fluorescein Angiographic Features of Three Cases of Frosted Branch Angiitis in Egypt

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Abstract: We examined three previously healthy young patients who reported to our hospital (Research Institute of Ophthalmology, Egypt) with unilateral/bilateral acute visual loss associated with diffuse retinal peri-phlebitis in the form of thick, inflammatory infiltrates surrounding all of the retinal veins creating the appearance of frosted tree branches. Occlusive vasculitis occurred in two out of three patients. The clinical appearance of these patients matched that of a previously described condition known as Frosted Branch Angiitis. In this report, we describe the clinical course, as well as fluorescein angiographic features of these three cases.
[Sherif Karawya, and Mahmoud Leila**. Fluorescein Angiographic Features of Three Cases of Frosted Branch Angiitis in Egypt.** *Life Sci J* 2013; 10(4): 2086-2091]. (ISSN: 1097-8135). http://www.lifesciencesite.com. 277

Key words: Frosted Branch Angiitis, retinal vasculitis.

1- Introduction

Acute bilateral retinal vasculitis was first described in a 6-year-old boy by Ito *et al.* in 1976. The development of peri-vascular sheathing extending towards the periphery resembled frosted branches of a tree, hence the term 'Frosted Branch Angiitis'[1].

To date, only 57 cases were reported. It affects young, otherwise healthy individuals. Most patients (75%) have bilateral disease. Most common symptoms are visual loss, floaters or photopsia. Typically, the fundus shows florid, translucent retinal peri-vascular sheathing in association with variable uveitis and retinal edema. The condition is considered a 'primary periphlebitis'. Fluorescein angiography demonstrates late leakage, no vascular occlusion, & late vascular staining[2-6].

Idiopathic Frosted Branch Angiitis probably represents an immune response to an underlying stimulus such as a viral infection, in which vascular sheathing is caused by immune- complex deposition in vessel walls. This hypothesis is supported by the favorable response of FBA to systemic steroids[7,8].

2. Case Reports

Case 1. A previously healthy 35-years-old male presented with drop of vision in his left eye for the past three days. His BCVA was 20/400. Anterior segment was free of inflammation.

Ophthalmoscopy revealed mild to moderate venous dilatation and tortuosity, extensive peri-venous white exudates extending from the posterior pole to the periphery, intraretinal hemorrhages, & marked macular edema **(Figure 1)**.

Fluorescein angiography revealed extensive late staining of the vein walls, but without evidence of vascular occlusion **(Figures 2a,b,c,d)**.

CBC, ESR, total lipid profile, & Serology for HIV, HZV, & CMV were negative. Chest X-ray was normal.

Three days after the onset of symptoms, oral corticosteroids were started at a daily dose of 80 mg prednisolone for 10 days. Ten days later, his BCVA improved to 20/125. Ophthalmoscopic picture improved dramatically with almost complete resolution of intra-retinal hemorrhages, & peri-venous exudates. Macular edema improved substantially though with residual star-shaped macular exudates **(Figure 3)**.

Hence, we started tapering steroids over six weeks. The patient reported in three weeks complaining of dropped vision in the same eye. He mentioned that he stopped steroids on his own after two weeks of initiating steroid tapering.

His BCVA in the left eye was 20/400. Fundus examination revealed the same picture seen at initial presentation **(Figure 4)**. Steroids were re-instated; one week later, he developed vitreous hemorrhage and ended up with PL vision.

Case 2. A previously healthy 35-years-old male presented with drop of vision in both eyes for the past two days. His BCVA was 20/400 OU. Anterior segment was free of inflammation.

Ophthalmoscopy revealed mild to moderate venous dilatation and tortuosity, extensive peri-venous white exudates extending from the posterior pole to the periphery along with intra-retinal hemorrhages. Fluorescein angiography revealed extensive staining of the vein walls, along with vascular occlusion. **Figures (5a,b,c,d)-(6a,b,c)**.

ESR, total lipid profile, & CBC as well as chest X-ray were normal. Patient was offered steroid treatment but he did not accept it nor did he come for follow-up.

Case 3. A previously healthy 25-years-old male presented with drop of vision in his left eye for the past two days. His BCVA was 20/200. His anterior segment was free of inflammation.

Ophthalmoscopy revealed extensive peri-venous white exudates extending from the posterior pole to the periphery along with intraretinal hemorrhages. Ghost vessels were also seen **(Figures 7a,b,c)**.

Fluorescein angiography revealed extensive staining of the vein walls, with diffuse vascular occlusion **(Figures 8a, b, c, d)**.

ESR, total lipid profile, & CBC & chest X-ray were normal. Two days after the onset of symptoms, oral corticosteroid regimen was advised, consisting of 80 mg prednisolone daily for 10 days. The patient did not come for follow-up.

Discussion. Our three patients were all young, & healthy. Their age group fell in the third & fourth decades. In his review Walker *et al.,* stated that this condition predominantly affects young and healthy individuals, and that it has a bimodal age distribution with two peaks, one in childhood and the other in the third decade[2].

Two of three patients had unilateral involvement. This finding is in accordance with Stephanie et al., who stated that FBA may occur unilaterally as well as bilaterally[4].

As in previously reported cases of frosted branch angiitis, our three patients had rapid onset of visual loss, & sheathing of the retinal veins. We were able to demonstrate a prompt response to corticosteroids in one of three patients as the other two did not continue follow-up. In addition, these reports stated that the vasculitis in Frosted Branch Angiitis is usually non-occlusive. Two of our patients, however, showed angiographic evidence of diffuse vascular occlusion[2,4-6,8].

Published reports describe Frosted Branch Angiitis as primary periphlebitis. Even when arteries are affected, veins are usually more severely involved. In accordance with this statement, all three patients included in this report had venous affection only[8].

In our cases, initial visual acuities ranged from 20/400 to 20/200. We were able to monitor the response to corticosteroids in one patient who improved from a baseline BCVA of 20/400 to 20/125 (equivalent to 15 letters on ETDRS scale), 10 days following a daily dose of 80mg oral corticosteroids. That same patient later on did not follow-up the therapeutic regimen as advised, developed vitreous hemorrhage and ended up in PL vision.

Compared to previous reports, in one series the authors demonstrated improvement of BCVA to 20/30 from a baseline level of 20/400 without resorting to systemic steroids, only topical steroids were given for associated anterior uveitis[6]. In a second series that included two patients[4], BCVA improved to 20/20 from an initial level of counting fingers in one case, and a BCVA of 20/80 from an initial level of counting fingers in a second case. In both cases oral corticosteroids were given.

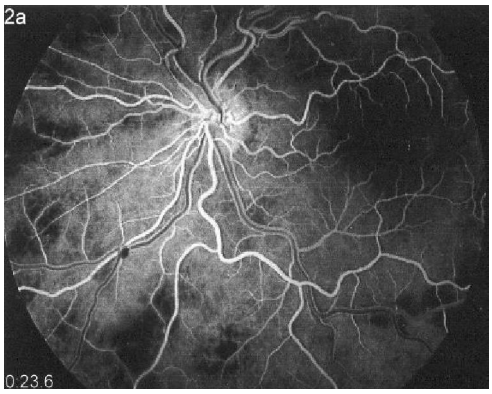
In a third case series including three cases[8], the authors recorded improvement of BCVA to 20/300 OD, and 20/20 OS from a baseline level of 4/200 & 20/400 respectively in their first case. In the second case, they demonstrated improvement of BCVA to 20/20 OU. The baseline BCVA was 20/20 OD, and 20/30 OS. Their third case had a final BCVA 20/20 OU from an initial level of counting fingers OU. All three cases received oral corticosteroids.

A fourth case report demonstrated a final BCVA of better than 6/6 OU after an initial BCVA of 6/60 OD, & 2/36 OS. The patient received systemic corticosteroids[2].

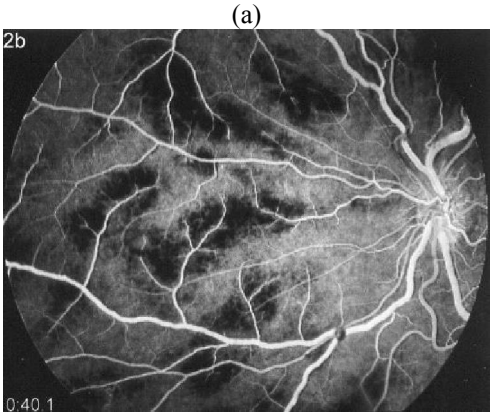
In our series, we encountered a single complication, which was vitreous hemorrhage in the first patient. Reported complications include macular scarring[4,8], vein occlusion, & retinal tear formation[8].

Although we could not delineate the natural course of the disease due to small sample size, & lack of patients' compliance with treatment regimen we were able to demonstrate the dramatic response to systemic steroids with resolution of vasculitis. This suggests that in cases of idiopathic frosted branch angiitis early diagnosis, and prompt initiation of steroid treatment is mandatory.

Figure 1. Case 1. A composite fundus photo of the left eye demonstrating thick perivascular sheathing affecting the retinal veins. Note that Intra-retinal hemorrhages & macular edema are also present.



(a)



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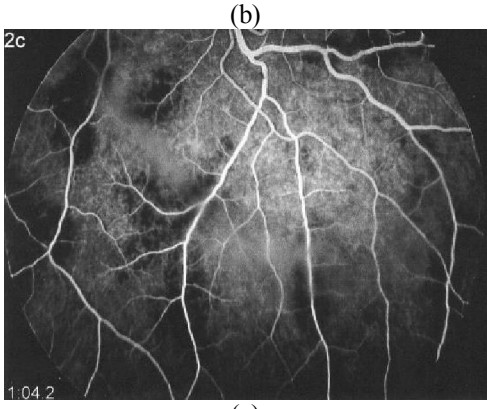

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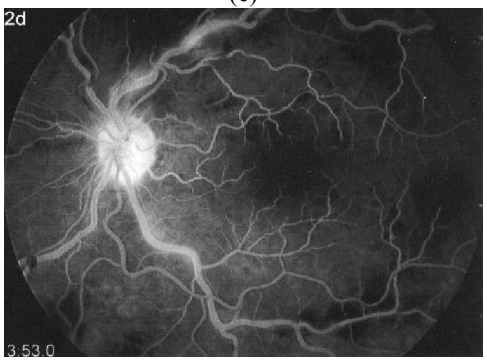


(d)

Figures 2a, 2b, 2c, 2d. Case 1. Fluorescein angiography of the same patient demonstrating normal venous flow with late staining of venous walls.

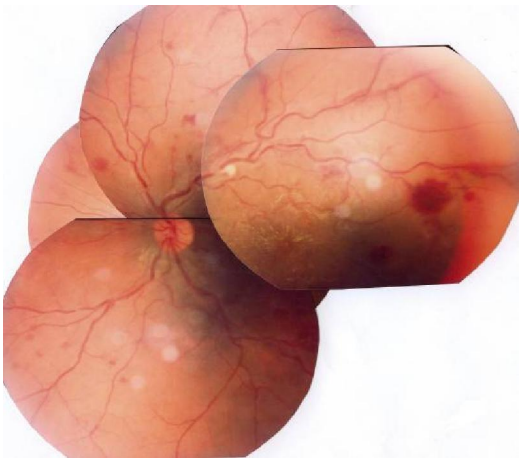


Figure 3. Case 1. Fundus appearance of the same patient 10 days after oral corticosteroids with marked improvement of perivascular exudates, & intra-retinal hemorrhages. Macular edema has resolved leaving a residual macular star.

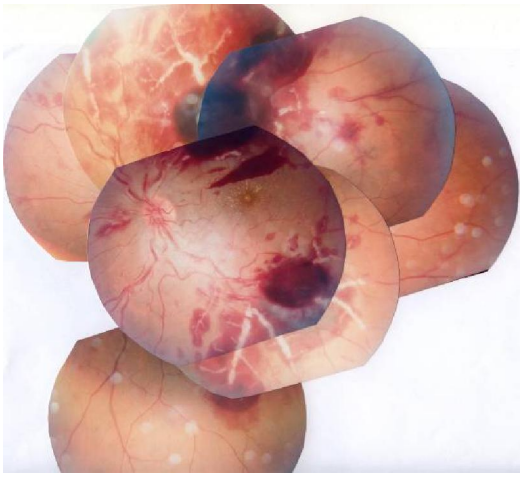


Figure 4. Case 1. Composite Fundus photo of the same patient after relapse following cessation of steroids.

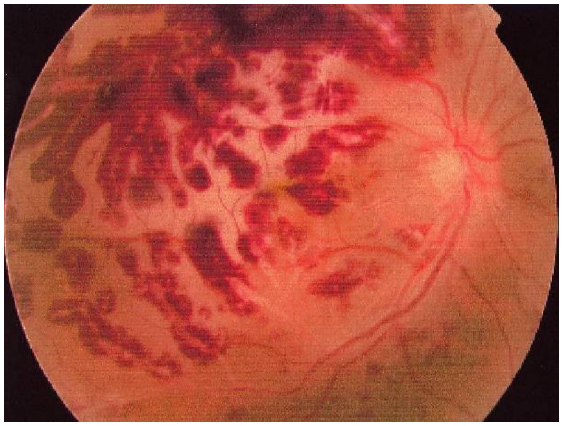
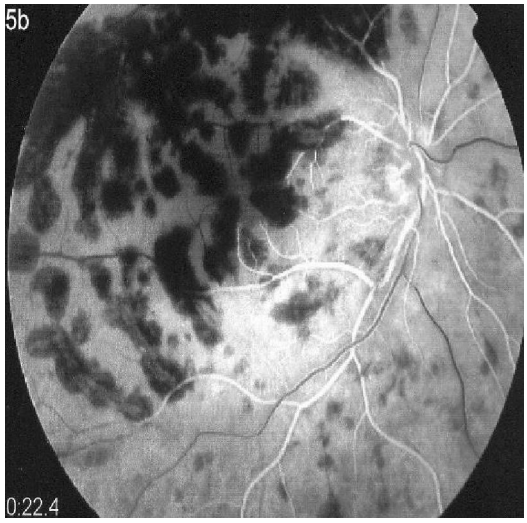


Figure 5a. Case 2. Fundus photo of the right eye showing the posterior pole with thick peri-vascular exudates affecting retinal veins, and intra-retinal hemorrhages.



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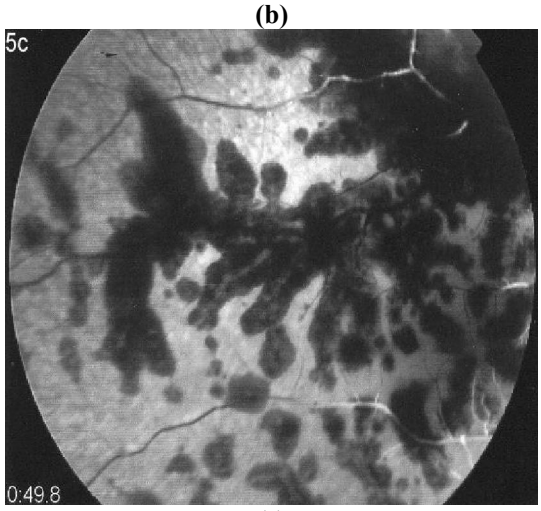

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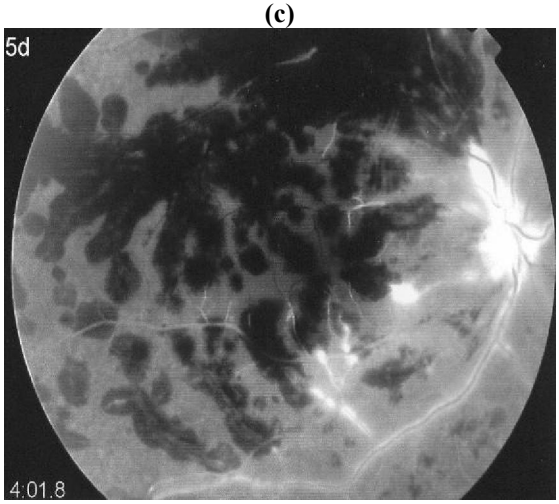

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Figures 5b, 5c, 5d. Case 2. Fundus fluorescein angiography of the same patient showing diffuse vascular occlusion along with late staining of venous walls.

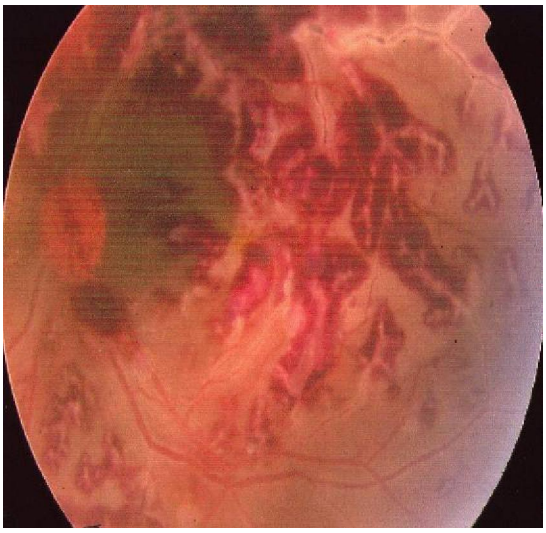
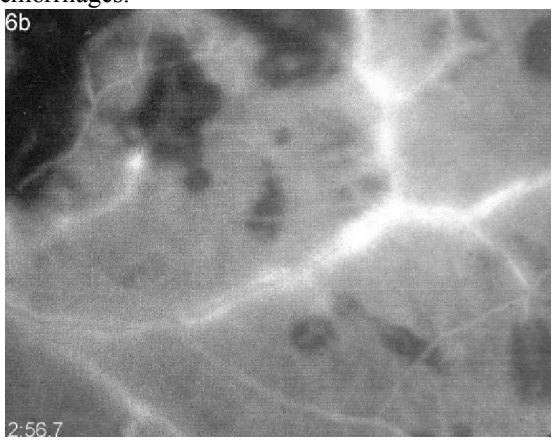


Figure 6a. Case 2. Fundus photo of the left eye showing the posterior pole with thick peri-vascular exudates affecting retinal veins, and intra-retinal hemorrhages.



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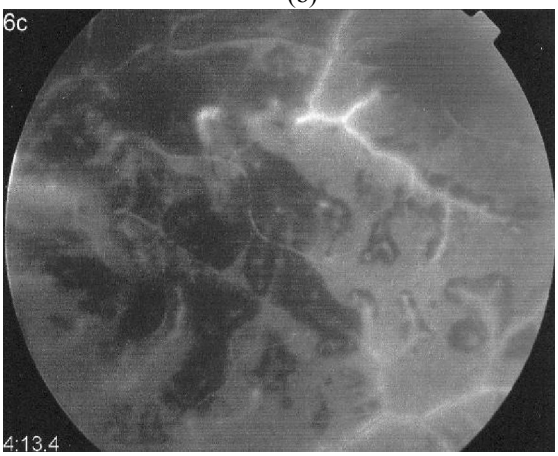

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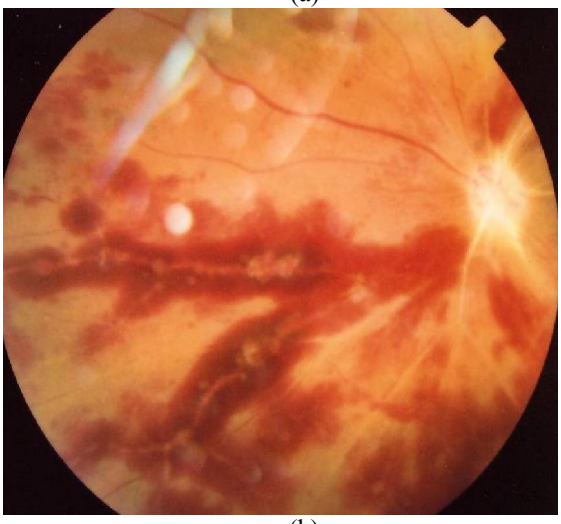
Figures 6b, 6c. Case 2. Fundus fluorescein angiography of the same patient showing late staining of venous walls.

(a)



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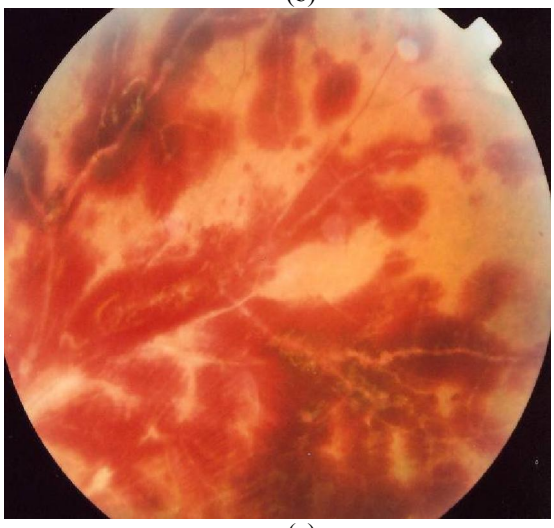

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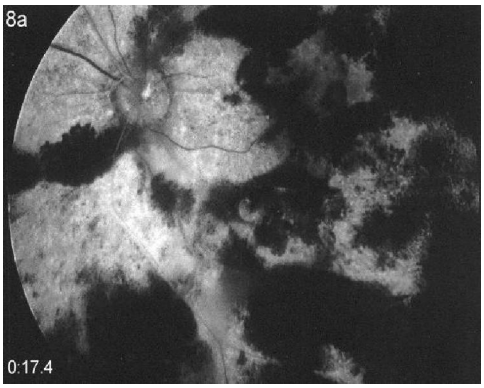
Figures 7a, 7b, 7c. Case 3. Fundus photos of the left eye showing thick peri-vascular exudates affecting retinal veins, along with intra-retinal hemorrhages. Ghost vessels are seen.



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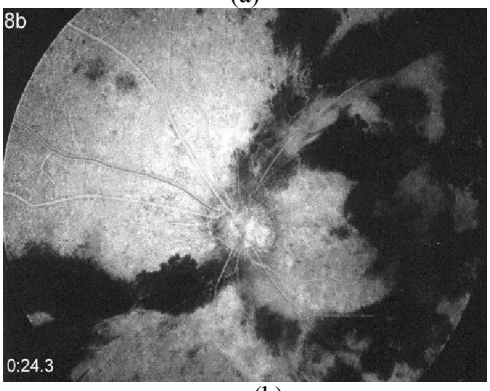

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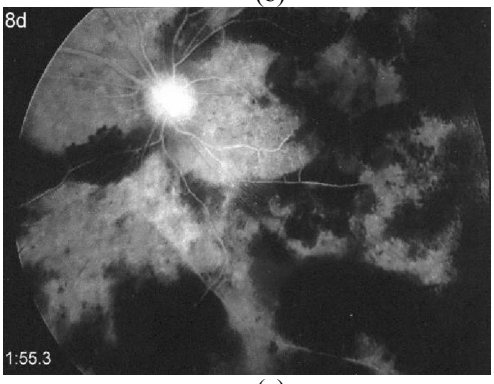

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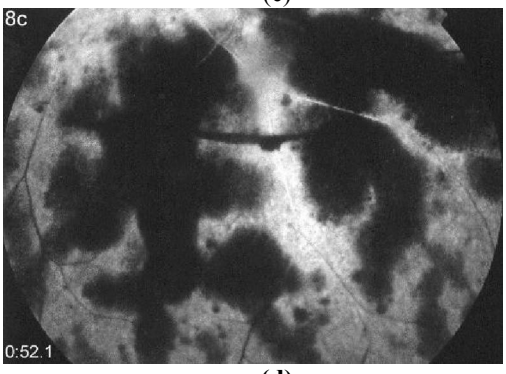

(d)

Figures 8a, 8b, 8c, 8d. Case 3. Fundus fluorescein angiography of the same patient showing vascular occlusion along with late staining of venous walls.

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11/10/2013