

BRIEF REPORTS**Frosted-branch angiitis
with central-retinal-vein occlusion:
angiographic and OCT findings**

Harvey S. Uy, MD

*Asian Eye Institute
Makati, Philippines**Sentro Oftalmologico Jose Rizal
University of the Philippines–Philippine General Hospital
Manila, Philippines***ABSTRACT****Objective**

To report angiographic and optical-coherence-tomography findings in a case of frosted-branch angiitis (FBA) with central-retinal-vein occlusion (CRVO).

Methods

This is a case report.

Results

A healthy Japanese female presented with monocular blurring of vision. Fluorescein angiography (FA) revealed extensive dye leakage from the retinal blood vessels and optic-nerve head. Indocyanine-green (ICG) angiography demonstrated tortuous retinal veins containing foci of hypofluorescence, foci of hyperfluorescence, and diffuse dye leakage. Late choroidal hypofluorescent patches were observed. Optical coherence tomography demonstrated swelling of the optic-nerve head and thickening of the sensory retina and nerve-fiber layer. Aggressive steroid treatment resulted in improved vision and angiographic appearance.

Conclusion

CRVO may be an initial presentation of FBA. FA, ICG, and nerve-fiber-layer OCT examinations are useful in monitoring disease resolution. Early and aggressive steroid treatment can help control inflammation and improve visual outcome.

FROSTED-branch angiitis (FBA) is a rare form of retinal vasculitis characterized by decreased visual acuity, white perivascular sheathing of the retinal blood vessels, anterior-chamber cells, absence of systemic involvement,

and response to steroid treatment.¹⁻¹⁰ Central-retinal-vein occlusion (CRVO) may develop in some patients with FBA.¹¹ We describe here the fluorescein (FA) and indocyanine-green (ICG) angiographic and optical-coherence-tomographic (OCT) findings in a patient with FBA and CRVO.

A healthy, 34-year-old Japanese female experienced rapidly progressive blurring of vision in the left eye. On presentation, the best-corrected visual acuity was 20/400. There were 2+ cells in the anterior chamber. Dilated-fundus examination revealed extensive white perivascular sheathing of engorged, tortuous retinal veins. Macular and papillary edema were present. FA demonstrated diffuse dye leakage from the retinal veins, optic-nerve head, and macula. Areas of ischemia and intraretinal hemorrhages appeared as hypofluorescent patches (Figure 1A-B). The right eye was completely normal.

ICG revealed beading of the retinal veins, intravascular-filling defects, and diffuse posterior-pole hyperfluorescence which obscured visualization of the choroidal blood vessels. Foci of intense intravascular hyperfluorescence suggestive of focal intravascular inflammation and foci of intravascular hypofluorescence suggestive of vascular occlusion were visible both in the retinal arteries and veins. Late-phase patches of choroidal hypofluorescence were observed suggesting involvement of the choriocapillaris or larger choroidal blood vessels (Figure 1C).

The patient was diagnosed to have FBA with CRVO and was treated with a periocular injection of triamcinolone acetonide (40 mg), prednisolone acetate eye drops, and oral prednisone (1 mg/kg/day). Over the next 3 months, the visual acuity returned to 20/50. The fundus improved in appearance with reduction of cotton-wool spots and foveal hard exudates. Multiple retinal arterial plaques were visible. FA demonstrated return of retinal veins to a near-normal appearance with residual patches of ischemia and late disc hyperfluorescence. ICG demonstrated disappearance of retinal blood vessels abnormalities but persistence of late-phase choroidal hypofluorescence (Figure 2).

Optical coherence tomography (Stratus OCT, Carl Zeiss Meditec, Dublin, CA) performed after 2 weeks of steroid therapy revealed optic-nerve-head (ONH) swelling with reduction of the optic-cup area (0.072 mm² compared to 0.435 mm² for the uninvolved eye). The nerve-fiber layer was thickened (114.9 μm compared to 87.1 μm for the uninvolved eye). OCT of the macula showed normal retinal configuration and thickness (217 μm) (Figure 3).

FBA is a clinically distinct form of retinal vasculitis. Since its initial description by Ito in 1976, several reports of FBA

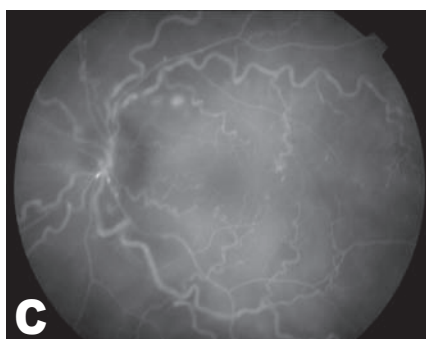
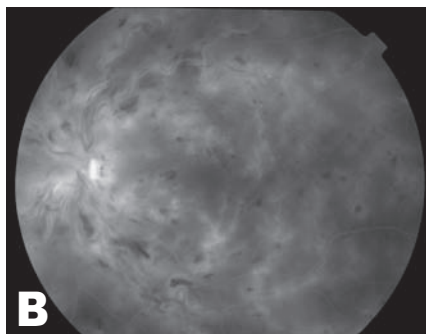
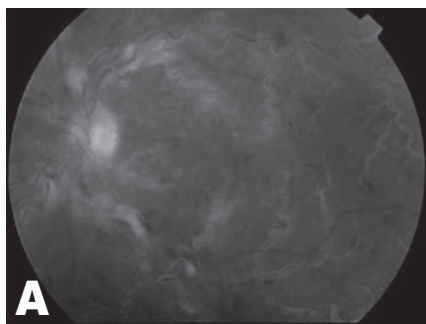


Figure 1. Fundus photograph demonstrating extensive, white perivascular sheathing resembling frosted tree branches. Engorged retinal veins, hemorrhages, and papillary and macular edema characteristic of central-retinal-vein occlusion can be seen (A). Fluorescein angiography demonstrates dye leakage from retinal blood vessels and the optic-nerve head (B). Late-phase indocyanine-green angiography demonstrates abnormalities of vascular caliber with prominent foci of intravascular hypofluorescence and hyperfluorescence as well as patches of choroidal hypofluorescence (C).

have been published; however, the underlying pathophysiologic processes are still poorly understood. While most cases are idiopathic in nature, some cases have been associated with hematologic malignancies, viral infections, and autoimmune disease.¹⁻¹² It is unclear whether FBA is a unique disease entity or a common presentation resulting from several causes.¹² In this patient, no systemic associa-

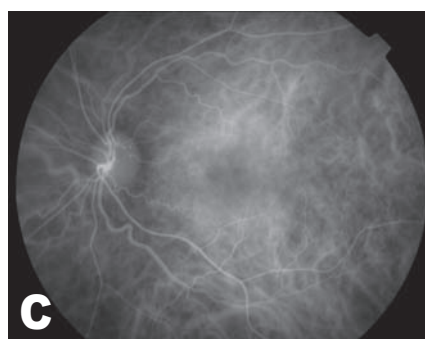
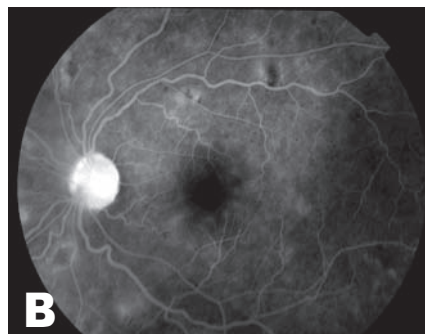
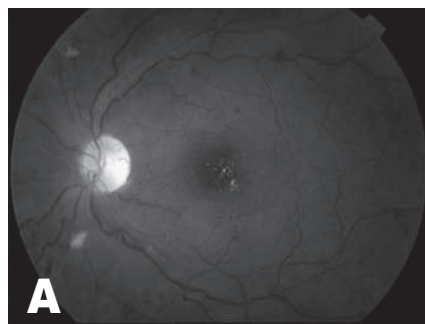


Figure 2. Fundus photograph taken 2 months after steroid treatment demonstrates return of retinal blood vessels and optic-nerve head to near-normal appearance. Residual hemorrhages, hard exudates, cotton-wool spots, and a few foci of intra-arterial plaques are visible (A). Fluorescein angiography demonstrates a few residual ischemic areas (B). Indocyanine angiography demonstrates normal retinal vessels and persistence of choroidal hypofluorescent patches (C).

tions were found and no etiologic cause was established.

Masuda and coworkers found areas of choroidal hypofluorescence during ICG angiography in a case of FBA without CRVO. They surmised that areas of choroidal hypofluorescence are suggestive of choroidal vascular occlusion or choroiditis.¹⁰ In our patient, the retinal blood vessels exhibited three types of findings

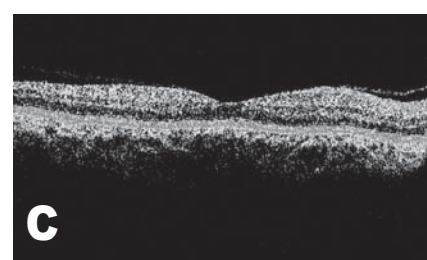
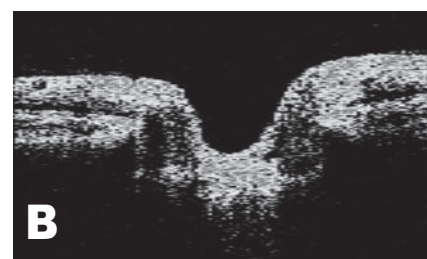


Figure 3. Optical coherence tomography demonstrates optic-nerve-head swelling of the involved eye (A); normal optic-cup configuration of the uninvolved eye (B); and normal foveal configuration and thickness (C).

during FA and ICG angiography:

1. foci of intravascular hypofluorescence indicating an occlusive process,
2. foci of intense hyperfluorescence suggesting multifocal inflammatory processes, and
3. segments with mild dye leakage indicating diffuse vascular leakage and blood-retinal-barrier disruption.

The involvement of both retinal and choroidal blood vessels suggests that a common vascular antigen acts as the inciting agent for FBA.

The most prominent OCT finding was ONH swelling similar to that found in central-vein occlusion. There was also mild thickening of the sensory-retina and nerve-fiber layers. We are not able to determine whether CRVO in conjunction with FBA stems from occlusive vasculitis causing

thrombosis or optic neuritis causing luminal occlusion. Retinal OCT did not demonstrate cystic changes or retinal thickening and was not helpful in establishing nor monitoring the disease.

Kaburaki and associates reported two cases of FBA associated with CRVO.¹¹ Both patients developed neovascular glaucoma (NVG) and progressive worsening of vision despite initial improvement with systemic steroids; CRVO and NVG developed when the steroid dosage was tapered. In contrast, the initial presentation of our patient was FBA with CRVO. Prolonged high-dose oral-steroid intake given over 3 months resulted in steady improvement of visual acuity and angiographic appearance. This case suggests that prolonged steroid intake may be necessary in order to prevent progression of a vascular occlusive process. The good response to steroid treatment with return to normal angiographic appearance of the retinal and choroidal blood vessels also suggests that the occlusive process is primarily inflammatory in nature and is reversible. While care should be taken prior to instituting steroid treatment particularly in the background of an immunocompromised status, caution should likewise be exercised when reducing steroid dosage especially when there is an initial good response to steroid intake.

This case report reveals that CRVO may be an initial presentation of FBA and that early, vigorous steroid treatment may be beneficial for these patients. FA, ICG, and nerve-fiber-layer OCT examinations were useful in monitoring disease resolution. Early and aggressive steroid treatment was beneficial in controlling inflammation and improving visual outcome.

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A case of intracranial chordoma associated with multiple-cranial-nerve palsy

Ali A. Bodla, MD
Syed A. Ali, MRCS
Bruce Hutchison, FRCS Ed, FRCOphth

*Eye Unit, The Ayr Hospital
Scotland, United Kingdom*

ABSTRACT

Objective

To report a case of intracranial chordoma associated with multiple-cranial-nerve palsies, highlight its presenting features, and evaluate treatment options for this rare neoplasm.

Methods

This is a case report.

Results

A 77-year-old man presented with diplopia and left sixth-nerve palsy. Further investigations revealed an intracranial chordoma. Surgical decompression was performed which resulted in complete resolution of the diplopia. Three months postsurgery, he was noted to have a right-third-nerve palsy. Repeat MRI showed tumor recurrence. Targeted proton-beam-charged-particle therapy was performed.

Conclusion

Radiation therapy using targeted proton-beam-charged particle was shown effective in the treatment of intracranial chordoma, resulting in significant improvement in ocular motility and resolution of ptosis.

CHORDOMAS are rare neoplasms that constitute only 0.2% of central-nervous-system (CNS) tumors. The most common site is the sacrococcygeal region (50%), followed by the intracranial region (30%).¹ We report a case of an intracranial chordoma presenting with diplopia and describe its response to treatment.

A 77-year-old man was referred to the clinic complaining of diplopia. On examination, a left-sixth-nerve palsy was noted. Magnetic-resonance imaging (MRI) revealed an expanding sellar mass consistent with a pituitary tumor (Figure 1). Surgical decompression was performed, with complete resolution of the diplopia. Complete resection, however, was not possible. Histological studies showed an