

A Rare Case of Combined Vascular Block

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Abstract

Papillophlebitis or central retinal vein occlusion (CRVO) of the young is a condition affecting healthy adults, less than 50 years of age. We report a case of papillophlebitis with cilioretinal artery occlusion along with its fundus fluorescein angiography (FFA) and Spectral Domain Ocular Coherence Tomography (SD-OCT) features. A 27 year old female with no systemic illness reported with sudden onset diminution of vision for 1 day. Fundus examination revealed optic nerve head edema, dilated tortuous veins and whitening of papillomacular bundle suggestive of cilioretinal artery occlusion. Fundus fluorescein angiography showed normal filling of cilioretinal artery and no arteriovenous delay. OCT showed hyperreflective inner retinal layers with obscuration of external limiting membrane. Prompt therapy with high dose intravenous steroids lead to early resolution and better visual prognosis.

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Introduction

Papillophlebitis or central retinal vein occlusion (CRVO) of the young is a condition affecting healthy adults, less than 50 years of age.¹ The patient presents with slight blurring of vision with or without photopsia.² It is largely an idiopathic phenomenon, however, association with certain underlying systemic conditions have been described.³ Serology and imaging are important to rule out other etiologies of disc edema.

We report a case of papillophlebitis with cilioretinal artery occlusion, along with its fundus fluorescein angiography (FFA) and spectral domain ocular coherence tomography (SD-OCT) features.

Case Report

A 27 year old female with no systemic illness reported with sudden onset diminution of vision for 1 day. The diminution of vision was non progressive and there was no pain on ocular movements or retro-orbital pain. There was no history of previous similar episode. There was no history of any oral contraceptive use or any coagulopathy.

Cardiovascular examination was within normal limits with no added sounds.

The best corrected vision in the right eye was finger counting at 1 meter while in the left eye, it was 6/6 on Snellen's chart. Color vision and contrast sensitivity were normal in both eyes and there was no afferent pupillary defect.

Fundus examination consisted of slight blurring of disc margins, with elevation of the optic nerve head and filling of cup. The veins entering the disc were dilated and tortuous, with arteriovenous ratio of 1:3. Few flame shaped hemorrhages were noted on the posterior pole and mid-periphery. An area of retinal whitening was noted in the papillomacular bundle with surrounding cuff of fluid (Figure 1). The left fundus examination was normal (Figure 2). FFA showed normal filling of cilioretinal artery in the early phase, and normal arteriovenous transit time. In the late phase, however, there was staining of the temporal disc margin with minimal leakage (Figure 3).

OCT examination was suggestive of hyperreflective inner retinal layers, with obscuration of external limiting



Figure 1: Colored fundus photograph of the right eye showing venous stasis and combined central venous and cilioretinal artery occlusion



Figure 2: Colored fundus photograph of the left eye

membrane (ELM). There was associated intraretinal cystoid spaces and increased retinal thickness (Figure 4).

On hematological investigations, there was neutrophilia and inflammatory markers like Erythrocyte Sedimentation Rate and C - reactive protein were raised. Investigations to rule out any rheumatologic and infective disorders

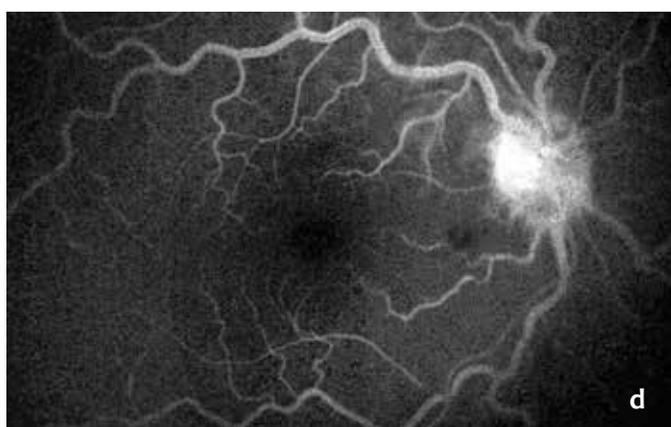
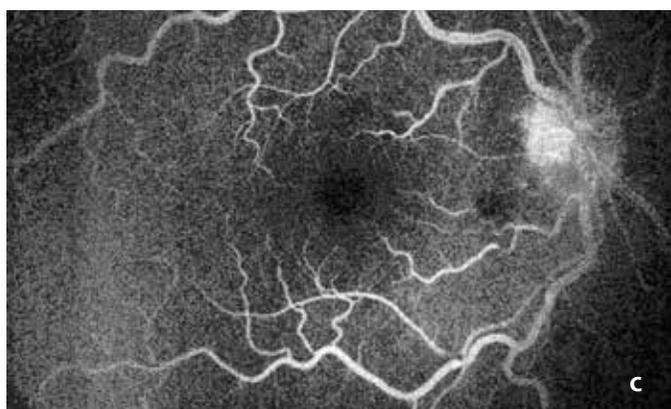
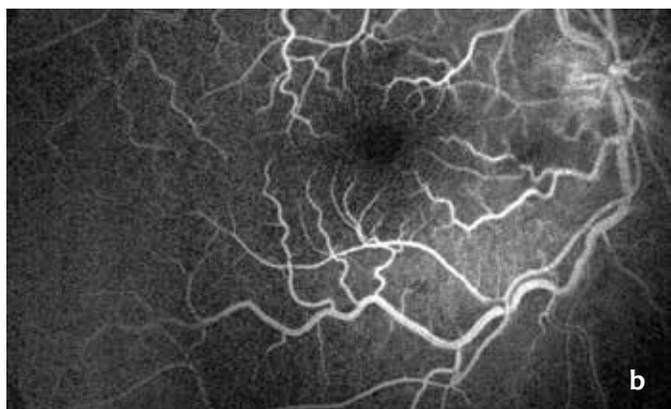
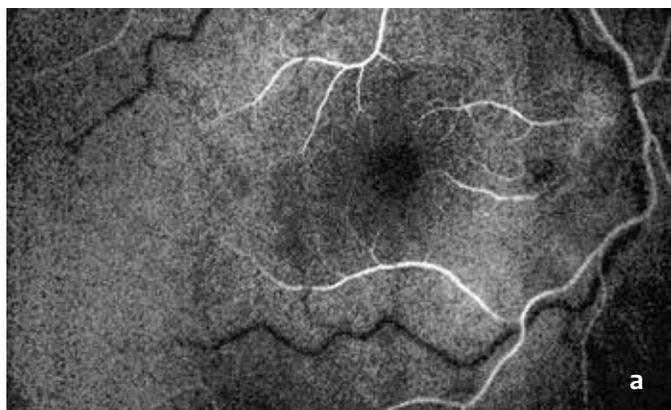


Figure 3: Fundus fluorescein angiography, (a) showing normal cilioretinal artery filling (b) normal AV transit phase (c) venous dilation and tortuosity (d) late staining of the disc.

were negative. Detailed workup for any thromboembolic phenomenon including complete coagulation profile, serum homocysteine markers were within normal limits.

On the basis of the clinical picture and raised inflammatory markers, we diagnosed the patient as a case of papillophlebitis, with cilioretinal artery occlusion and managed the patient with intravenous pulse dexamethasone, 100 mg in 150 ml Ringer Lactate for three days. On the third day, the best corrected visual acuity in the right eye was 6/12, with complete resolution of macular edema. The inner retinal layer hyper reflectivity also decreased and ELM was visible (Figure 5).

The patient was continued on oral prednisolone 1 mg/kg weight for 11 days, with taper over 3 days.

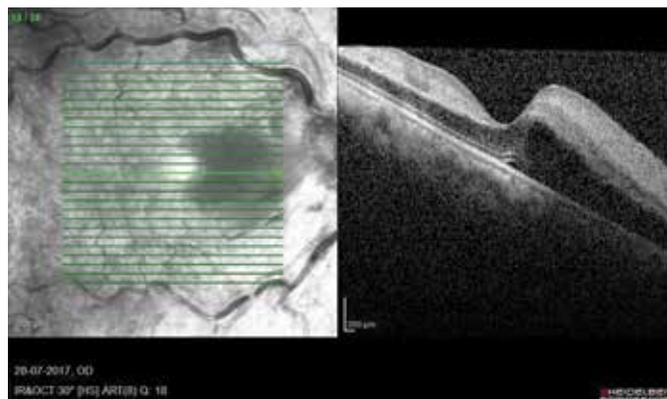


Figure 4: Macular OCT of the right eye before treatment showing hyperreflectivity of inner retinal layers

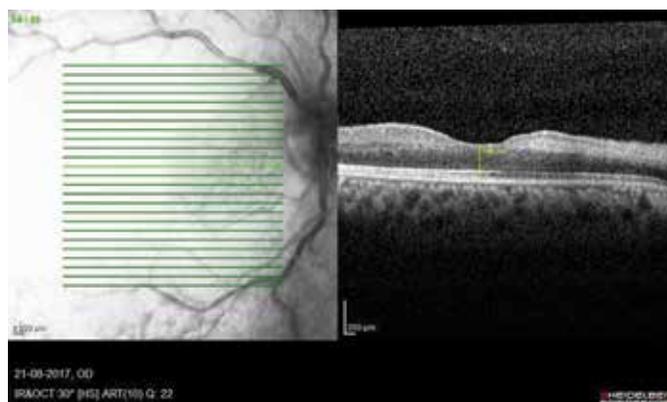


Figure 5: Macular OCT of the right eye after treatment showing decreased reflectivity of inner retinal layers

Discussion

Papillophlebitis as a syndrome was first described by Lonn and Hoyt⁴ in 1966 consisting of painless optic disc edema, venous dilatation and tortuosity, variable hemorrhages and macular edema in a young adult. Over past decades, this syndrome has been variously known as big blind spot syndrome⁵, optic disc vasculitis⁶ or presumed phlebitis of optic disc⁷ etc. The patient is usually asymptomatic, or may present with slight unilateral blurring of vision. Males may be more affected than females; however, there is no consistent report on the sex predilection. Idiopathic

inflammation of the veins at the optic nerve head, along with disc edema, plays the major role in the pathophysiology. Congestion at the disc leads to impaired venous flow in the central retinal vein and resultant venous tortuosity and dilation. Hayreh⁶ summarized the previous cases described in literature and his own case series and categorized these patients in two subgroups, one with paucity of hemorrhages and no macular involvement and the other with extensive hemorrhages, macular edema and poor visual prognosis.

Our patient presented with retinal whitening in the area of the papillomacular bundle and severe visual loss suggestive of a cilioretinal artery occlusion (CLRAO). A combined central retinal venous occlusion with a cilioretinal artery occlusion has the above described fundus findings, along with delayed arterial dye filling and prolonged arteriovenous transit time on FFA. The angiographic features were absent in our case, and so combined vascular block was ruled out.

The mechanism of CLRAO in a patient with papillophlebitis is based on edema and disc congestion, which leads to disruption of retinal blood flow, and venous stasis. This venous stasis and mechanical compression leads to decreased hydrostatic pressure in the cilioretinal artery giving a picture of CLRAO.⁸

Papillophlebitis is a non specific inflammatory phenomenon and is a diagnosis of exclusion. This was supported by our finding of increased inflammatory markers and negative serology.

The treatment comprises of suppression of inflammation by high dose steroids, which as described above, resulted in a significant improvement in visual acuity. This view was also shared by Hayreh⁶, who suggested that steroid therapy leads to early visual rehabilitation and better visual prognosis.

An extensive workup for thromboembolic phenomenon was done in our case for two reasons. The first was to distinguish papillophlebitis from an embolic vascular block. Secondly, as patients with papillophlebitis have a higher lifetime risk of thrombophlebitis, care must be taken to recognize and manage any predisposition to embolus formation.

Follow up should include monthly vision assessment, color fundus photography; visual field charting and a look-out should be done for connective tissue disorders and coagulative states.

We also suggest that in a young female patient such as ours, oral contraceptive pill use should be discouraged to prevent blood hypercoagulability.

The present case aims to emphasize the importance of clinical suspicion and prompt institution of anti-inflammatory therapy for better prognosis.

Conclusions

Papillophlebitis is a rare cause of combined vascular occlusion. A high index of suspicion in a young patient with venous occlusion and prompt therapy can result in a good visual recovery.

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