# Papillophlebitis, Optic Disc Vasculitis

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Papillophlebitis, or optic disc vasculitis, is an uncommon ophthalmologic condition of unknown cause that is often confused with other ocular pathology. In 1880, Henry Eales first described this condition as *primary retinal hemorrhage* in young men.[1] Since then, papillophlebitis has been termed *retinal vasculitis,*[2] *benign retinal vasculitis,*[3] and *optic disc vasculitis.*[4] Lonn and Hoyt[5] reported this same condition as papillophlebitis in 1966.

Papillophlebitis is identified by historical and clinical characteristics. The condition usually occurs in healthy young adults, more commonly in men, and is invariably unilateral. Most patients complain of blurred vision and minor impairment of visual acuity. Typical ophthalmologic findings include significant optic disc edema with distended retinal veins and frequent retinal hemorrhages. Retinal arterial involvement is minimal. Visual field testing demonstrates enlargement of the blind spot. Residual deficits are rare. Treatment with corticosteroids has produced disappointing results. The following case exemplifies an unusual finding of papillophlebitis.

## Case Report

A 19-year-old male university student complained of a grayish blurring of his right central visual field when he awoke from sleep one morning. In addition, he described a frontal headache, which was partially relieved by the ingestion of two aspirin tablets. The student reported no recent trauma, hypertension, nausea or emesis, fever, associated neurologic deficits, or history of a similar event in the past. He had experienced a significant concussion following a rugby injury 4 years before the date of presentation. The patient reported that he did not abuse drugs or use tobacco, but that he did consume beer occasionally. He had no chronic medical problems or allergies and was not taking medication at the time of presentation. He did wear hard contact lenses, but denied using them incorrectly.

Physical examination revealed an apparently healthy young man. His vital signs were normal. His best-corrected visual acuity was 20/25 in the left eye and less than 20/400 in the right eye. The sclera and conjunctiva were normal. He had a 1+ to 2+ Marcus Gunn pupil in the right eye, with moderate disc edema, venous engorgement, and local areas of hemorrhage (Figure 1). The left eye was completely normal, as was the remainder of the physical examination, including a neurologic examination. The patient was admitted to the hospital for evaluation of unilateral papilledema.

Laboratory studies, including a complete blood count, coagulation studies, and a 12-test serum profile, were normal. A computed tomography scan of the head and orbits showed no evidence of intracranial masses, edema, or optic nerve or orbital abnormalities. The lumbar puncture opening cerebrospinal fluid pressure was 180 mm $H_2O$ and the closing pressure was 174 mm $H_2O$. The fluid was clear, and protein electrophoresis studies, cytology, cell count, Gram stain, and cultures were negative. An erythrocyte sedimentation rate and antinuclear antibody test were normal.

An ophthalmologist was consulted and, following his recommendation, the patient was started on intravenous methylprednisolone, 1 g every 12 hours. Steroids were recommended for two reasons: (1) the case presentation was somewhat atypical for papillophlebitis; and (2) the real existence of papillophlebitis is controversial. Other entities were included in the differential diagnosis, such as optic neuritis, for which the patient was also initially treated. A fluorescein angiogram demonstrated a serous macular detachment, which probably explained his visual deficit. Other angiographic characteristics for papillophlebitis, retinal vein-filling delay, and contrast agent leakage from dilated veins and the optic nerve were not observed. The patient was discharged and given a

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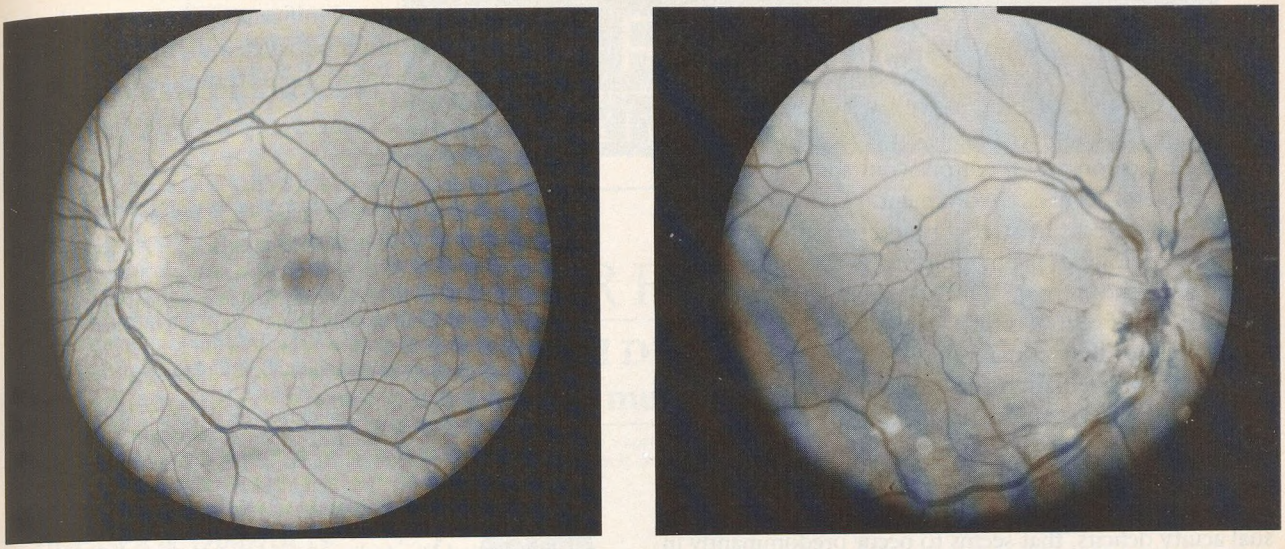

Figure 1. Left: Angiogram shows that the left eye of 19-year-old student is normal. Right: The right eye reveals characteristics of papillophlebitis. The optic disc is edematous, retinal veins are mildly dilated, and there are exudates and hemorrhages present.

prescription for oral methylprednisolone, the dosage of which was to be tapered over the subsequent 2 weeks.

The patient returned for a follow-up examination 3 months after being discharged. His fundoscopic examination was normal, and the vision in his right eye had refracted to 20/25 +1. Symptomatically, the patient had a complete recovery, which further supported a diagnosis of either papillophlebitis or optic neuritis.

## Discussion

The case demonstrated the typical classic features of papillophlebitis, except for the noted loss of vision, which is very unusual. Consequently, other ocular abnormalities were included in the differential diagnosis.

Optic neuritis and papillophlebitis have similar common features. Papillophlebitis is thought to be an inflammatory process of retinal veins. Optic neuritis is an inflammatory disease of the optic nerve, resulting from demyelinative, autoimmune, or infectious processes. Optic neuritis affects young adults but generally results in an acute, profound, unilateral loss of vision, usually in proportion to the degree of disc changes visualized. Retrobulbar pain and tenderness, particularly with extraocular movement, are common symptoms of optic neuritis. Central visual field defects are seen, as was noted in this case. Clinically, afferent papillary defects (Marcus Gunn pupil) are also common. Ophthalmologic findings vary. Frank inflammation and edema of the nerve head may be observed; however, retrobulbar inflammation can occur without clinically apparent nerve head changes. In contrast to papillophlebitis, retinal venous changes are not seen in cases of optic neuritis. Visual deficits frequently resolve spontaneously in cases of optic neuritis. Recently, high-dose intravenous steroids have been used successfully to treat optic neuritis; however, this practice is still somewhat controversial. Nevertheless, visual acuity improves more rapidly when steroids are used, and steroids may prevent further neurologic impairment or reverse existing impairment.[6] Because of these similarities and the atypical presentation, the patient described was treated initially with steroids.

The fundoscopic finding of central retinal vein occlusion (CRVO) or thrombosis is similar to that found in papillophlebitis. CRVO-associated hemorrhages are more centrally located, more extensive, and more numerous, however.[2] The macula is damaged more extensively, which results in central visual field deficits, as seen in this case. However, the vascular changes that occur in patients with hypertension and arteriosclerosis are not seen with papillophlebitis, and those changes are typically bilateral.

Papillophlebitis can be confused with optic nerve edema secondary to increased intracranial or intraorbital pressure, since both exhibit papilledema. Yet, the hemorrhages and venous dilatation of papillophlebitis are not limited to the disc, and often extend more peripherally. Each condition can cause visual acuity deficits, but the defect associated with papillophlebitis is usually temporary. Papilledema from increased intracranial pressure usually causes transient visual obscurations. A protracted increase in intracranial pressure may result in optic nerve

atrophy and more permanent acuity changes. Evidence of an intracranial space-occupying lesion or tumor would support optic nerve edema. Hypertensive retinopathy in contrast to papillophlebitis is identified by arteriolar narrowing, arteriovenous crossing changes, possible nerve-fiber-layer hemorrhages, cotton-wool spots and, invariably, bilateral ocular involvement. The patient also exhibits an increase in arterial blood pressure.

The etiology of papillophlebitis is controversial and is not well understood.[7] One explanation proposed by Hayreh[4] is that central retinal vein occlusion results from a phlebitis of the retinal veins surrounding the optic nerve. The inflammation then promotes venous thrombosis, which produces papilledema, while sparing arterial compromise.

In summary, papillophlebitis is a unilateral, clinically severe-appearing retinopathy, usually with minimal visual acuity deficits, that seems to occur predominantly in healthy young people and subsequently results in a complete recovery. Other, significant optic nerve and retinal disease processes must be ruled out in the differential diagnosis through appropriate studies and consultation.

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