Case Report

Prepapillary Arterial Loop Associated with Central Retinal Artery Occlusion: A Case Report

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Abstract

Purpose: To report a case of central retinal artery (CRA) occlusion secondary to prepapillary loop in a 13-year-old girl.

Methods: A 13-year-old girl presented with a history of sudden visual loss in her left eye.

Results: Fundus examination confirmed thrombosis in a prepapillary arterial loop causing CRA occlusion and extensive retinal ischemia. Macular region was watered by an anomalous macular branch, which explained her 20/20 vision central vision.

Conclusion: Congenital prepapillary vascular loops are rare and usually asymptomatic. We report a case of central artery occlusion confirmed by multimodal imaging.

Keywords: Central retinal artery occlusion, Multimodal investigation, Prepapillary vascular loop, Vascular thrombosis

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Introduction

Prepapillary vascular loop (PVL) is a rare retinal vascular anomaly with an incidence of approximately 0.01% in the total population.[1] Fundus examination, fluorescein angiography (FA), and histopathological examinations confirmed that PVL originates from a branch of either retinal artery or vein ending at the disk and projects for a variable distance into the vitreous.[2] The majority of the cases are unilateral and asymptomatic, presenting as occasional findings in ophthalmologic examination.[1] However, they can complicate with thrombosis and branch retinal artery occlusion, amaurosis fugax, and vitreous hemorrhage, more commonly in young patients.[2] Occlusion of the central retinal artery (CRA) has been never described.

Case Report

A 13-year-old girl presented to the emergency department due to sudden visual field loss in her left eye. Initially, she noticed a bottom field defect that progressed to nasal and upper defects. There were no other ocular or systemic symptoms, and her medical history was unremarkable. Her best corrected visual acuity was 20/20 bilaterally. Anterior segments were normal, and the intraocular pressure was 10 mmHg bilaterally. In the left eye fundoscopy, there were extensive retinal pallor, peripapillary retinal hemorrhage, and perimacular cotton wool spots [Figure 1a]. There was also a PVL, extending anteriorly from the disk into the vitreous [Figure 1b]. The right fundus was normal.

In the left eye, FA showed normal choroidal fluorescence. A macular branch was filled after choroidal flush during the early arterial phase. This branch left from CRA before the formation of the vascular loop and extended through the macula, suggesting an anomalous intraneural branch of the

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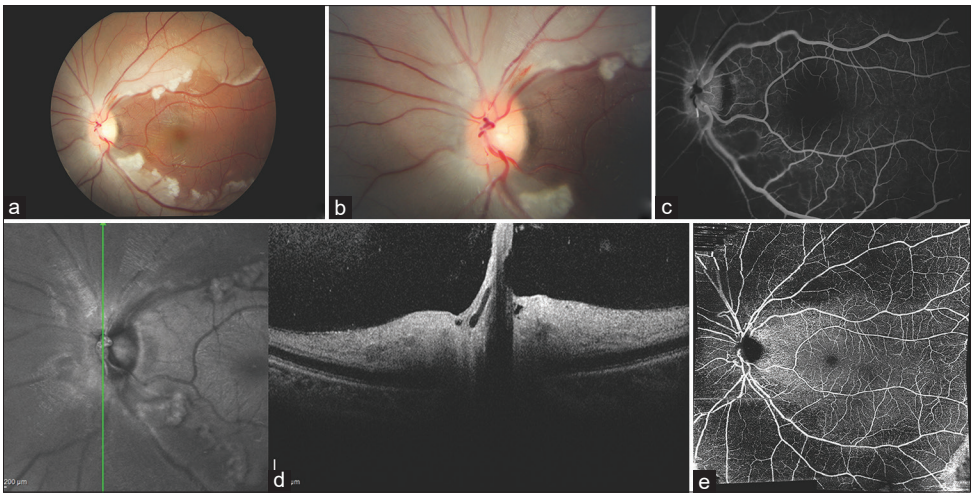

Figure 1: (a) Left eye retinography showing retinal pallor, sparing the macular region. Note peripapillary retinal hemorrhage and perimacular cotton wool spots. (b) Left eye retinography. Details of the peripapillary region. Note the arterial macular branch and the vascular loop leaving the disk area. (c) Arterial-venous phase of fluorescein angiography of the left eye. Note filling of the macular branch leaving the optic nerve head and the darkness of the peripheral retina. Note absence of dye on the vascular loop. (d) Optical coherence tomography (OCT) of the optic nerve of the left eye of the patient taken at 2 weeks after presentation. OCT shows a tubular structure leaving the optic head disc toward vitreous cavity and diffuse intracellular retinal edema. (e) OCT angiography (image overlap of multiples 8 × 8 scans) of the left eye. Note reduced blood flow in periphery with capillary dropout

CRA emerging at the level of the optic disk [Figure 1c]. There was no filling of the vascular loop, neither the temporal nor nasal arterial arcades, due to a thrombus within the loop [Figure 1c], characterizing a case of "presumed CRAO". Visual field testing revealed a peripheral defect in the left eye, sparing her central vision. Ocular coherence tomography (OCT) demonstrated diffuse intracellular retinal edema [Figure 1d]. We conducted an extensive systemic investigation including hemoglobin electrophoresis, autoantibodies, cholesterol and triglycerides levels, erythrocyte sedimentation rate, and C-reactive protein levels, as well as transesophageal echocardiography and Doppler imaging of the carotid.

No therapy was undertaken, and the patient was followed for 4 months. Visual acuity in the right eye remained 20/20. The nerve fiber layer edema subsided, and the preretinal arterial loop appeared as a subtle white ghost vessel devoid of blood. At this time, we decided to perform ocular coherence tomography angiography (OCTA) to study retinal perfusion. This revealed reduced blood flow in the retinal capillaries, sparing the macular area [Figure 1e]. Since OCTA demonstrated an ischemic pattern in the periphery, we decided to perform panretinal laser therapy to reduce the risk of neovascularization. The patient is now being followed at 3-month intervals, and there has been no evidence of neovascularization or changes in visual field defects.

Written informed consent was obtained from the study participant, and the research was conducted with approval and ethical review.

Discussion

In this report, we discuss a case of a 13-year-old child with CRA occlusion secondary to thrombus in a vascular loop. To our knowledge, we are the first to report a case of CRA occlusion secondary to a vascular loop. Previous studies have only reported branch artery retinal occlusion.[3-7] Furthermore, we are the first to describe retinal ischemic changes using OCTA in these cases.

Prepapillary loop is a rare congenital vascular anomaly, first described by Liebreish in 1871,[2] and since then, few cases have been published. They originate from the retinal arterial system (not related to the hyaloid artery), travel anteriorly into the vitreous cavity, and return to the optic disk to participate in the retinal vascular system.[1,6] The loops may be single and may take one or more spiral turns, as in our case. Recently, Mansour *et al*. attempt to classify them according to their morphologic type, with different pathogenesis during early embryogenesis.[2] We believe our patient would be classified as type four (loop protrude into vitreous). Ninety-five percent have arterial origin, with few cases originating from veins.[1]

Eyes with PVL usually present other concomitant vascular anomalies: presence of a cilioretinal artery is the most prevalent,[8,9] occurring in 75% of cases. Our patient presented an uncommonly long macular branch, extending through the macula up to the temporal periphery [Figure 1c]. This was the first branch to leave CRA, followed by the vascular loop. Since the thrombus was located within the loop, only the territory irrigated by this branch was able to maintain perfusion, allowing the uncommon finding of good central vision of 20/20 in a case of CRA occlusion.

Complications associated with PVL are rare. There are reports of associated thrombosis, amaurosis fugax, vitreous hemorrhage, hyphema, and branch retinal artery secondary to thrombus formation within the loops.[7,8] In a previous series report, Bisland reported the incidence of 4% of arterial

occlusion in PVL, all of them presenting as branch occlusion.[10] The retinal territory irrigated by the loop varies considerably; however, it appears to be most commonly associated with inferior retinal vessels.[1,2,10] Our case is unusual in that it presented with CRA occlusion, a finding we believe is first-ever described.

After 4 months of follow-up, peripheral visual field defects persisted, and the retina acquired normal color, had no edema, hemorrhages, or exudates. There was no neovascularization, and the angle was open. We conducted an OCTA that revealed reduced perfusion in the peripheral retina, suggesting ischemia. These findings emphasize the importance of the vascular loop in the retinal arterial system. Since the patient was only 13 years old, we decided to perform customized peripheral pan photocoagulation to reduce the risk of neovascularization.

We reported the first case of CRA occlusion due to PVL thrombosis. Although most cases are asymptomatic, we must be aware of the possibility of finding this anomaly on routine examination owing to the risk of severe complications. Surveillance of these patients is recommended by maintaining long-term follow-up.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal patient identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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