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REVIEW

Retinal Artery Occlusion Associated with Prepapillary Arterial Loop: An Isolated Ocular Disease? Case Report and Literature Review

Ahmad M Mansour[1], Maurizio Battaglia Parodi[2], Antônio Marcelo Barbante Casella[3], Wajiha J Kheir[1], Ziad Fayez Bashshur[1]

[1]Department of Ophthalmology, American University of Beirut, Beirut, Lebanon; [2]Department of Ophthalmology, University Vita-Salute Milan, Milan, Italy; [3]Department of Ophthalmology, Universidade Estadual de Londrina, Paraná, Brazil

Correspondence: Ahmad M Mansour, Department of Ophthalmology, American University of Beirut, Beirut, Lebanon, Tel +961-337-7633, Email ammansourmd@gmail.com

Purpose: When branch retinal artery occlusion is linked to prepapillary artery loop, is a systemic work-up necessary?

Methods: Case Report and literature review. The databases searched were MEDLINE OVID, Scopus, Web of Science, and Embase. The search was for all relevant articles published from inception until October 31, 2024.

Results: A total of 33 cases of prepapillary artery loop with retinal artery occlusion were reviewed. Median age at diagnosis was 24 years (range 10–64). Twenty five cases had no systemic disease while 9 had unrelated systemic disorder. Systemic workup was carried in 17 cases and was negative. The location of the branch retinal artery occlusion was inferior in 21 cases, superior in 6 cases, and combined superior and inferior in 2 cases. Paracentral acute middle maculopathy was noted in 2 cases. Precipitating factors included exercise in 4 cases, head trauma in 2 cases, blunt ocular trauma, severe sneezing, ocular rubbing and sun exposure in 1 case each.

Conclusion: A branch retinal artery occlusion linked to a prepapillary loop appears as a distinct ocular condition in young healthy individuals. There is no definite need for additional diagnostic testing in such patients with a negative systemic history.

Keywords: prepapillary arterial loop, branch retinal artery occlusion, systemic workup

Introduction

Prepapillary arterial loop is a rare congenital vascular[1–34] anomaly originating from a branch retinal artery and ending at the optic disc with projection into the vitreous cavity. Distinguishing it from other pathologies such as optic disc collaterals or neovascularization is crucial. There is no known association between prepapillary capillary loops and specific systemic illness. This variant of normal retinal vasculature, occurs when a developing retinal artery diverges into the vitreous cavity within Cloquet's canal and returns back to the retina. It is usually unilateral in 80% of cases and the number of loops can vary from 1 to 10.[15] Prepapillary arterial loops, also known as spirals, are classified based on their morphological appearances.[15] The majority are detected on routine fundus exam and cause no symptoms. The most dreaded complication is occlusion of the branch retinal artery emanating from the loop.[1–32] Many such cases undergo extensive workup to find the cause of the vascular occlusion.[3,7,13,16,19,24–26,28,29,34] The laboratory vascular workup has included complete blood count, platelet count, erythrocyte sedimentation rate, lipid profile, antiphospholipid antibodies, and coagulation screen. In addition, imaging studies have included echocardiography and carotid Doppler ultrasonography.[35] We present such a case and wonder whether any workup is obligatory based on an extensive literature review.

Case Report

A 29-year-old Caucasian man complained of a sudden right superior visual field loss at noon time 10 days prior to presentation at a major eye facility overseas. Corrected visual acuity (from myopic astigmatism) was 20/20 in both eyes

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with a normal intraocular pressure. Funduscopy revealed sectorial whitening of the inferior temporal retina sparing the foveal region (Figures 1 and 2). Fluorescein angiography confirmed obstruction of the inferotemporal arteries sparing the macula with nonperfusion of the prepapillary arterial loop (Figure 3). The branch retinal artery occlusion spared the fovea by optic coherence tomography (Figure 4). A right superior visual field was documented (Figure 4). Extensive blood work-up (complete blood count, platelet count, fasting blood sugar level, lipid profile, coagulation profile, anti-nuclear antibody, anti-cardiolipin, lupus anticoagulant), as well as electrocardiogram, echocardiography, and carotid duplex ultrasound were unrevealing. The patient denied intravenous drug abuse. He was placed on acetyl salicylic acid and clopidogrel. A second

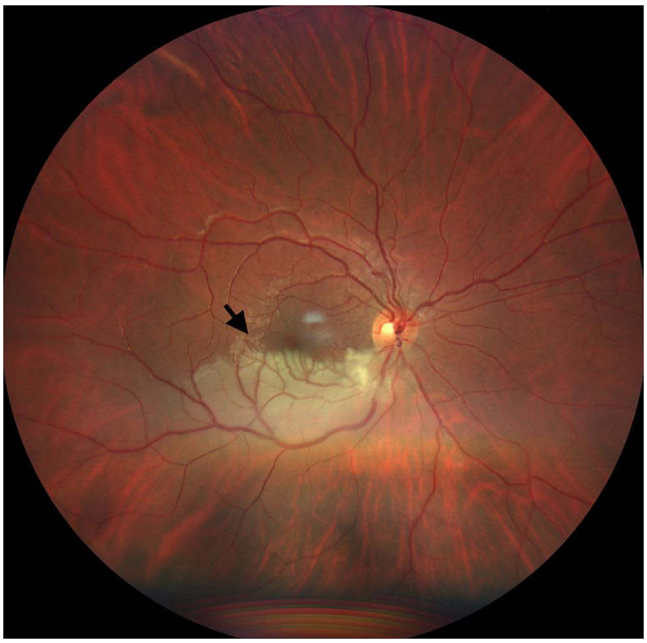

Figure 1 Widefield pseudocolor fundus of the right eye showing inferotemporal branch retinal artery occlusion (arrow).

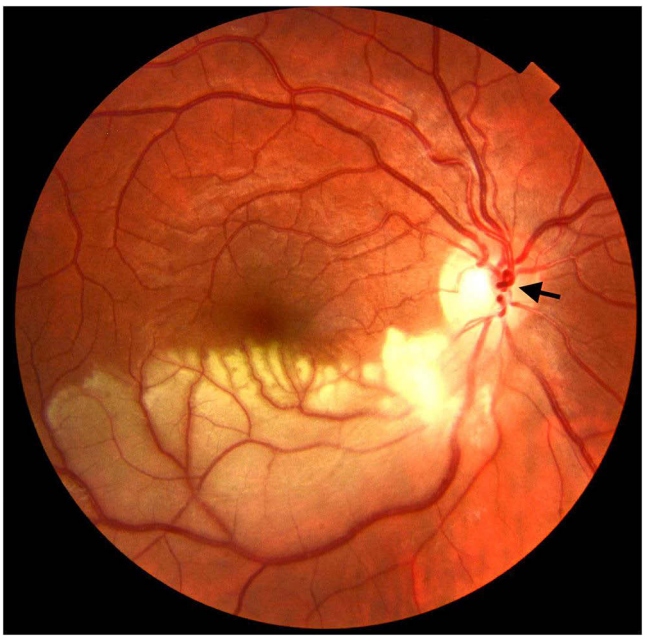

Figure 2 Prepapillary arterial loop takes the figure of eight configuration (arrow).

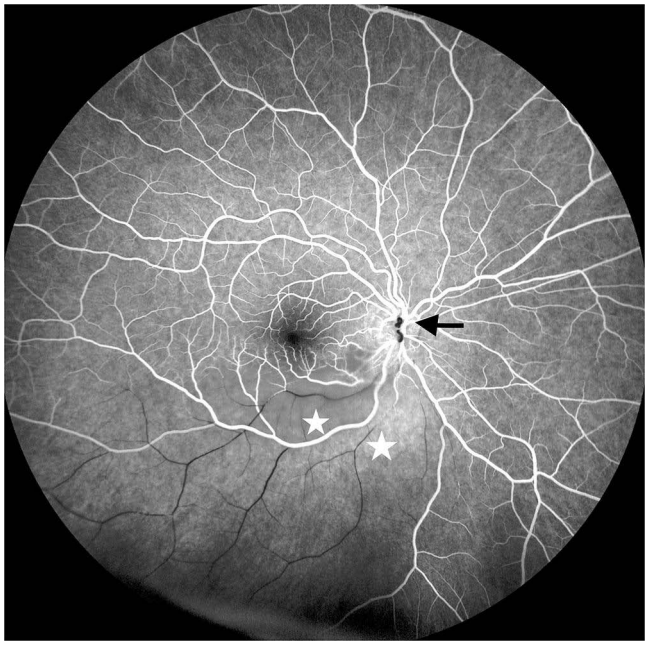

Figure 3 Venous phase of the right eye shows nonperfusion of the loop (arrow) and of the inferior temporal retina (star).

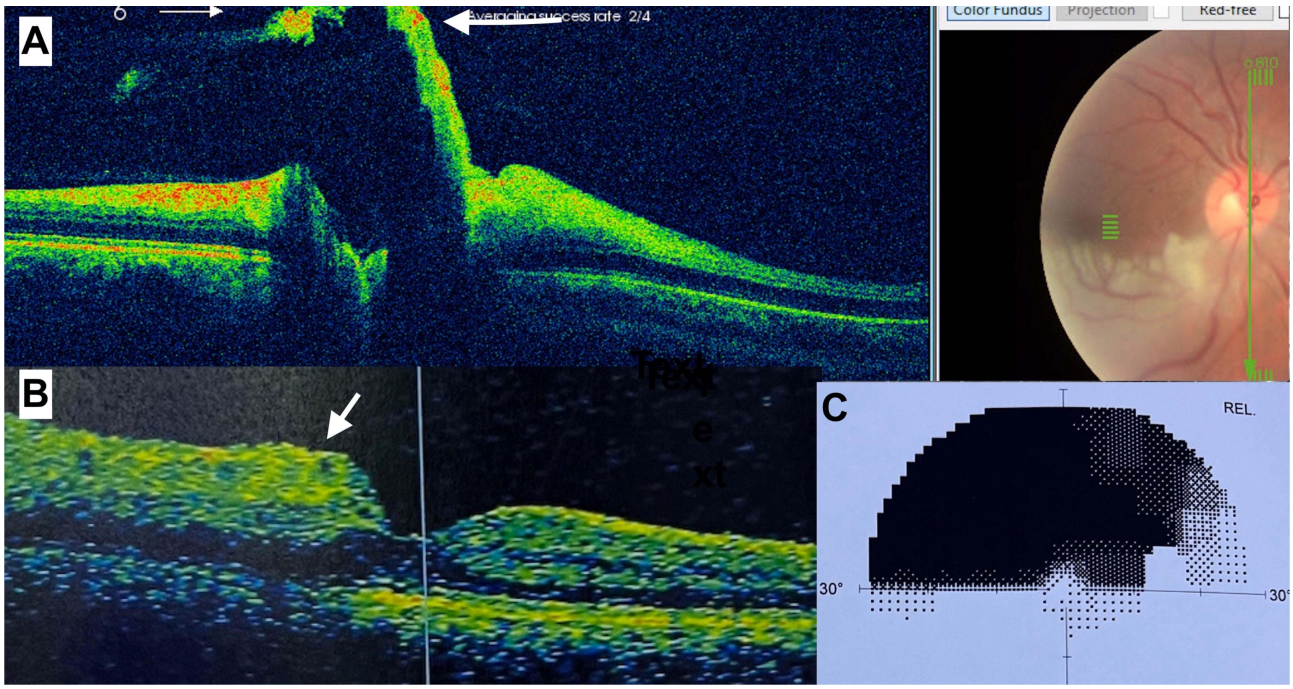

Figure 4 (**A**)- Vertical raster of the right prepapillary loop (arrow). (**B**)- Vertical raster through the fovea revealed hyperreflectivity and increased thickness of the inner retinal layers in the superior (arrow) compared to inferior retina. (**C**)- Superior visual field defect of the right eye.

opinion was sought, and a CT angiography of the brain was negative based on the differential diagnosis of arteriovenous anastomoses. A third opinion attested to the entity of branch retinal artery occlusion from prepapillary arterial loop with no need for further diagnostic tests. The visual status was unchanged at the one-month follow-up, before leaving the country.

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Literature Review

The databases searched were MEDLINE OVID, Scopus, Web of Science, and Embase. The search was for all relevant articles published from inception until October 31, 2024 following the Prisma guidelines. No limitation was placed on language, year of publication, or study design. MEDLINE was used with the following MeSH (Medical Subject Headings): prepapillary arterial loop or prepapillary vascular loop AND vascular occlusion or retinal vascular occlusion. All interventional and observational studies (case report, case series) were included. A manual search was also conducted to ensure the inclusion of potentially missing relevant articles. We searched for 1) similar articles of the finally included articles in our review, 2) reference list of finally included articles in our review, and 3) Google scholar as grey literature. The following data was collected using an Excel (Microsoft, Redmond, WA, USA) extraction sheet: baseline characteristics (year of publication, country of corresponding author, age, gender, systemic disease, possible etiology of arterial occlusion), initial best corrected visual acuity and workup for retinal vascular occlusion.

Results

A total of 33 cases (31 articles) were collected from 1901 to 2024 (including the current case report and after deleting 2 duplicates) (Tables 1 and 2). The median age was 24 (range 10–64), with equal gender distribution 15 vs15 and more involvement of the right eye (19 right vs11 left). The occlusion was inferior in 21, superior in 6, and both in 2 cases. The

Table 1 Summary of a Literature Review of Branch Retinal Artery Occlusion (BRAO) with Prepapillary Arterial Loop

Variables	Findings
Male:Female	15:15
Age	Mean=29.4 SD=16.2 Median 24 range (10–64)
Systemic disease	None 25 Yes 4 Carotid stenosis 1 Controlled hypertension 1 Controlled hypertension with diabetes mellitus 1 IgA nephropathy 1
Workup done for BRAO Yes:No	17:12
Vision	Mean 20/36 Median 20/20 range (20/15-CF) logMAR mean=0.26 SD= 0.50
BRAO right:left	19:11
BRAO superior:inferior:both	6:21:2
Loop unilateral:bilateral	25:7
Precipitating cause	Exercise 4 Head trauma 2 Ocular trauma 1 Severe sneezing 1 Ocular rubbing 1 Exposure to sunlight 1 Endodiathermy 1
Top 5 country of corresponding author	US 8 UK 5 France 4 Mexico 3 India 2

Table 2 Excel Sheet of the Detailed Literature Review of Branch Retinal Artery Occlusion (BRAO) with Prepapillary Arterial Loop

Author	Publication Year	Country	Age	Gender	Laterality BRAO	Vascular Loop	Vision	Sector BRAO	Systemic Disease	BRAO Workup	Precipitating Factor
Mansour	2025	Lebanon	27	Male	right	unilateral	20/20	inferior	No	Yes	Eye rubbing
RiaÑo	2023	Mexico	37	Female	right	unilateral	20/30	inferior	No	Yes	
Sánchez-Ramos	2022	Mexico	42	Female	right	unilateral	20/25		No	No	
Susanna	2021	Brazil	13	Female	left	unilateral	20/20	superior +inferior	No	Yes	
Mansour	2020	multinational				unilateral	Counting finger		No	No	
Esra	2020	South Africa				unilateral		superior +inferior			
Zermeno-Arce	2019	Mexico	13	Male	right	unilateral	20/20	inferior	No	Yes	
Dhawan	2015	India	54	Male	right	unilateral	20/125	superior	Hypertension, diabetes	Yes	
Rahimy	2015	US	13	Female	left	unilateral	20/25	inferior	No	No	Head rauma
Rahimy	2014	US	13	Male	right	bilateral	20/15	inferior	No	No	Head trauma
Singh	2014	Japan	10	Female	right	bilateral	20/20	inferior	No	Yes	Exercise
O'Keefe	2014	US	15	Male	left	unilateral	20/25	inferior	No	Yes	Exercise
Codenotti	2013	Italy	64	Female	right	bilateral	Vitreous hemorrhage	inferior	No	No	Endodiathermy
Ruan	2012	Australia	12	Female	right	bilateral		inferior	No	Yes	
Zekraoui	2010	Morrocco	45	Female	left	bilateral	20/400	inferior	No		
Delyfer	2008	France	22	Male	right	unilateral	20/20	inferior	No	No	
Delyfer	2008	France	38	Male	right	unilateral	20/20	inferior	No	No	
Kim	2007	Korea	26	Female	right	unilateral	Vitreous hemorrhage	superior	IgA nephropathy	Yes	
Bonneric	2007	France	16	Male	left	unilateral	20/20	inferior	No	Yes	
Rubinstein	2006	UK	57	Male	left	unilateral	20/15	superior	Carotid stenosis	Yes	
Vedantham	2005	India	16	Female	left	unilateral	20/20	superior	No	No	Severe sneezing

(*Continued*)

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Table 2 (Continued).

Author	Publication Year	Country	Age	Gender	Laterality BRAO	Vascular Loop	Vision	Sector BRAO	Systemic Disease	BRAO Workup	Precipitating Factor
Mireskandari	2001	UK	24	Male	right	bilateral	20/20	inferior	No	Yes	Exercise
Reichel	1994	US	18	Female	left	unilateral	20/20	inferior	No	Yes	
Walland	1993	UK	26	Female	right	unilateral	20/20	inferior	No	Yes	Blunt eye trauma
Regenbogen	1981	Israel	adult	Male	right	unilateral	Vitreous hemorrhage				
Amzallag	1989	France	adult	Male	right	unilateral					
Limaye	1980	US	14	Female	right	unilateral	20/20	superior	No	Yes	
Limaye	1980	US	26	Female	left	unilateral	20/15	inferior	No	Yes	
Limaye	1980	US	60	Male	left	unilateral	20/25	superior	Hypertension	Yes	
Brown	1979	US	19	Male	right	unilateral	20/20	inferior	No	No	
Harcourt	1967	UK	22	Female	left	bilateral	20/20	inferior	No	No	Exercise
Walker	1903	UK	22	Male	right	unilateral	20/20	inferior	No	No	Sun exposure
Bar	1901	Germany	adult					inferior		No	

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Table 3 Detailed Laboratory Investigations in 17 Patients with Branch Retinal Artery Occlusion (BRAO) and Prepapillary Arterial Loop Reported in the Literature

Author	Year of Publication	Age	Systemic Disease	CBC, Lipid Profile	Coagulation Screen	Hemoglobin Electrophoresis	Vasculitis Screen	Brain Imaging	Carotid Doppler	EchocardioGram or EKG Plus Cardiac Consult
Mansour	2025	27	No	Yes	Yes	No	Yes	Yes	Yes	Yes
RiaÑo	2023	37	No	Yes	Yes	No	Yes	No	No	Yes
Susanna	2021	13	No	Yes	Yes	Yes	Yes	No	Yes	Yes
Zermeno-Arce	2019	13	No	Yes	Yes	No	Yes	No	No	No
Dhawan	2015	54	Diabetes mellitus, Hypertension	Yes	Yes	No	No	No	Yes	Yes
Singh	2014	10	No	Yes	Yes	No	No	No	No	No
O'Keefe	2014	15	No	Yes	Yes	No	No	No	No	No
Ruan	2012	12	No	Yes	Yes	No	Yes	No	No	No
Kim	2007	26	IgA nephropathy	Yes	Yes	No	Yes	No	No	No
Bonneric	2007	16	No	Yes	Yes	No	Yes	Yes	No	Yes
Rubinstein	2006	57	Carotid stenosis	Yes	No	No	No	No	Yes	No
Mireskandari	2001	24	No	Yes	Yes	Yes	Yes	No	No	No
Reichel	1994	18	No	Yes	Yes	No	Yes	No	Yes	Yes
Walland	1993	26	No	Yes	No	No	Yes	No	Yes	Yes
Limaye	1980	14	No	Yes	No	Yes	No	No	No	No
Limaye	1980	26	No	Yes	No	Yes	No	No	No	No
Limaye	1980	60	Hypertension	Yes	No	Yes	No	No	No	No
		Mean 26.4	**4 (23.5%)**	**17 (100%)**	**12 (70.6%)**	**5 (29.4%)**	**10 (58.8%)**	**2 (11.1%)**	**6 (35.3%)**	**7 (41.2%)**

median presenting best corrected visual acuity was 20/20. Three cases had secondary vitreous hemorrhage and two had paracentral acute middle maculopathy. Twenty-five patients had no known systemic disease while 4 had unrelated systemic disorder. Workup for retinal vascular occlusion (complete blood count, platelet count, erythrocyte sedimentation rate, coagulation screen, carotid ultrasound, echocardiogram, and brain imaging) (Table 3) was done in 17 and turned up to be negative in all such cases while work-up was deemed unnecessary in 12. Precipitating factors included Valsalva maneuvers (strenuous exercise in 4 and severe sneezing in 1) and some kind of trauma (head trauma in 2, blunt trauma in 1 and strong ocular rubbing in 1).

Discussion

Generally, patients with branch retinal artery occlusion need an urgent stroke workup. However in the case of prepapillary arterial loop, as in both this case report and literature review attest to, there is no need for systemic workup. Occlusion in this entity is intrinsic to the loop physiology and pathology. Pathologically, prepapillary vascular loop does not have an internal elastic lamina and is supported by a connective tissue sheath lined by a cellular lamina.[36] This weakened vessel wall is hence more prone to vasoocclusion. Physiologically, it is known that twisted vessels are more prone to occlusion with several cases of twisted cerebrovascular vessels reported to lead to occlusion of blood flow.[37,38] Prepapillary loops, and more so the coil-like structures, appear to alter the hemodynamics and increase the risk of thromboembolic events due to turbulent blood flow.[15] Prepapillary loops can assume several shapes: figure of eight, corkscrew, or hairpin turn.[15] It seems that the torsional forces accompanied by turbulence lead to twisting and strangulation of the loop. Hemodynamic turbulence inside the loop results in endothelial damage with secondary intraarterial clot. Occasionally there is vitreous traction on the loop.[39] There is no known association between prepapillary capillary loops and specific systemic illness.

Conclusions

In conclusion, the presence of a prepapillary arterial loop in a young healthy patient with branch retinal artery occlusion does not necessitate a thorough and multidisciplinary approach to diagnosis and management. Detailed systemic evaluation and proactive management strategies are not crucial to optimizing patient outcomes. This entity is a local ocular problem with inherent tendency of the loop to twist occurring mostly in young subjects probably initiated by exercise, Valsalva and ocular trauma or rubbing.

Ethics Approval and Informed Consent

This case report was conducted in accordance with the declaration of Helsinki. Written informed consent was provided by the patient to have the case details and accompanying images published. Institutional approval was not required to publish the case details.

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Author Contributions

All authors made a significant contribution to the work reported, whether that is in the conception, study design, execution, acquisition of data, analysis and interpretation, or in all these areas; took part in drafting, revising or critically reviewing the article; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

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Disclosure

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