Contents lists available at ScienceDirect

American Journal of Ophthalmology Case Reports

Bilateral retinal artery occlusions with carotid artery occlusions-ocular and cerebral hemodynamic changes

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A B S T R A C T

Purpose: To introduce a case of bilateral retinal artery occlusions with carotid occlusions to achieve a fuller understanding of hemodynamic flow changes and the origin of emboli.
Observations: A 58-year-old male presented with binocular vision loss. Fundus examination revealed bilateral retinal whitening with multiple emboli. Cherry red spot was surrounded and shaped by white edematous ischemic retina. FAG showed retinal refilling but not to the macula and choroidal background. Carotid arteriography and color Doppler images demonstrated bilateral carotid occlusions. CT angiography showed compensatory flows perfused from vertebral arteries for the brain. Orbital color Doppler images revealed bilateral reversed ophthalmic flows indicating another compensatory flow arising from external carotid/ophthalmic collaterals to both eyes.
Conclusions and Importance: Reversed ophthalmic flow indicates the presence of external carotid/ophthalmic collaterals as the source of ocular blood supply and the origin of emboli. In this case, all the embolic, hemodynamic, and serotonin mechanisms may be responsible for the pathogenesis. RAO with multiple emboli is an important sign warning critical flow changes of carotid occlusions.

1. Introduction

Retinal artery occlusion (RAO) is a serious vision-threatening disorder. RAO is often associated with ipsilateral carotid artery occlusion disease (CAOD); however, ocular and cerebral hemodynamic changes have been less thoroughly described. Here, we report a case of bilateral RAO with bilateral carotid occlusions. Combined images of ocular and carotid/cerebral studies, including ophthalmic flow changes in the orbit, are demonstrated for further delineation of the profile of RAO.

2. Case report

A 58-year-old male presented with acute onset of blurred vision in both eyes for 4 days. The patient reported sudden onset of tingling pain in the left eye and corners of the mouth, followed by blurred vision of the left eye. One day later, right eye vision also became blurred. He also complained of weakness and numbness in the left limbs. Although symptoms of limbs resolved later, bilateral blurred vision persist. So he

came to our clinic for help. The patient was a nonsmoker and denied any history of systemic disease. On general physical examination, no sign of arteritis or focal neurologic deficits were noted. On ophthalmic examination, his visual acuity was hand motion in the right eye (OD) and 0.02 in the left eye (OS). Intraocular pressures were relatively low (3 mmHg OD and 7 mmHg OS). Slit-lamp examinations and extraocular muscles motility were bilaterally normal. Ophthalmoscopy revealed bilateral retinal whitening in different opacities and a cherry red spot in the macula. In both eye, there was a major embolus with multiple minor emboli with corresponding retinal opacities while some emboli dispersed in normal colored retina (Fig. 1). Fluorescein angiography (FAG) revealed refilling to the vasculature of retinal arcade but failed to the macular and temporal areas. Choroidal background of bilateral posterior pole sustained dark in color. (Fig. 1C–E).

Thrombolysis therapy under carotid arteriography was arranged immediately but discontinued due to the finding of bilateral carotid occlusions (Fig. 2A). The patient was admitted to our neurology department for further evaluation and treatment. Laboratory data

revealed only mildly elevated serum cholesterol level. Other blood data including ESR, Protein C, Protein S, Anti-cardiolipin, and bleeding tendency were normal. CT angiography (CTA) performed the next day demonstrated bilateral carotid occlusions with compensatory flows perfused from bilateral vertebral arteries posteriorly to the circle of Willis (Fig. 2B). Color Doppler images (CDI) showed severe atherosclerosis with complete occlusions of right common carotid artery (CCA) and bilateral internal carotid artery (ICA), slow flow in stenotic left CCA, and reversed flow in right external carotid artery (ECA). There were increased flows in bilateral vertebral arteries. Orbital CDI revealed reversed ophthalmic flows indicating another compensatory flow arising externally from branches of ECA (Fig. 2CD). Findings of CDI are summarized in Table 1. Aggressive treatments including carotid stenting and endarterectomy were not indicated for this patient. He took conservative medical treatment and maintained stable for six months. Unfortunately, he passed away before eight months follow-up.

3. Discussion

Embolism originated from carotid or cardiac lesion is the most common cause of RAO while marked carotid stenosis or occlusion will reduce the ocular blood flow and produce hemodynamically induced retinal ischemia.[1] Most hemodynamically significant CAOD's are unilateral with decreased perfusion to the brain transiently so that there will be compensatory flow response, arising from the contralateral carotid artery and/or the vertebral arteries for the redistribution of intracranial circulation via the circle of Willis. In cases with poor function of Willisian collaterals, such as bilateral CAOD, inadequate intracranial circulations for the brain or to the eye may lead to the recruitment of external carotid/ophthalmic collaterals.[2,3] The retina can tolerate transient ischemia but suffers irreversible damage after prolonged occlusion.[4,5] Thus, delayed or insufficient compensatory flow from the collaterals may aggravate the retinal ischemia. Prolonged duration of retinal ischemia may result in necrosis of the ganglion cells with intracellular edema which accounts for the clinical gray retinal opacity.[6,7] At first glance, this case present with typical pictures of central RAO, but we noticed that there are regions of more intense whitening at the boundaries of the ischemic retina and many emboli with corresponding retinal opacity while some emboli without; and

more, there is derangement of the cherry-red spot contour in the left eye (Fig. 1). The variation in retinal opacities and in shapes of cherry-red spots could be the result of multiple retinal infarctions by emboli.

FAG of this case revealed refilling to the retinal vasculature with filling failure to the macula similar to the transient RAO as described by Hayreh.[2] It is noticeable that bilateral posterior poles sustained dark which is seldom observed in RAO cases clinically. These dark areas are due to retinal non-perfusion of embolic infarctions or blockage from the overlying opacified ischemic retina (Fig. 1C ~ E). However, some dark areas may result from deficits of the choroidal background because of the simultaneous perfusion failure of posterior ciliary arteries (PCA). In addition to embolic and hemodynamic mechanisms, the third mechanism, arterial spasm induced by serotonin during carotid occlusive event,[1] may act as an autoregulation of CAOD and is responsible for the PCA occlusions. With sympathetic innervation, PCA is more prone to be affected by serotonin than intraocular arteries. This will ensure adequate blood perfusion for the retinal circulations to preserve visual function during critical carotid occlusions.

In this case, marked hemodynamic changes were well demonstrated by carotid and cerebral evaluations performed soon after RAO attack. Carotid arteriography, CDI and CTA all demonstrated bilateral carotid occlusions with many perfusion changes in different aspect (Fig. 2). CTA revealed vanished carotid flows with fair intracranial compensatory flow only from the posterior vertebral arteries to the anterior cerebral territories (Fig. 2B), also confirmed by CDI with relatively high vertebral flow bilaterally. Since there was no neurologic deficit except RAO at presentation, vertebral compensatory flow appears to be adequate for the brain but insufficient to the eye, as there were reversed ophthalmic flows noted by orbital CDI indicating the recruitment of external carotid/ophthalmic collaterals (Fig. 2C). Slower ophthalmic flow velocity in deeper orbit tallied with external compensatory flow (Table 1). The major source of blood supply to bilateral ophthalmic arteries is from the branches of left ECA in this case. Deep in the right orbit, slow reversed flow with faint antegrade signals may indicate a head-on collision between the vertebral and external compensatory flows (Fig. 2D). Fig. 3 demonstrated these hemodynamic flow changes according to above evaluations.

Since the major sources of blood supply to both eyes are compensatory flows from an external carotid/ophthalmic anastomosis, there is
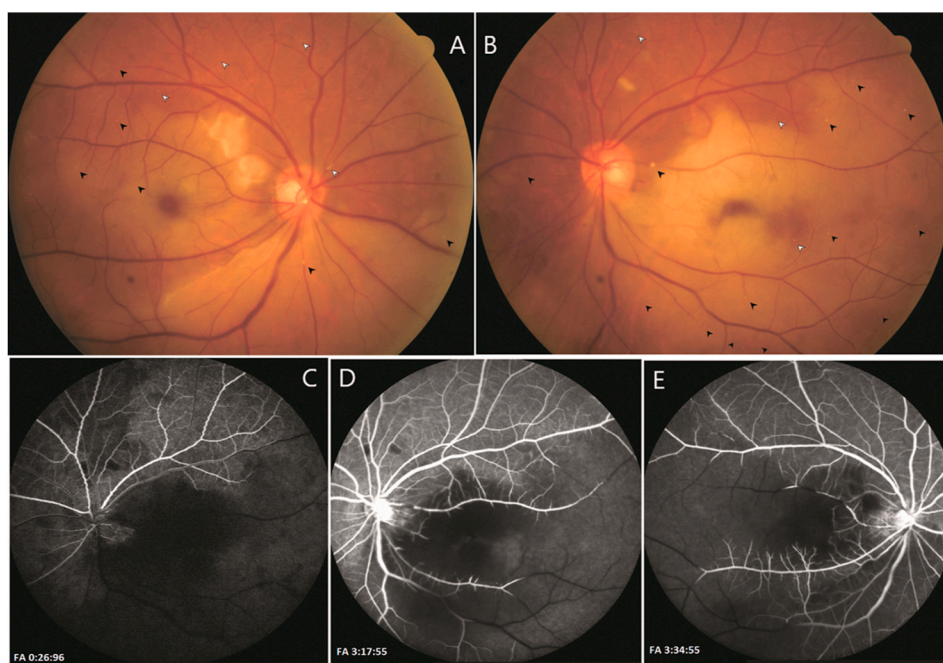

Fig. 1. Bilateral RAO at presentation. A. Right eye. There is a major embolus impacted on the optic disc with numerous micro-emboli with corresponding retinal opacities (arrowhead). Cherry-red spot is round. There are regions of more intense opacities at the boundaries of some ischemic retina. B. Left eye, a major embolus is impacted in the macular branch. Most emboli have corresponding retinal opacities (arrowhead) while some emboli dispersed in normal colored retina (white arrowhead). There is derangement of the cherry-red spot contour. C. FAG of left eye during retinal arterial phase (at 26 seconds) shows refilling to the superior retinal vasculature bed with absence of filling in the macular region and inferior retina. There are deficits of choroidal background. Choroidal defect of superior part soon refill via the anastomosis but it persist under the posterior pole. D, E. Three minutes after dye injection, FAG shows normal filling of the retinal vasculature bed with absent filling in the macular region and temporal retina. Bilateral background of the posterior pole sustain dark in color. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

an enlargement of the anastomosis and flow direction changes which may facilitate debris scaling off the vessel wall and become emboli in the eye. We believe in the present case most retinal emboli originated from external carotid/ophthalmic collaterals. It has been reported by vascular surgeons that with distal embolic protection device to occlude distal ICA temporarily during carotid angioplasty and stenting to prevent embolic stroke, RAO and retinal embolization could still occurr.[8,9] The passage of debris through the collaterals poses a significant risk as flushing debris through the orbital branch of the ECA to ophthalmic artery may lead to retinal embolization. Transient blindness in patients with a fully occluded ipsilateral ICA unequivocally indicating the collateral pathway to be a mechanism for retinal embolization.[8,9]

Bilateral RAO is reported in only 1–2% of central RAO,[10,11] and in 20% of arteritic RAO cases.[1] When both eyes are affected simultaneously by RAO, valvular heart disease and vasculitis including giant cell arteritis, Henoch-Schonlein purpura, Churg-Strauss syndrome, and Wegener granulomatosis should be considered.[12–15] Arteritic RAO is potentially treatable with better systemic and visual outcome.

Most RAO patients had ipsilateral carotid lesions, however, retinal embolus and type of RAO were considered a poor predictor of hemodynamically significant CAOD.[1,16,17.] Our recent report revealed that RAO with multiple emboli can be an important sign warning hemodynamic changes of severe CAOD.[18] In recent years, more timely clinical information and evidences of carotid and cerebral evaluations can be collected and surveyed to facilitate the understanding of disease process

Table 1
Findings of Doppler ultrasonography.

	Right	Left
Carotid artery		
Internal	Total occlusion. No flow	Total occlusion. No flow
External	Reversed slow flow	Antegrade flow
Common	Total occlusion, No flow	Stenosis: 63%, slow flow
Vertebral artery	Increased flow	Increased flow
Ophthalmic artery	Reversed slow flow	Reversed flow
Mean Flow velocity		
Depth 3.0–3.9cm	11.0 cm/s	35.5 cm/s
Depth 4.0–4.5cm	5.35 cm/s	25.9 cm/s

and relationships between RAO and CAOD. Factors that influence the retinal ischemia including hypo-perfusion by the carotid occlusion, duration to re-establish the intracranial and orbital circulations, adequacy of the compensatory flow, pathway for the collaterals, vasospasm of ophthalmic territories by serotonin, impaction of the major embolus, and retinal infarctions by the micro-emboli were all displayed in this case. These findings provide valuable insights about the relationships between RAO and CAOD.

4. Conclusion

In this case of RAO with multiple emboli, cherry red spot is surrounded and shaped by white edematous ischemic retina. Reversed
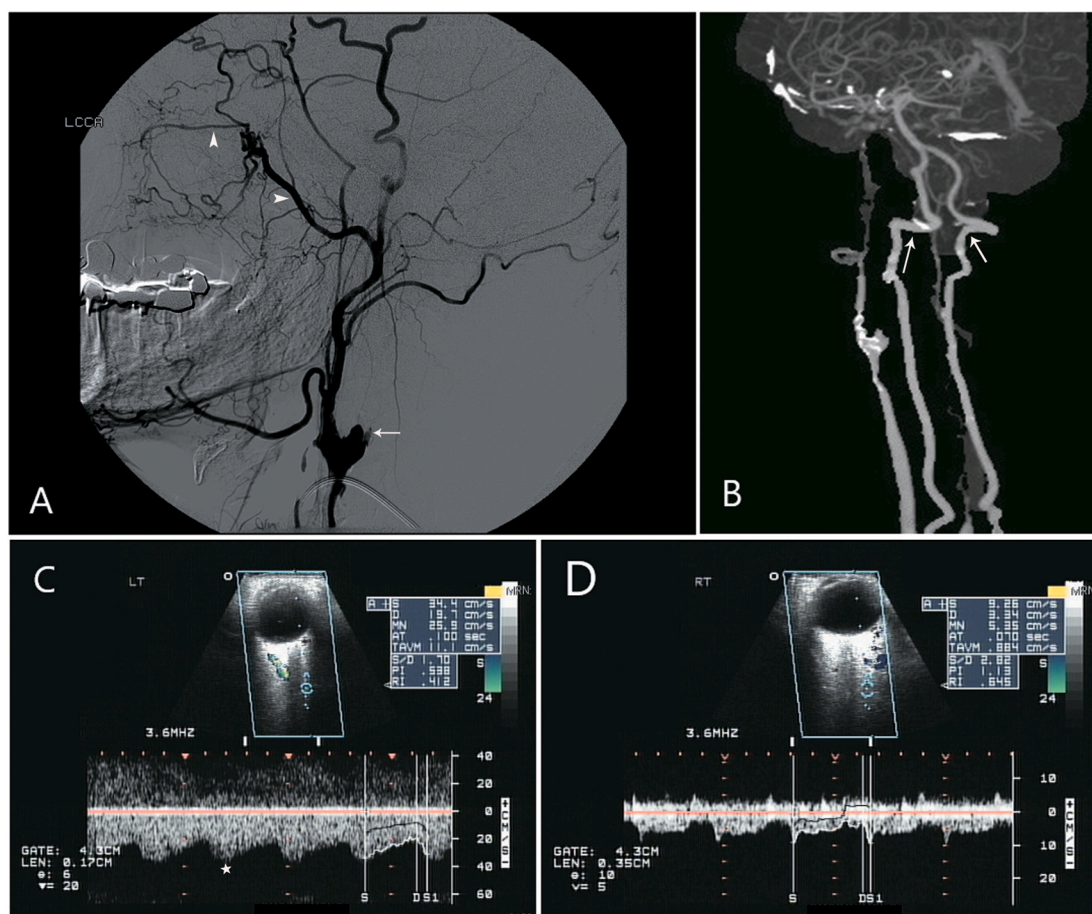

Fig. 2. Hemodynamic changes demonstrated with ocular and carotid/cerebral evaluations. A. Carotid arteriography of left CCA reveals completely occluded ICA. (Arrow) There is blood supplied from left ECA to the orbital area. (Arrowhead) B. CT angiography shows total occlusion of bilateral ICA without carotid flow for the brain. There are compensatory flows from bilateral vertebral arteries posteriorly perfusing to anterior cerebral territories. (arrow) C. Color Doppler images in left orbit at 4.3cm depth shows reversed ophthalmic flows parallel with the optic nerve. (Star) D. Color Doppler images in right orbit at 4.3cm depth also shows reversed flows in relatively slow velocity. There are faint signals of antegrade flow. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)
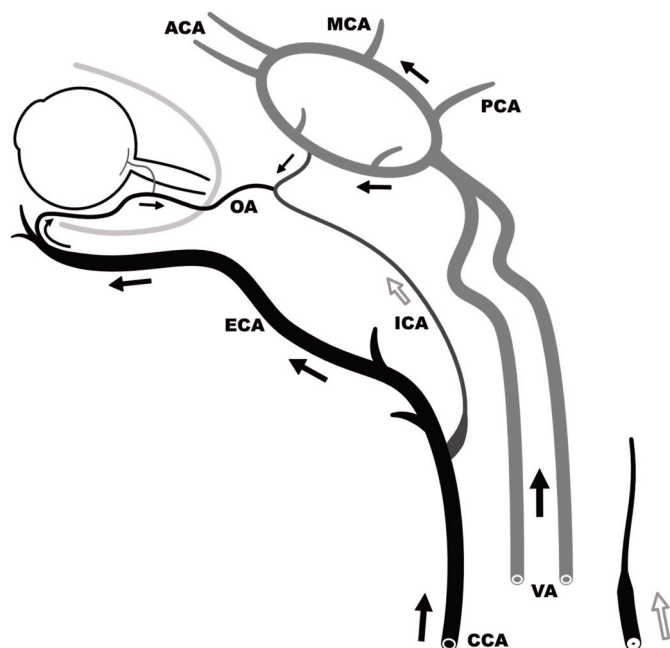
Fig. 3. Schematic diagram illustrates the collateral pathways in the status of bilateral carotid occlusion. There are compensatory flows from the vertebral arteries for the brain but may not be sufficient to the eye. The source of ocular blood supply is then from the branch of left ECA and the ophthalmic flow become retrograde in the orbit. (Abbreviations: ICA, internal carotid artery; ECA, external carotid artery; ACA, anterior cerebral artery; MCA, middle cerebral artery; VA, vertebral artery; OA, ophthalmic artery.)

ophthalmic flow found by orbital CDI indicates the presence of external carotid/ophthalmic collaterals as the source of ocular blood supply and the origin of emboli. Complex mechanisms including hemodynamic, embolic, and serotonin may contribute all together for the pathogenesis. Such patient should have thorough ocular and carotid/cerebral evaluations, including orbital CDI.

Funding

No funding.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

Declaration of competing interest

All authors declare that there are no financial or other conflicts of interest.

1. Conflicts of interest

Potential conflict of interest exists:
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We wish to draw the attention of the Editor to the following facts, which may be considered as potential conflicts of interest, and to significant financial contributions to this work:
The nature of potential conflict of interest is described below:
V No conflict of interest exists.
We wish to confirm that there are no known conflicts of interest associated with this publication and there has been no significant financial support for this work that could have influenced its outcome.

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