Carotid artery disease and central retinal artery occlusion

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Central retinal artery occlusion (CRAO) is a devastating ocular event with a dismal visual prognosis.[1] The visual crisis caused by CRAO may be the first manifestation of an underlying disease process such as atherosclerosis of the carotid artery. A number of studies based on clinical, nonangiographic methods have estimated the incidence of ipsilateral carotid artery disease to be 8% to 25% of those affected with CRAO.[2-4] In contrast, one roentgenographic study demonstrated that nine of nine patients with CRAO had angiographically demonstrable ipsilateral carotid artery disease.[5]

Because of the great disparity between the coincidence of carotid artery disease and CRAO in clinical and angiographic studies, we reviewed our cases of CRAO with special attention to the incidence of angiographically abnormal carotid arteries. Our results confirm a high association between carotid artery disease and CRAO. Three cases of CRAO with normal carotid angiograms are reported to emphasize that the association between these two events is significant, and clearly less than 100%.

Patients

Thirty cases of CRAO occurring in 27 patients

were seen at The Cleveland Clinic Foundation between 1969 and 1978. The age range of the patients, 17 men (63%) and 10 women (37%) was from 21 to 75 years (mean, 57 years).

Men were most often affected in their 60s and women in their 70s (*Table 1*). The right eye was affected in 14 patients (52%), the left in ten patients (37%), and three patients had bilateral CRAOs (11%).

Results

Associated diseases. The associated diseases in patients with CRAO are shown in *Table 2.* Most patients had hypertension and atherosclerosis. Other diseases included vasculitis, cardiac valvular disease, trauma, and meningioma.

Procedure-related CRAO. Approximately one fourth of our cases of CRAO occurred after a diagnostic or surgical procedure (*Table 3*). The greatest number of cases occurred after coronary bypass surgery.* CRAO also followed carotid angiography, carotid endarterectomy, carotid ligation for carotid-cavernous sinus fistula, and cervical spine decompression.

Carotid angiography and CRAO. Carotid angiography was performed in 13 patients who had CRAO. Angiography revealed ulcerated or stenosed carotid arteries on the same side as CRAO in ten patients (76%). However, three patients had normal carotid angiograms.

Case 1. A 74-year-old man suffered a CRAO OD. The medical history documented diffuse atherosclerotic disease, chemical diabetes, hyperlipidemia, and hyperuricemia. Bilateral carotid angiograms were normal.

Case 2. A 67-year-old man had presenting symptoms of CRAO OD. The medical history documented hyperuricemia. Results of the following studies were normal: right carotid angiogram, echocardiogram, temporal artery biopsy, and carotid compression tonography. The admission electrocardiogram showed atrial fibrillation, but a HIS bundle electrocardiogram was normal.

Case 3. A 75-year-old woman was examined in the Department of Neurosurgery for evaluation of dementia. She had suffered attacks of blurred vision in the left eye for

Table 1. Central retinal artery occlusion

Age, yr	Men	Women	Total
20–29	1	0	1
30–39	1	2	3
40–49	1	2	3
50–59	3	1	4
60–69	10	1	11
70–79	1	4	5

Table 2. Associated diseases in CRAO

Disease	No. of cases
Hypertension and/or atherosclerosis	18 (67%)
Vasculitis	3 (11%)
Cardiac valvular disease	2 (7%)
Trauma	2 (7%)
Meningioma	1 (4%)
None	1 (4%)

Table 3. Procedure-related cases of CRAO

Procedure	No. of cases
Postcoronary bypass	3
Postcarotid angiography	1
Postcarotid endarterectomy	1
Postcarotid ligation for carotid-cavernous sinus fistula	1
Postcervical spine decompression	1
Total	7

* *The total number of coronary revascularization procedures performed at the Cleveland Clinic during the course of our study approximates 18,500. From 1974 through 1977, 1761 carotid angiograms were performed. At present, 250 to 300 carotid endarterectomies are done yearly. Thus, the incidence of CRAO following these procedures is extremely small.*

one year and then CRAO OS developed one month before her admission. Carotid angiography showed no abnormality of the common or internal carotid arteries proximal to the left ophthalmic artery. However, angiography did reveal a meningioma of the falx cerebri fed by the left middle meningeal artery and by the left anterior cerebral artery.

Neurological symptoms and CRAO. Nine patients (33%) had neurological symptoms prior to the onset of CRAO. These included stroke, vertigo, amaurosis fugax, numbness, weakness, dizziness, blurred vision, headache, diplopia, and aphasia. Fourteen patients (52%) had no prior neurological symptoms. A complete history was unavailable in four cases (15%).

Discussion

Much of our data agree with other reports of CRAO. For example, we found that men in their 60s were affected most often by CRAO, whereas women were most often affected in their 70s. A similar observation was made by Liversedge and Smith.[2] In addition, most of our patients were men, and the right and left eyes were affected almost equally in both sexes. These findings also agree with previous reports of CRAO.[1-4]

Approximately two thirds of our patients had hypertensive or atherosclerotic cardiovascular disease (*Table 2*). In other series the association between these two entities and CRAO has ranged from 30% to 77%.[1-3]

Vasculitis was an associated finding in three of our patients (11%). Two were younger than 40 years, and had diffuse nonspecific vasculitis that resulted in multiple neurological deficits. The third patient had a malignant form of biopsy-proved temporal arteritis that precipitated bilateral CRAOs and a left hemiparesis. The incidence of vasculitis in our series is somewhat higher than in other series reported in which the incidence has been 3% to 6%.[1,3]

Cardiac vascular disease, including atrial myxoma, has been associated with CRAO in 2% to 17% of reported cases.[1-3] In our series, the association was 7%. In these cases of CRAO, embolization of rheumatic vegetations or myxoma tissue to the central retinal artery is thought to be the cause.[6-8]

About one fourth of our cases of CRAO were temporally associated with surgery or carotid angiography. Three cases followed coronary revascularization operations. The interval between surgery and CRAO was 6, 10, and 21 days respectively. In two of these patients retinal emboli were noted. Williams[9] has shown that retinal emboli after open heart surgery consist of platelets and other blood cell aggregates, as well as fat and silicone material derived from the extracorporeal oxygenation system. One case of CRAO occurred immediately after carotid angiography. Because embolic material was identified on the optic disc, embolism of atheromatous material dislodged during the procedure most likely precipitated the event. Another case followed carotid endarterectomy during which embolic atheromatous material may have been dislodged. Similar observations have been made by others.[10-12] In addition, one case of CRAO occurred after ligation of the internal carotid artery for treatment of a carotid-cavernous sinus fistula, and another occurred after a cervical spine decompression operation. The cause of the CRAO in this last case is only speculation, but may have been related to inadvertent long-standing pressure on the globe during surgery.[13,14]

Carotid angiography was performed

in 13 patients with CRAO as noted before. Ten patients had identifiable lesions in the ipsilateral carotid artery for an incidence of 76%. The degree of occlusion ranged from 40% to 60% stenosis (four cases) to greater than 95% stenosis (six cases). These results are notable in several respects. First, to our knowledge our series represents the largest number of patients who have had carotid angiography following CRAO. Second, our findings corroborate a high association between CRAO and ipsilateral carotid disease as first shown by Kollarits et al.[5] That this association is clearly less than 100% is important. Furthermore, the three cases in which carotid angiograms were normal emphasize again that pathological mechanisms not obviously related to carotid disease may be associated with CRAO in elderly patients.

The first patient with presenting symptoms of CRAO and normal carotid angiograms had many factors predisposing him to atherosclerotic disease, including hyperlipidemia, hyperuricemia, and chemical diabetes. In view of the normal angiograms, and no obvious source of emboli, the most likely cause was thrombotic occlusion of the central retinal artery.

The second case of CRAO with normal carotid angiograms occurred in an apparently healthy elderly patient. Appen et al[3] noted that 16% of their patients had no associated disease at the time of CRAO. However, carotid angiograms were not performed.

Our third case of CRAO with normal carotid angiograms is of special interest. This patient had visual symptoms for almost a year preceding a CRAO in the left eye. However, angiography did reveal a meningioma of the falx, which was fed by the left middle meningeal artery and the left anterior cerebral artery. It is worth noting that the left anterior cerebral artery is a branch of the left internal carotid artery, and that the left middle meningeal artery is a branch of the left external carotid artery. Furthermore, an anastomosis between the internal carotid system and the external carotid system usually occurs via the lacrimal artery, a branch of the ophthalmic and the middle meningeal arteries.[15] We suggest that this patient's visual symptoms and CRAO may have resulted from a steal syndrome in which blood was diverted from the central retinal artery to the tumor via the anastomosis from the lacrimal to the middle meningeal artery. Although CRAO has been associated with carcinoma of the colon, cecum, and bronchus,[4] to our knowledge this is the first reported case of CRAO associated with an intracranial meningioma.

Most of our patients had no neurological symptoms prior to CRAO. Only 33% of cases had symptoms and in Liversedge and Smith's[2] series the incidence was 21%. Thus, CRAO may be the first sign of systemic vascular disease that can increase mortality and morbidity.

Summary

Thirty cases of CRAO occurring in 27 patients are reviewed. About two-thirds of the patients had hypertension or atherosclerosis or both as associated systemic findings. Carotid angiography was performed in 13 patients with CRAO and significant ipsilateral carotid artery disease was observed in 10 (76%). In approximately 25% of cases, CRAO was related to a surgical or invasive diagnostic procedure. The majority of patients in our series had no neurological symptoms prior to CRAO. This study, which reports the largest

series of carotid angiograms in patients with CRAO, emphasizes and confirms a significant correlation between CRAO and ipsilateral carotid artery disease.

References

1. Karjalainen K: Occlusion of the central retinal artery and retinal branch arterioles; a clinical, tonographic and fluorescein angiographic study of 175 patients. Acta Ophthalmol [Suppl] (Kbh) **109:** 1–95, 1971.
2. Liversedge LA, Smith VH: Neuromedical and ophthalmic aspects of central retinal artery occlusion. Trans Ophthalmol Soc UK **82:** 571–588, 1962.
3. Appen RE, Wray SH, Cogan DG: Central retinal artery occlusion. Am J Ophthalmol **79:** 374–381, 1975.
4. Ellis CJ, Hamer DB, Hunt RW, et al: Medical investigation of retinal vascular occlusion. Br Med J **2:** 1093–1098, 1964.
5. Kollarits CR, Lubow M, Hissong SL: Retinal strokes. I. Incidence of carotid atheromata. JAMA **222:** 1273–1275, 1972.
6. Penner R, Font RL: Retinal embolism from calcified vegetations of aortic valve; spontaneous complication of rheumatic heart disease. Arch Ophthalmol **81:** 565–568, 1969.
7. Jampol LM, Wong AS, Albert DM: Atrial myxoma and central retinal artery occlusion. Am J Ophthalmol **75:** 242–249, 1973.
8. Anderson JD, Lubow M: Atrial myxoma as a source of retinal embolization. Am J Ophthalmol **76:** 769–772, 1973.
9. Williams IM: Retinal vascular occlusions in open heart surgery. Br J Ophthalmol **59:** 81–91, 1975.
10. Cogan DG, Kuwabara T, Moser H: Fat emboli in the retina following angiography. Arch Ophthalmol **71:** 308–313, 1964.
11. Haney WP, Preston RE: Ocular complications of carotid arteriography in carotid occlusive disease; a report of three cases. Arch Ophthalmol **67:** 127–137, 1962.
12. Shillito J Jr, Rockett FX: Retinal artery embolism; a complication of carotid endarterectomy. J Neurosurg **20:** 718–720, 1963.
13. Paton D, Goldberg MF: Management of Ocular Injuries. Philadelphia, Saunders, 1976, p 225.
14. Jampol LM, Goldbaum M, Rosenberg M, et al: Ischemia of ciliary arterial circulation from ocular compression. Arch Ophthalmol **93:** 1311–1317, 1975.
15. Grant JCB: An Atlas of Anatomy, 6th ed. Baltimore, Williams and Wilkins, 1972, p 524.