SIMULATANEOUS OCCLUSION OF THE CENTRAL RETINAL ARTERY AND VEIN*

BY *Richard D. Richards,* MD

THE CHARACTERISTIC APPEARANCE OF OCCLUSION OF THE CENTRAL RETINAL ARTERY is well known. The clinical appearance of occlusion of the central retinal vein is more variable but equally well recognized. Whether retinal ischemia plays any part is in question, although observers have noted that retinal artery disease is often present in combination with central retinal vein occlusion. Occlusion of both the central retinal artery and vein in one eye at the same time is unusual despite their close relationship in the optic nerve. Cases have been described in the literature, but the clinical characteristics have not been clearly defined and emphasized.

We have observed six patients with acute combined central retinal artery and vein occlusion, with characteristic clinical aspects.

CASE REPORTS

CASE 1

A 25-year-old woman, developed blurred vision in her left eye which progressed to total blindness in 24 hours. She also had retrobulbar pain, headache, fever and leucocytosis of 13,000. Neurological examination, skull roentgenograms and lumbar puncture were within normal limits. The ophthalmoscopic examination of the left eye revealed papilledema with hemorrhages around the disc and a grey-white retina with a cherry-red spot (Fig 1 & 2). Fluorescein angiography showed occlusion of both central retinal vessels. She eventually developed neovascular glaucoma and the left eye was enucleated.

Histologic examination of the eye, removed 18 months after loss of vision, showed obliteration of the angle from anterior synechias and a neovascular membrane extending along the surface of the iris. The retina was atrophic. The optic nerve showed complete atrophy and a diffuse interstitial lymphocytic infiltration.

During the time between the initial loss of vision and enucleation, and subsequently as well, the patient was hospitalized several times for extensive evaluation. Aphthous ulcers developed on the mucous membranes, and neurological signs

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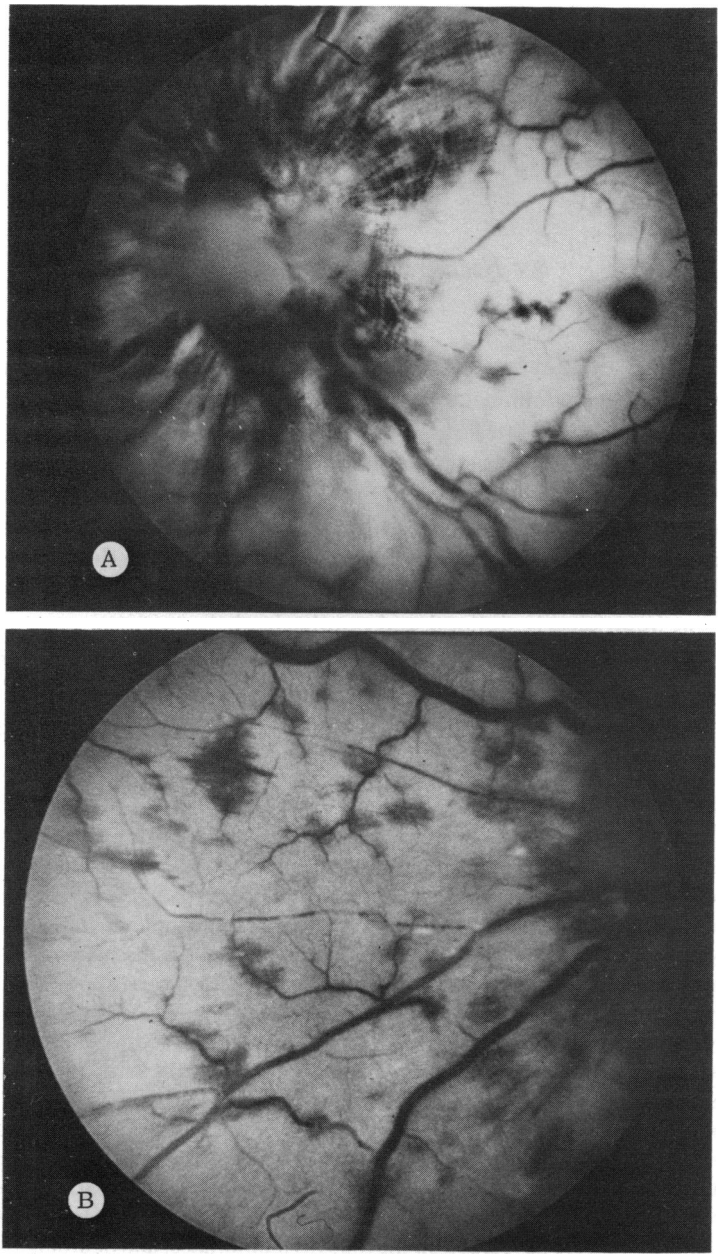

FIGURE 1

Case 1. A: Papilledema, hemorrhages adjacent to disc and milky-white retina caused by combined CRAO/CRVO. B: Nasal to disc. Hemorrhages are adjacent to retinal vessels but are mainly near the disc. Retina shows cloudy swelling.

and symptoms became evident. A diagnosis of Behcet's disease was finally substantiated.

CASE 2

A 48-year-old woman, noted blurred vision in the left eye for two days, progressing to counting fingers at six inches and finally to no light perception. There also was increased lacrimation and pain over the left eye and frontal area. The ophthalmoscopic appearance of the left eye was papilledema with a few hemorrhages around the disc and macular area, and a grey-white retina with a cherry-red spot in the macula (Fig 3). Neurological examination, skull roentgenograms and lumbar puncture showed no significant abnormalities. Final diagnosis was vasculitis of unknown etiology.

CASE 3

A 39-year-old man, had fever and chills with marked swelling of the right orbital area. Vision decreased to no light perception in the right eye in 48 hours. Skull

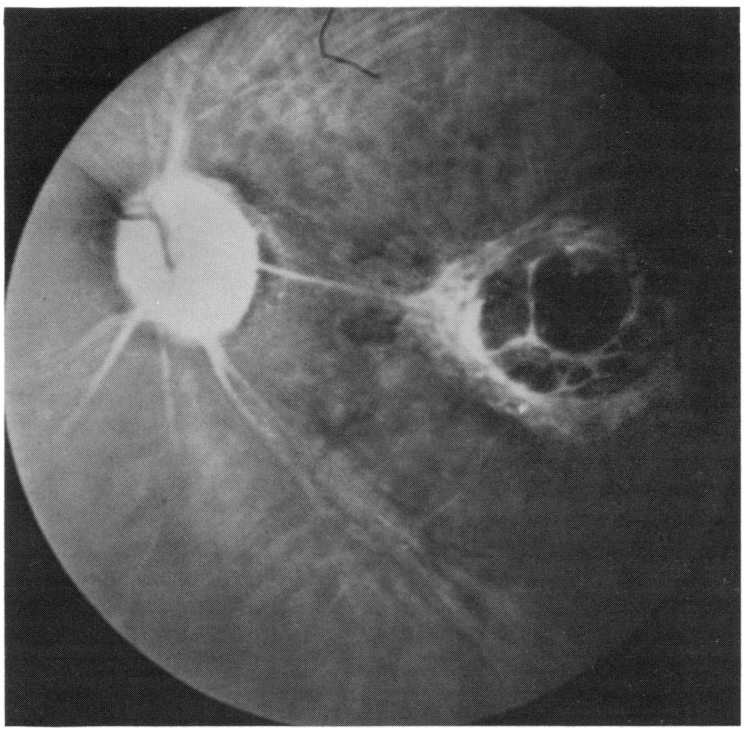

FIGURE 2

Case 1. Two months later, the disc is white, and all retinal vessels are obliterated except for a superior nasal vein. The macula shows cystic degeneration.

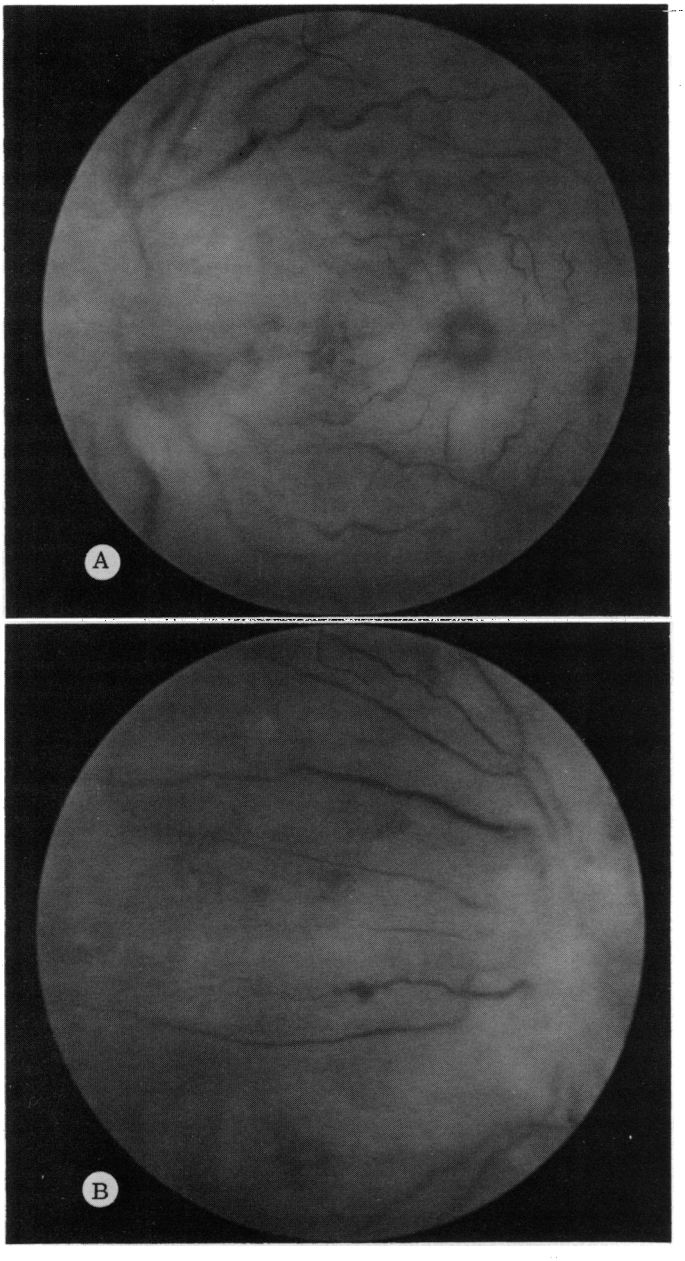

FIGURE 3

Case 2. A: Papilledema with a few hemorrhages around the macular area, and milky-white retina. B: Nasal to the disc. Scattered retinal hemorrhages and a milky-white retina.

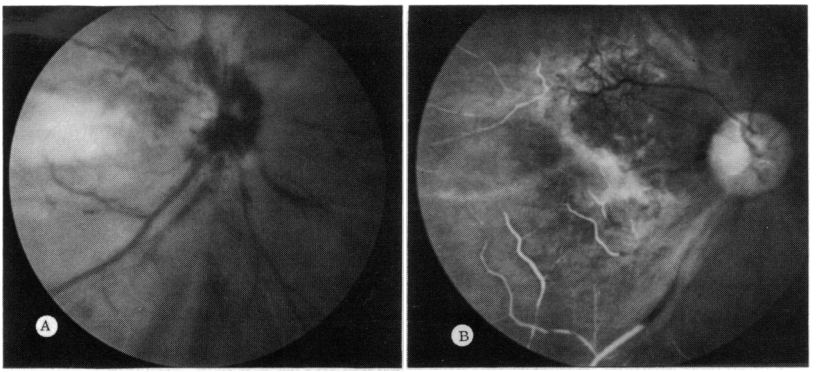

FIGURE 4
Case 3. A: Mild papilledema, hemorrhages over the disc edges, and a milky-white retina. White material in the peripheral part of the retinal vessels did not move. B: Two months later, the disc is white, and two upper branch veins are patent. Neovascularization is present above the macula. White material in the retinal vessels is more prominent. Macula shows cystic degeneration.

roentgenograms showed bilateral maxillary and ethmoid sinusitis. Ophthalmoscopy showed slight papilledema with hemorrhages at the disc and a grey-white retina (Fig 4A). The retinal blood vessels, both arteries and veins, were filled in a segmented fashion with blood cells and white plasma. No movement of the plasma and blood column was seen, indicating total occlusion of the central retinal artery and vein. Optic atrophy and neovascularization around the disc subsequently developed (Fig 4B), and later neovascular glaucoma occurred. Final diagnosis was septic cavernous sinus thrombosis.

CASE 4

A 52-year-old man, developed fever, weight loss, and had a heart murmur. At the time of admission, blood cultures were positive for streptococcus viridans. Ocular examination showed no light perception in the right eye for an unknown period of time. Ophthalmoscopy showed a pale disc with mild papilledema and retinal hemorrhages around the disc and macula, and a grey-white retina with a cherry-red spot (Fig 5). The patient died shortly thereafter with a final diagnosis of subacute bacterial endocarditis. (Cases 1, 3, and 4 were previously reported.)[1]

CASE 5

A 51-year-old man, noted sudden onset of blurred vision, left eye, with pain behind and around the eye. Vision was reduced to 20/200, with the greatest decrease in the lower half of the visual field. He was also being treated for poorly differentiated lymphocytic lymphoma and lymphomatous lepto-meningitis. A Marcus Gunn pupil was present, and ophthalmoscopy of both eyes revealed normal fundi. Despite treatment with systemic corticosteroids, vision gradually decreased to counting

fingers at one foot after three weeks. Ten days later he developed increased pain behind the left eye, and vision was reduced to no light perception. EMI scan of the orbit showed no abnormalities. Ophthalmoscopy showed papilledema with many hemorrhages, and a grey-white retina with a cherry-red spot (Fig 6). Fluorescein angiography showed total occlusion of the central retinal artery and vein, with a normal choroidal vascular pattern (Fig 7).

He was treated with 3000 rad to the left orbital apex, with no observable effect. Two months later, EMI scan showed a positive area in the left ventricular horn. Final diagnosis was poorly differentiated lymphocytic lymphoma with central nervous system involvement.

CASE 6

A 38-year-old woman, had sudden loss of vision in the left eye, one week duration. She was also being treated for acute lymphocytic leukemia. Vision of the left eye was light perception only. Ophthalmoscopy showed slight papilledema with massive pre-retinal and retinal hemorrhages around the disc, with a grey-white retina and a cherry-red spot, and focal narrowing of retinal arteries and veins (Fig 8). The final diagnosis was acute lymphocytic leukemia.

Our patients all had similar histories of loss of vision in one eye, usually

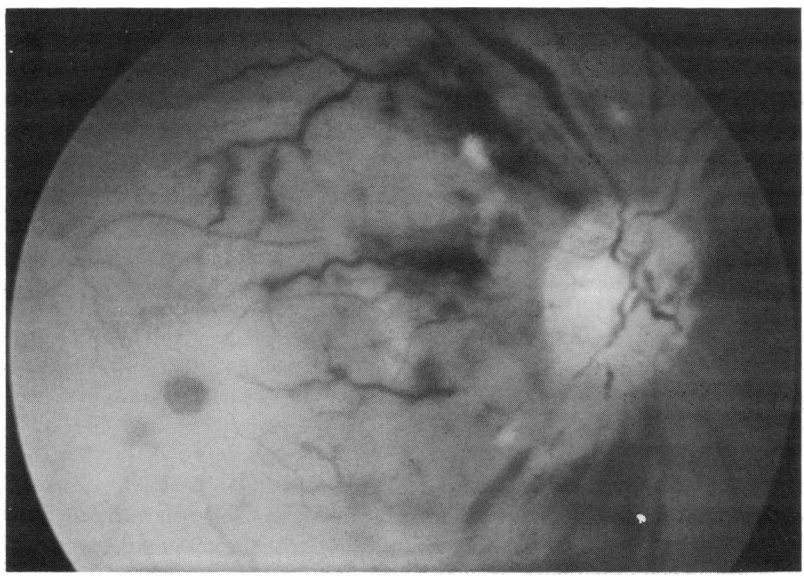

FIGURE 5

Case 4. Slight blurring of the disc margins with narrowed vessels. Retinal veins are engorged away from the disc. Hemorrhages are adjacent to the engorged retinal veins. Retina is milky-white with a cherry-red macula.

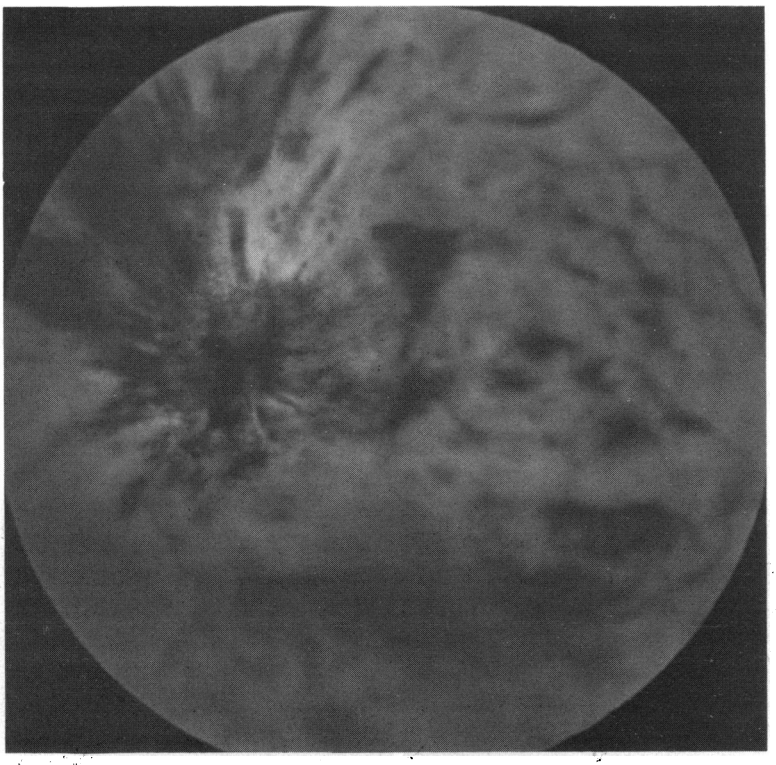

FIGURE 6
Case 5. Papilledema with many hemorrhages adjacent to the disc and in the posterior pole. Retina is grey-white.

over a few hours. All patients had some retrobulbar pain, usually worse with ocular movements. Patients 1, 2, 3 and 4 also had external evidence of inflammation, with erythemia of conjunctiva and lids.

The ophthalmoscopic appearance of the disc and retina was similar in all cases. The disc showed papilledema with blurred edges, and with hemorrhages around the disc area. The retina was grey-white, with a cherry-red spot in the macula. The periphery of the retina was relatively free of hemorhages. When fluorescein angiography was possible, the choroid showed normal filling, and the central retinal vessels were occluded. After the acute phase, optic atrophy with marked permanent narrowing of the retinal vessels developed. The retinal vessels often became totally obliterated and were seen as white cords. The macula usually showed marked cystic changes.

Neovascularization on the retina near the disc and neovascular glaucoma

developed in two patients. Three of the other four patients died from the underlying disease before the late changes could occur. The other patient did not return for re-evaluation after the acute episode.

DISCUSSION

The clinical differentiation of central retinal artery occlusion (CRAO) from central retinal vein occlusion (CRVO) is well established. The description of CRAO includes sudden loss of vision with the appearance of milky-white retina with a cherry-red spot in the macula. The retinal arteries may appear narrower than usual. Retinal hemorrhages are not characteristically present. The basis for the clinical picture CRAO is firmly established as an ischemic infarct due to the obstruction, either temporarily or permanently, of the central retinal artery.

Visual loss in CRVO is usually slower in developing and may range from a

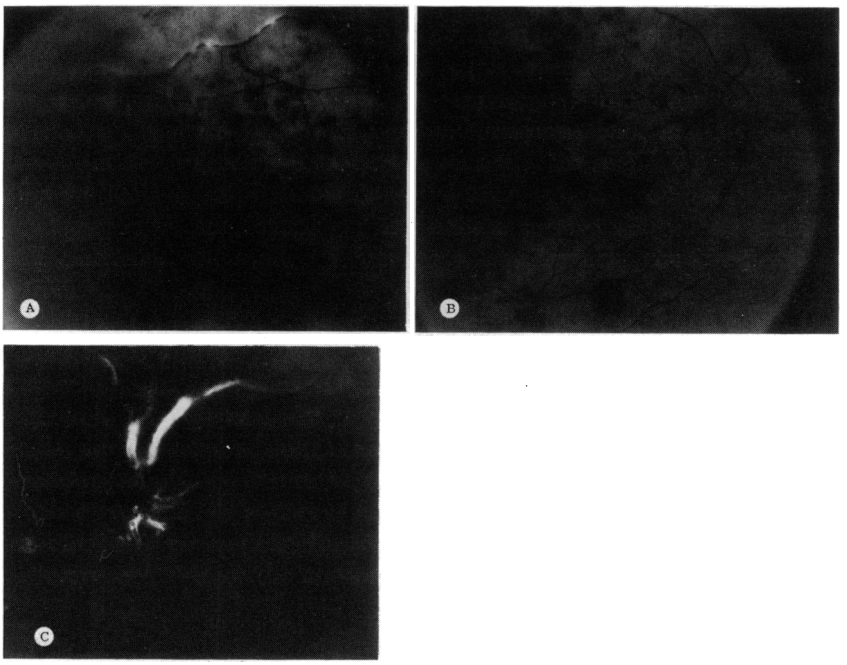

FIGURE 7

Fluorescein angiography, Case 5. A: Retinal hemorrhages obscure the choroidal pattern. Slight staining of vein walls in upper portion is the only indication of any fluorescein flow in the retinal vessels, 36 sec after injection. B: Periphery shows fading choroidal pattern and no retinal blood flow, 64 sec after injection. C: Staining of vessel walls at the disc, 10 minutes after injection.

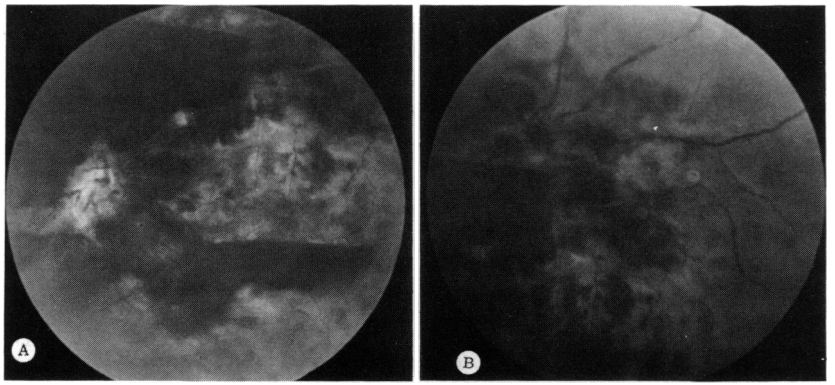

FIGURE 8
Case 6. A: Slight papilledema, large and numerous hemorrhages in the posterior pole, and grey-white retina. B: Hemorrhages are concentrated around the disc, and retinal periphery is relatively clear.

slight decrease to a profound loss. The disc is usually hyperemic with blurred margins, and superficial hemorrhages are present over the entire retina. The veins are engorged and retinal edema of varying degree may be present. The retinal changes in CRVO are more variable than in CRAO and the basis for CRVO is not as fully established.

Hayreh postulated there are two types of CRVO; one type, "venous stasis retinopathy,"[2] due to CRVO alone, and the second type, "hemorrhagic retinopathy,"[3] having the element of transient retinal ischemia combined with CRVO. Hayreh based his concept on experiments carried out on monkeys. In his first experiments[4] he occluded the central retinal vessels with diathermy as they emerged from the optic nerve in the orbit. In six monkeys he occluded the CRV, and observed that the retinal veins became engorged with the disc hyperemic but no retinal hemorrhages developed. He occluded the CRA in one monkey and the ophthalmoscopic appearance was typical of the clinical picture of CRAO. He occluded both the CRA and CRV in three monkeys at the point they emerged from the optic nerve in the orbit and produced changes very similar to those seen in our patients. Twenty hours after occlusion, the disc was elevated with blurred margins. A few hemorrhages were present adjacent to the disc and/or the macula, and the posterior pole was milky-white with a cherry-red spot in the macula. Later, more hemorrhages developed in the retina, probably due to re-establishement of the arterial circulation. Hayreh cited these findings as support for his concept of retinal ischemia plus CRVO causing hemorrhagic retinopathy.

In a later report[5] he cited a patient with chronic CRVO with a superim-

posed cilio-retinal artery occlusion, and described the appearance of hemorrhagic retinopathy in the area supplied by the cilio-retinal vessel. He stated that hemorrhagic retinopathy was really due to "retinal vascular occlusion," and that "simultaneous occlusion of the central retina artery and vein at their site of entry and exit from the optic nerve produces immediately the classic clinical picture usually associated with so-called central retinal vein occlusion, i.e., retinal hemorrhages, engorged retinal veins and retinal edema."

However, in a still later paper,[6] he reported further experimental work in monkeys. In these experiments he cut and cauterized the retinal vessels in the orbit as they entered the optic nerve. In one group, only the CRV was occluded; in other groups, both CRV and CRA were occluded, and the CRA occlusion being temporary for variable time periods, or permanent.

Permanent simultaneous occlusion of the CRA and CRV (both cut and cauterized in 11 eyes) produced the retinal changes described for our patients. After approximately 24 hours, disc edema was present, with adjacent hemorrhages in half the eyes. Cloudy swelling of the retina was present, more marked in the macular area, with a few hemorrhages in that region. (One animal of the group had many more hemorrhages than the others.) Many of the vessels subsequently developed sheathing and became progressively narrower. Retinal capillaries in the macular area became obliterated. The changes were not characteristic of those seen in CRVO in humans, either hemorrhagic retinopathy or venous stasis retinopathy.

In the monkeys with CRVO associated with temporary occlusion of the CRA, the retinopathy was similar to his hemorrhagic retinopathy type of CRVO. He concluded that retinal ischemia was necessary to produce that picture, but that combined CRAO/CRVO did not result in the classical picture of so-called CRVO.

Hayreh's experimental work[4,6] on combined CRAO/CRVO in monkeys resulted in an ophthalmoscopic picture like that seen in our patients. The experimental occlusions were done in healthy animals, acutely, in the area of the optic nerve behind the globe. Our patietns also had an acute process, and we believe the occlusion occurred in the retrobulbar portion of the optic nerve. We agree that the clinical appearance of combined CRAO/CRVO is distinctive and not the same as CRVO, either hemorrhagic retinopathy or venous stasis retinopathy. We believe our patients showed the result of complete and permanent (more than 48 hours) combined CRAO/CRVO.

Although others have not accepted Hayreh's concepts concerning CRVO and retinal ischemia, their reports do support the concept of combined

CRAO/CRVO as a distinct entity. McLeod[7,8] has published reports dealing with CRVO and cilio-retinal artery circulation, and in one,[8] retinal photographs of CRVO with an occluded cilio-retinal artery showed very similar changes in the area supplied by the cilio-retinal artery to those seen in our patients. He also produced experimental combined CRAO/CRVO,[9] and has reported these did not produce a fundus picture resembling clinical CRVO with hemorrhages. Details of the retinal changes were not included.

There are other clinical reports of central retinal artery disease in patients with CRVO. Niesel[10] reported that the intermediate pictures between venous thrombosis and retinal ischemia due to arterial obstruction occurred in retinal thrombosis. Paton, Rubenstein and Smith[11] reported a high incidence of central retinal artery changes in patients with CRVO. However, whether retinal ischemia secondary to retinal artery disease is necessary along with CRVO to cause the clinical picture of CRVO is still in question. Nonetheless, the ophthalmoscopic picture in so-called CRVO is distinctly different from the fundus changes described for our patients with combined CRAO/CRVO.

The changes we have described are different from those observed in total posterior infarction of the retina and choroid, such as occur with pressure on the globe during anesthesia and associated with a lowered blood pressure. In these patients the immediate ophthalmoscopic view is that of a CRAO. Hemorrhages and papilledema are not seen. Later, retinal pigmentary changes develop because of the choroidal ischemia. The retinal vessles may show the same late changes as observed in our patients, ie, almost total obliteration of vessels and absence of flow in retinal vessels during fluorescein angiography.

Neuroretinitis has some ophthalmoscopic aspects similar to those seen in our patients. The description of acute syphilitic neuroretinitis[12] includes marked disc edema, engorgement of veins, and scanty hemorrhages, leading to sheathed and obliterated vessels with an atrophic disc. One difference is the pigment proliferation that develops in the periphery with syphilitic neuroretinitis. Acute inflammation of the blood vessels at the optic nerve head fits our concept of a possible cause of combined CRAO/CRVO, and Smith[13] reported a patient with combined CRAO/CRVO in early syphilis. The patient was 46 years old and developed loss of vision and uveitis in one eye. The fundus was seen 18 days after the onset. A vitreous hemorrhage, multiple retinal hemorrhages, and extremely narrowed arterioles led to the diagnosis of combined CRVO/CRAO. The cause was thought to be syphilitic vasculitis.

There are other reports of combined CRAO/CRVO in the literature. Egerer[14] reported a patient with partial closure of the CRA and CRV

associated with retrobulbar optic neuritis. The fundus showed perimacular edema, peripapillary exudates and retinal hemorrhages, similar to the ophthalmoscopic changes described for our patients. Fluorescein angiography showed closure of arterioles and veins, particularly around the macular area. Histologic examination showed a retrobulbar neuritis associated with round cell infiltration around the central retinal vessels, impeding the flow.

Ridgway[15] and associates reported on the high incidence of optic nerve infiltration by white blood cells in children with leukemia. They reported that combined occlusion of the CRA and CRV caused permanent blindness unless radiation therapy was used at the first sign of vascular involvement. They noted spontaneous venous pulsation as the first phase, followed by progressive venous obstruction and complete loss of arterial and venous circulation. They did not describe the appearance of disc and retina at the time of combined CRAO/CRVO, but a fundus photograph showed essentially the same changes described for our patients.

Coppeto and Lessell[16] reported a 32-year-old woman with bilateral visual loss secondary to systemic lupus erythematosus. The retinal findings were swollen discs, pale retina and hemorrhages of several types, due to total and persistant arrest of the retinal circulation. Fluorescein angiography showed lack of retinal circulation and normal choroidal circulation. The authors noted that thrombotic and vasospastic occlusions of larger arterioles, often associated with evidence of vasculitis, occurs in systemic lupus erythematosus. The retinal photographs showed changes similar to those seen with our patients.

Stowe et al,[17] reported a 40-year-old woman with combined CRAO/CRVO which occurred while taking oral contraceptives. The brief clinical description stated diffuse flame-shaped hemorrhages and dilated veins consistent with CRVO were seen, and did not suggest any similarity to the retinal changes reported in our patients. Histological examination of the eye 16 months later showed occlusion of both the CRA and CRV in the optic nerve.

Cullen[18] described a patient with temporal arteritis who had CRAO in her left eye and a combined CRAO/CRVO in her right eye, with no light perception in either. Retinal photographs[19] of the right eye showed the typical changes described in our patients with combined CRAO/CRVO. Wagener and Hollenhorst[20] also reported a patient with combined CRAO/CRVO, also with temporal arteritis. No description of the retinal changes was given.

Gresser[21] reported a patient with bilateral partial occlusion of CRA and CRV, associated with thromboangiitis obliterans. Both eyes had similar

retinal changes, with edematous and blurred discs, faint cherry-red maculas, and retinal hemorrhages mainly in the posterior pole. The retinal vessels were attenuated with segmented blood columns showing reduced and irregular blood flow. Bender[22] reported a patient with bilateral occlusion of all the central retinal vessels. A woman, aged 32, had multiple tooth extractions and developed orbital inflammation and total loss of vision. The fundi could not be seen during the acute phase, but the description 1½ months later was bilateral optic atrophy, retinal vessels obliterated and visible only as white lines, and macular changes. A drawing was very similar to the retina in Fig 2.

Our patients and the case reports in the literature, except for Stowe's, have a common background of acute inflammation, vasculitis, and/or cellular infiltration in or adjacent to the optic nerve just behind the globe. In our patients, visual loss was rapid, from a few hours to one or two days, except in one patient. In that patient, retrobulbar neuritis apparently secondary to infiltration by lymphomatous cells was present for a month, progressing during that time and then resulting in combined CRAO/CRVO. Systemic vascular disease, ie, hypertension, diabetes, arteriosclerosis, atherosclerosis, was not a factor in any of our patients. The fundus changes were distinctive and characteristic. Treatment of the underlying cause was not effective in restoring vision.

SUMMARY

Combined CRAO/CRVO in our patients occurred with rapid visual loss, usually over a few hours, associated with evidence of inflammation and/or cellular infiltration of the retrobulbar portion of the optic nerve. The ophthalmoscopic appearance was characteristic, with papilledema and hemorrhages of various types in the posterior pole. The retina also showed ischemic changes, with a milky-white color and cherry-red macula. Fluorescein angiography, when possible, showed no retinal vascular flow, and normal choroidal flow.

After six to eight weeks, optic atrophy was evident and the retinal vessels were markedly narrowed or obliterated. The macula showed typical cystic changes. Neovascularization often developed, leading to neovascular glaucoma as the end result.

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DISCUSSION

Dr Thomas R. Hedges. I want to thank Dr Richards for sending me his paper far in advance so I could consider how he had seen six such patients whereas I had seen none—at least not in pure culture. Since many of my colleagues were of similar experience, we might conclude that we are misinterpreting the diagnosis or, as I suspect, such cases are extremely rare.

Common denominators in these patients are: young or middle age—average 42; acute onset and total loss of vision; a common anatomic location in the retro-laminar optic nerve where the artery and the vein come together in a common adventitia. Three patients were of possible septic origin, one had acute lymphatic leukemia, another lymphocytic lymphoma with lepto-meningitis, and one had no associated septic or aseptic process.

The mechanism of simultaneous central retinal artery (CRAO) and central retinal vein (CRVO) occlusion is intriguing. One might suggest that the artery is most probably involved first and the vein subsequently or in sequence. The author states "a common background of acute inflammation, vasculitis and/or cellular infiltration . . ." is responsible. "Septic" patients may, indeed, embolize inflammatory particles to the eye or develop vasculitis per se. They may also develop platelet aggregation and intra-vascular coagulation without vasculitis and still end up with the clinical picture of thrombosis seen here. Acute lymphatic leukemia patients with a rapidly rising lymphocyte count or one over 100,000 are noted to have central nervous system (CNS) involvement. Previous studies of acute leukemia in childhood have shown close association between the eye and CNS wherein nodular leukemic infiltrates and peri-vascular infiltration of the septa and meninges occur in the optic nerve (Trans Am Ophthalmol Soc 76:90-101, 1978). Thus other causes than "vasculitis" in the ordinary sense of the word can lead to vascular obstruction. The high white counts with increased blood viscosity may be the important factor leading to leukostasis and thrombosis, and the associated thrombocytopenia is thought to be an important factor leading to the massive retinal hemorrhages seen in two patients illustrated in this paper.

The striking clinical picture described combines diffuse retinal ischemia with a cherry-red spot, swollen optic nerve and a varying degree of retinal hemorrhage either confined to the disc area or massive as seen in the leukemia patients. Disc swelling does not occur in CRAO, but does occur in combined CRAO-CRVO. Papillitis or disc swelling does occur with posterior ciliary infarction—namely in anterior ischemic neuropathy and must be differentiated from combined CRAO-CRVO. Interior ischemic neuropathy, however, does not produce a wipeout of vision but rather a typical inferior altitudinal hemianopia.

Another type of patient which may be confused with partial CRAO-CRVO would be the hemorrhagic infarct patient with papillitis due to ischemic neuropathy. These patients are all part of a wide spectrum of arterial occlusive disease in one form or another and, if viewed thoughtfully, should be readily differentiated.

The author has used Hayreh's reports to show that the picture of CRVO can only be seen experimentally when the CRA is also occluded. This may be true for acute CRVO. However, Hayreh's postulate that venous stasis retinopathy is a form of CRVO is misleading, since venous stasis retinopathy is really a retinopathy of *chronic* reduced arterial blood flow (hypoxia)—hardly a similar clinical entity to CRVO.

In summary: The common denominators of young age, acute onset, blindness and retrolaminar location along with retrobulbar pain, sepsis or leukemia appear to justify Doctor Richards' conclusion that either acute inflammation, vasculitis and/or cellular infiltration are the common mechanism in this rare group of patients. However, intravascular coagulation and agglutination and leukostasis may be important factors as well and not just "vasculitis." Something unusual produces this rare clinical picture, that is for sure. It is unfortunate more pathologic studies of the acute condition were not obtained, but such is often the case.

The author is to be congratulated in bringing these patients into the spectrum of

optic nerve retinal vascular occlusive disease.

DR THOMAS P. KEARNS. This slide shows the fundus picture of a patient that I saw about two months ago. As can be seen, there is papilledema, venous engorgement, and widespread retinal hemorrhages. In addition to these features representing venous occlusion, the eye was almost blind—note the cherry-red spot in the macula—from an arterial occlusion. Simultaneous occlusion of the central retinal artery and vein is unusual, but I have seen a few examples of this over the last 30+ years.

Doctor Hedges pointed out in his discussion that we must not confuse this entity with venous stasis retinopathy of carotid occlusive disease. This (slide) is the classical picture of venous stasis retinopathy of carotid occlusive disease. This photograph is one that I showed at this meeting last year and appears in the 1978 Transactions in my paper on the ocular aspects of carotid bypass surgery.

Doctor Hedges first reported venous stasis retinopathy of carotid occlusive disease in 1962. Although he used the term "venous stasis" he did not use the term "venous stasis retinopathy." Doctor Hollenhorst and I must take the blame for coining the term "venous stasis retinopathy" to describe the retinal changes associated with severe occlusive disease of the internal carotid artery.

Venous stasis retinopathy of carotid disease is produced by the anoxia of the retina. It may be confused with an occlusion of the central retinal vein. However, one never sees the marked irregularity in the caliber of the veins such as you see here (slide) in a central retinal vein occlusion. The final test in differentiating the two is the retinal artery pressure. It will always be low in venous stasis retinopathy of carotid occlusive disease.

Doctor Hayreh has further confused the issue by using the term venous stasis retinopathy to describe a central vein occlusion. He even has said that there is no such thing as the venous stasis retinopathy of carotid occlusive disease. He believes that what Doctor Hollenhorst and I described in 1963 was not caused by carotid disease but was due to venous occlusion! Anyone who sees patients with carotid occlusive disease should have no trouble distinguishing these two entities.

In summary, when you see a patient with a retinopathy such as Doctor Richards has presented, there may be several causes. It may be central vein occlusion, a combination of a central vein and a central artery occlusion, or it may be the venous stasis retinopathy of carotid occlusive disease. The differentiation is not just academic. If you miss the diagnosis of venous stasis retinopathy of carotid occlusive disease the delay of treatment may result in a disaster for your patient.

DR J. TERRY ERNEST. I would suggest that it is highly significant that the late fluorescein angiograms did not show filling of the retinal arteries or veins. The choroidal circulation appeared to fill normally and if the disease process were limited to the central retinal vessels behind; the lamina cribrosa, the fluorescein should have been able to shunt into the retinal vessels from the capillaries of the distal segment of the optic nerve which are supplied by the short posterior ciliary

arteries. Since this was apparently not the case, perhaps the disease was primarily a papillitis.

DR ROBERT W. HOLLENHORST. This excellent paper by Doctor Richards points out the rarity of combined arterial and venous occlusion in the retina and established the cause, in most cases, to be of inflammatory origin. This controverts the contention of Doctor Hayreh based on animal experiments that the usual occlusion of a central vein results from arterial stenosis. I could not find the photo of the patient with temporal arteritis that Doctor Wagener and I reported in 1958, but that combined occlusion was due to inflammation of the optic disc. The patient was totally blind in the eye. (slide) This young patient with a tetrology of Fallot has cyanosis retinae. The complicating subacute bacterial endocarditis produced combined occlusion of the central retinal artery and of the central retinal vein, with massive ischemic edema of the retina. This artery is completely closed, and there is a great deal of hemorrhage along these occluded veins. The second slide is one of two slides of young women with Behcet's disease showing inflammatory involvement of the optic disc along with arterial and venous occlusions. The next slide will show the occluded artery as well as a moderate amount of hemorrhage. The vitreous contains many white cells at this time. This photo was taken about three hours after this eye went blind. The other eye was normal. The patient received prednisone treatment and the next two photos show the situation one month later: the pale, ischemic disc, the vein that was occluded and the occluded arteries.

DR CLEMENT MCCULLOCH. I would like to ask Doctor Richards if he used radiation on any of his three leukemic patients. It strikes me that this is one instance when we might be able to apply some positive therapy. It wouldn't have to be a large dose since radiation in leukemia is amazingly quick and effective.

DR MAURICE LANDERS. Fools sometimes rush in where angels fear to tread. I'm a little bit hesitant in this group of experts to suggest what I am going to, but perhaps I will learn more from it than any of you. I would propose an attempted treatment of some of these patients with acute central retinal artery occlusion. I would also bring to your attention three items of experimental evidence which have been presented in recent years as background for this. Some ten years ago, Doctor Banks Anderson, Jr, in his thesis for admission to this Society clearly demonstrated that oxygen is the flow-limited metabolite in these patients, and thus people with an acute central retinal artery occlusion lose vision because of a lack of oxygen. Secondly, Doctor Hayreh has recently shown at this year's ARVO meeting that at least in rhesus monkeys (who have ocular structures quite similar to humans) that a lack of circulation of upwards to an hour in the retina can in most cases be tolerated with a return to essentially normal retinal anatomy and apparently retinal function as well. Thirdly, in a paper that we had the opportunity to prepare for this Society we found that the retina could be well oxygenated by the choroid even when the retinal circulation was completely occluded if one simply had the patient breathe 100% oxygen (the patients in our cases were rhesus monkeys). We have not done a control

study on humans but nevertheless I think the concept may be applicable. Now humans cannot tolerate prolonged 100% oxygen breathing but they can tolerate it for a long period of time, perhaps as much as 24 hours or so. I realize that the vast majority of central retinal artery occlusions don't open up within 24 hours. However, a few of them do. Thus it is our feeling that when a patient presents with an acute central retinal artery occlusion, let's say hopefully of less than an hour's duration, it may not be unreasonable to attempt to administer a high dose of inspired oxygen for some short period of time such as a day or so. The patient may be one of the fortunate few whose central retinal artery spontaneously reopens during that period.

DR JAMES O'ROURKE. Doctor Richards has given us a wonderfully clear description of a puzzling vascular change in the fundus. A question of possible clinical interest here is whether or not an elevated blood viscosity could contribute to this picture, especially in patients having leukemia.

As you know this can be rapidly checked in most hematology or rheology laboratories using a cone-plate viscometer. And an elevated viscosity can be lowered by various medical therapies, sometimes with visible improvement in the fundus appearance after retinal venous occlusion. Since the negative effect of viscosity on flow is especially strong in venules and capillaries (and is greatly magnified when the packed cell volume exceeds 55%), it may be of interest to check this factor in patients such as Doctor Richards has described.

DR TULLOS COSTON. This may not be apropos to these cases that have been presented but I would like to remind you that in acute central artery occlusion in the younger people, well, say those that you would think would have dilatable retinal arteries, a stellate ganglion block can be very dramatically effective. One such case, a young man 21 years of age, who only had one eye, came to my office with occlusion of the upper branches of the central retinal artery. A translucent embolus had lodged on the disc where the vessels emerged. Only in the lower fundus was circulation normal. So I sent him immediately to the hospital and asked the neurosurgeon to perform a stellate block because ophthalmologists are not very keen to try to hit the stellate ganglion. He did this and while observing the fundus a translucent embolus was seen to pass out of the disc area into the periphery and his vision returned, except for a tiny defect in the periphery where the embolus lodged at a bifurcation. All retinal arteries in that young man dilated about 4 or 5 times normal size. This procedure is something to consider in the appropriate circumstances.

DR JOHN DYER. It just occurred to me that Doctor Brian Younge of our neuro-ophthalmology department about a year ago had a patient with acute central artery closure that occurred following neurosurgery and about 24 hours later he took the patient into the operating room, cannulated the superior orbital artery and flushed it with heparin in a retrograde manner and cleared out the central retinal artery and the patient's vision was restored. Doctor Kearns says that Doctor Peter Watson of England had done this on several patients and reported it almost ten years ago.

DR RICHARD RICHARDS. I appreciate the comments of the many discussors, and in particular the usual thoughtful discussion and helpful suggestions of Doctor Hedges. In reply to Doctor Ernest, we did not believe this was a papillitis. The vitreous was crystal-clear. There were no cells. The one histologic examination in the literature is that by Doctor Egerer. He showed essentially the same clinical changes and pointed out on histological examination that there was round cell infiltration around the vessels in the retrobulbar portion of the optic nerve. Doctor McCulloch, radiation therapy is used for these patients. Doctor Ridgway has pointed out that at the first sign of venous stasis in children with leukemia, that is the time to give the radiation therapy. If you wait until the clinical picture develops that our patients demonstrated, it is far too late. Our patient did get radiation therapy with no help.