A GREAT VARIETY of ocular signs have been described. This ocular sign, we believe, may imply increased orbital blood flow and, therefore, carries additional physiological significance for the clinician in that it emphasizes the importance of maintaining a patent external carotid artery.

Two representative cases are described.

Case Reports

Case I (H.L.)

A 57 year old, right-handed, hypertensive, white male was admitted to the Neurology Service of the East Orange, New Jersey Veterans Administration Hospital in January, 1977, with a history of several episodes of transient weakness of his left side for three months prior to his admission. Frequently the episodes were accompanied by fleeting sensations of giddiness and on one occasion with a complete loss of consciousness reported to have lasted 24 hours. The left-sided weakness on this occasion is said to have lasted about 48 hours. However, the subsequent attacks lasted no more than a "few minutes" to 2 to 3 hours. The patient was a known hypertensive who had had two previous admissions for uncontrolled blood pressure elevations.

Upon hospital admission he was found to be generally healthy but aethenic with normal blood pressure, mild left-sided weakness, and a right carotid bruit. Ophthalmologic examination revealed markedly dilated and tortuous episcleral vessels in both eyes, which on slit lamp examination proved to be dilated arteries (fig. 1). Fundoscopic and formal field examinations were normal and vision was correctable to 20/20 bilaterally. Intraocular pressures were normal. Ophthalmodynamometry pressures were 40/20 in both eyes.

While in the hospital, the patient experienced several fleeting episodes of transient right upper extremity weakness which often seemed to be related to postural changes. However, he reported no frank syncope or giddiness. Aortic arch angiography with selective catheterization of both common carotids and the left vertebral arteries demonstrated complete occlusions of both internal carotid arteries and the right vertebral artery. There was modest stenosis at the origins of both external carotid arteries which were markedly dilated and filled in retrograde fashion the...
ophthalmic arteries, distal intracranial internal carotid arteries, and their respective distributions on both sides. The left vertebral artery filled the vertebral-basilar circulation and there was some retrograde filling of both distal anterior cerebral arteries via the posterior pericallosal artery on the left.

Case II (E.T.)

A 60-year-old, hypertensive, right-handed, white male was admitted to the Neurology Service of the East Orange Veterans Administration Hospital in June, 1976, one week after the sudden onset of a mild right-sided weakness. During the 48 hours prior to admission, the patient reported progressive worsening of his right-sided weakness and difficulty with his speech.

Examination on admission revealed a hypertensive with mild expressive aphasia, a moderately dense right hemiparesis, and a mild right cortical sensory loss. A left carotid bruit was audible. A TC 99m-technetium pertechnetate static and dynamic brain scan showed evidence of a moderately sized left frontal-parietal infarction and marked reduction in flow in the left middle cerebral artery distribution. His subsequent course of improvement was punctuated by a sudden complete loss of vision in the left eye and worsening of his aphasia and right hemiparesis on the 11th day after admission. The previously noted left carotid bruit was no longer audible. His aphasia and right hemiparesis improved somewhat after several hours although his visual loss remained unchanged. His fundoscopic examination was consistent with a central retinal artery occlusion presumed to be the result of emboli from his diseased left cervical carotid artery.

After repeat brain scan revealed no change and a cerebrospinal fluid analysis was normal, the patient was given anticoagulant therapy. After improvement in his aphasia and hemiparesis he was discharged. He remained amaurotic in the left eye.

Four months later the patient was readmitted to the Neurology Service for reevaluation. Though his neurological deficit had impressively improved and his anticoagulant therapy had been well controlled, bilateral carotid bruits were noted on a recent clinic visit. Ophthalmologic examination revealed dilated episcleral arteries in the left eye, especially when compared to the right (fig. 2). Visual acuity was 20/20 in the right eye and 20/800 in the left with a small residual island of vision in the temporal field. Fundoscopic examination was consistent with an old central retinal artery occlusion and ophthalmodynamometry was 30/10 L.E. and 100/40 R.E. Intraocular pressures were normal.

Selective arteriography showed occlusion of the left internal carotid artery at its origin and modest stenosis of the origin of the left external carotid artery which was dilated and filled the intracranial left internal carotid artery and its distribution by retrograde filling of the greatly enlarged left ophthalmic artery. There was no evidence of collateral flow from the opposite carotid which was widely patent, or from the posterior circulation.

Discussion

Pavlou and Wolff first described dilated episcleral vessels in eyes ipsilateral to an internal carotid artery occlusion. They demonstrated by slip lamp examination dilatation of the arteries, arterioles, veins, and venules in 7 patients: 4 patients had arteriograms documenting internal carotid occlusions; in one patient an internal carotid artery occlusion was presumed; two patients had known occlusions of the homolateral common carotid arteries. No mention was made as to the status of the external carotid or ophthalmic arteries. Reference was made to various visual symptoms, including transient monocular blindness; however, details of the visual status in their patients were not reported. They concluded that the vessel changes were manifestations of chronic ocular ischemia and hypoxia.

Knox, in 1965, reported five patients in whom dilated episcleral vessels were associated with occlusions of the carotid arteries. In his patients, however, visual loss, diffuse episcleral vascular congestion, rubeosis, retinal changes, and other signs of a fulminating ocular inflammation were prominent. All of his patients had either diffuse and severe aortic
arch occlusive disease and/or multiple small vessel occlusive disease from diabetes. He concluded that the visual loss and inflammatory ocular changes were indeed due to ischemia and hypoxia associated with multiple large vessel occlusions and/or severe oligemia in the distribution of the ciliary arteries as well as the central retinal artery.

In none of our 7 patients was there evidence of ocular inflammation. Nor was there evidence of ongoing ocular ischemia. Of the 7 patients whose carotid arteries were studied, in only 1 was there any visual deficit. In this patient the visual loss was of sudden onset, clearly the result of a central retinal artery occlusion, and appeared from arteriography to be from an embolus because the ophthalmic artery and its collaterals were widely patent and there was no evidence of propagated thrombus from the internal carotid artery. The dilated episcleral arteries developed over the course of several months and were associated with the insidious recurrence of a carotid bruit which appeared to represent increased flow through a dilated external carotid artery.

Although ophthalmic artery pressures were significantly reduced, on the affected side of all of our 7 patients, the ophthalmic artery on arteriograms in each were not only patent but markedly enlarged. This was associated with tortuously dilated arterial channels in the orbit which filled the ophthalmic artery as well as the intracranial internal carotid artery and its distribution in a retrograde fashion. Lowered ophthalmic artery pressures are probably necessary to facilitate retrograde flow. In patients without signs of ocular ischemia, visibly dilated episcleral arteries are evidence of prominent collateral retrograde flow through enlarged orbital collateral channels similar to the situation that Fisher has pointed out with anastomotic channels about the orbital rim.\(^3\)

Dilated episcleral vessels may be an important physical finding in patients suspected of having an ipsilateral internal
carotid artery occlusion. Dilated episcleral arteries, particularly in the absence of other stigmata of ocular ischemia, strongly suggest that the external carotid artery is the major source of blood supply to the homolateral cerebral hemisphere. Consequently, maintaining patency of the external carotid artery in these situations is important.

Cerebrovascular Disease in Sickle Cell Anemia: A Clinical, Pathological and Radiological Correlation

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SUMMARY An opportunity to study cerebrovascular changes in sickle cell anemia (SCA) presented itself when a black child with this disorder died of bihemispheric strokes. Angiography demonstrated severe occlusive vascular disease involving primarily the circle of Willis and major bifurcations of both internal carotid arteries. Collateral circulation to the distal branches of the internal carotid arteries occurred through transdural anastomoses from the external carotid system and via the leptomeningeal route. Perfusion of the basal ganglia was accomplished by vessels arising from the proximal internal carotid arteries. These changes resembled those of Moyamoya disease. Autopsy showed old and recent cerebral infarcts. Two vascular processes were responsible for the arterial occlusions: (1) exuberant intimal hyperplasia, and (2) old and recent thrombi with partial recanalization. The former has been described only once before in SCA. Small vessels in the basal ganglia were exceptionally numerous and dilated. We conclude that intimal hyperplasia within large cerebral arteries may be responsible for infarction and small vessel proliferation in basal ganglia in patients with SCA.

Clinical Features

A 7-year-old black boy was admitted to Jackson Memorial Hospital with an acute alteration of consciousness. The patient was originally diagnosed as having sickle cell anemia at the age of 7 months. Originally he had edema of both hands and feet, was anemic, and had a positive sickle cell preparation. Subsequently, he had several hemolytic crises. Seven months before this last admission, the child developed a right hemiparesis with hemianesthesia and Broca’s aphasia. This was accompanied by focal myoclonic seizures involving his right arm and the right side of his face. He was given phenytoin and improved, but 4 months later he suffered a similar episode, which left him with moderately severe expressive aphasia. A Tc99 (Technetium) brain scan was diagnostic for occlusion of the left middle cerebral artery. His hemoglobin ranged between 6–8 gm/dl, and his reticulocyte count between 10–40%. Prior to his last admission he had been irritable and was later found comatose. In the emergency room, he was stuporous, and had right spastic hemiparesis, a right gaze preference and right-beating jerk nystagmus. His hemoglobin was 6.9 gm/dl, with a white cell count of 17,400 per mm³ and 16% reticulocytes. The cerebrospinal fluid contained 11 white blood cells per mm³ (45% granulocytes). He was treated with phenobarbital and diazepam and became more alert. Subsequently, an EEG revealed bilateral slowing, more marked on the left. Three days later, he became febrile (102.4°F) and more lethargic. Examination revealed nuchal rigidity and a positive Brudzinski sign. A second spinal tap showed 6850 red cells and 340 white cells per mm³ with 55% granulocytes and a protein of 322 mg/dl; CSF glucose levels were normal. Cultures of spinal fluid, blood and urine were all negative. A brain scan (Tc99) showed another area of increased uptake in the posterior right hemisphere. The child was treated with penicillin and a transfusion of 1000 ml packed cells after which he improved slightly. Hemoglobin electrophoresis showed only hemoglobin A and S. Three days later he became comatose with bilateral hyperreflexia, Babinski signs, a right gaze preference with nystagmus, bilateral decerebrate posturing...
Dilated episcleral arteries--a significant physical finding in assessment of patients with cerebrovascular insufficiency.
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