VENOUS ANGIOMA OF THE RETINA, OPTIC NERVE, CHIASM, AND BRAIN

A Case Report with Postmortem Findings

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In the literature we find many references to anomalies of the retinal circulation—abnormal tortuousness and unusual communicating branches between two different vascular areas. Such vascular anomalies, particularly when unilateral and associated with exophthalmos, indicate the retinal lesion is only a part of similar vascular malformations of the central nervous system, especially of the brain.

The case presented here for record of the unusual ocular findings was first seen by one of us (Krug) in the fall of 1931.

S. F., a male, aged twenty-one years, complained of defective speech, unsteadiness in gait, noise in the right ear of seven years' duration, restlessness, fatigability, depression with suicidal trends, and mental deterioration. The patient was the only child of healthy parents. There was a history of instrumental birth, and the mother stated that at the time of birth the right cornea was imperfect. A few days after birth a peculiar mottling of the skin of the right side of the face was noted. His mental development seemed normal, but physically he had never been strong. As a child he had severe and prolonged bleeding from his gums when a dentist lanced them to hasten the eruption of the right molar teeth, and again following a tonsillectomy he had severe hemorrhage due to the involvement of the mucous membrane of the right soft palate and tonsillar fossa by the angiomatous lesion.

In October, 1925, he had had what was believed to be an attack of acute epidemic encephalitis, in which he slept continuously for six weeks, followed by a month of delirium. The right upper and lower extremities became paralyzed and speech became impaired.

We quote the following from a description of this patient given in a separate paper entitled, "Venous and Arteriovenous Angiomas of the Brain," by Drs. Samuel Brock and Cornelius G. Dyke, to be reported in the Bulletin of the New York Neurological Institute:

"Patient walks with an ataxic gait. On the right side of the face and temple a raised soft angiomatous lesion is seen. The right ear pulsates synchronously with the marked throbbing of the right temporal and right carotid arteries. Over the temporal region a loud systolic bruit is heard which can be followed into the adjacent neck. It is not heard over the rest of the head. When the carotid arteries are compressed with the fingers, the bruit can no longer be heard, and at the same time the tinnitus of which the patient complains disappears. When the pressure is removed, both bruit and tinnitus reappear.

"While the facial innervation is normal on voluntary action, a definite left mimetic weakness is noted. Speech is slowed and somewhat scanning.

"The motor, reflex, and sensory status is normal. There are no heart murmurs. An *x*-ray examination showed that the heart was not enlarged. The pulse rate is 86; blood-pressure, 110/80. He is right-handed. The patient is intellectually deteriorated and is depressed.

"The roentgenograms of the skull show two unusually large vascular channels in the right frontal bone which course in a vertical direction. The right middle meningeal channel proceeds upward along the coronal suture for a distance of 3 cm., and then takes a sharp turn anteriorly, to disappear in the large vascular channels previously described. There is no abnormal calcification.

"The electrocardiogram was normal."

Ophthalmologic examination revealed the right eyeball proptosed slightly forward, downward, and outward, and its movements limited inward and upward, suggesting retrobulbar extension of the process. The eyelids were thickened, and the conjunctival vessels were full. The right pupil was semidilated and fixed, and consensual pupillary reaction was absent, showing a definite involvement of the optic nerve. The media were clear except

for a small macula at the lower limbus at 7 o'clock, which was most probably the result of trauma from instrumentation at birth. The eye was blind.

Ophthalmoscopic examination of the right fundus presented an unusual picture (fig. 1). The veins were so greatly engorged and tortuous that the disc was almost completely obscured. Where visible, there was no elevation of the disc. The vessels showed much thickening, indicative of a prolonged, progressive process. Where distinguishable, the arteries were not enlarged, and at no point was there any visible communication between the vessels. There was no pulsation of the vessels. Some pigment migration along the vessels was evident (fig. 2).

The veins of the left fundus were full, but not tortuous. With correction, -1.00 sph. $\bigcirc$ -0.75 cyl. ax. 180°, the vision of the left eye was normal (6/6 and Jaeger 1), and the field was normal for form and colors.

On March 18, 1932, the patient committed suicide by strangulation. The right eye, orbital contents, and the brain were removed on March 20th. The pathologic findings of the brain will be reported in Brock and Dyke's paper, previously cited.

At autopsy an angiomatous lesion was found extending from the right retina along the right optic nerve, chiasm, and tract to the right midbrain and via the right superior cerebellar peduncle to the right cerebellum (dentate nucleus), which explained the entire symptomatology.

DISCUSSION

Blood-vessel tumors are of two distinct major types: angiomatous malformations and hemangioblastomas or true neoplasms of the blood-vessel elements. The angiomatous malformations are undoubtedly the result of some fault of development, and may be distinguished clearly from the true blood-vessel tumors by the presence of traces of compressed nervous tissue between the vascular loops comprising the lesion.

Our case is one of congenital angiomatous malformation.

Through the studies of Florence R. Sabin[1] we have acquired a better understanding of the manner of development of the blood-vessels from the primordial endothelial blood-

Fig. 1.—Photograph of fundus at disc, showing tangle of enlarged tortuous veins.

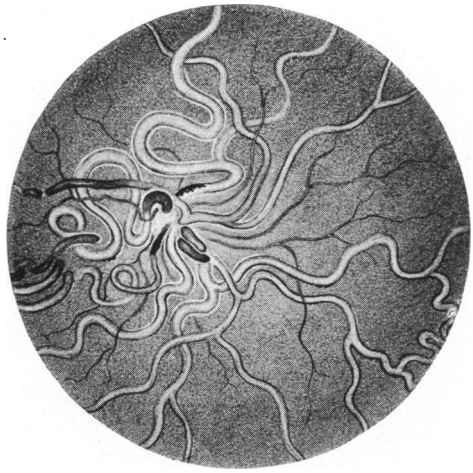

Fig. 2.—Drawing of nerve head and surrounding retina, showing migration of pigment cells along walls of vessels.

containing channels. These spaces are neither arteries nor veins, but constitute the source from which these vessels are derived, for they ultimately become linked into the circulatory blood channels. It is not difficult to understand how malformations within certain vascular areas can occur as the metamorphosis of primordial endothelial blood-containing channels into differentiated blood-vessels takes place. It is evident that the retention of the original vascular connections between arteries and veins will readily explain the origin of the adult arteriovenous aneurysms. The maldevelopment of the original vascular bed would similarly explain the nests of abnormal vessels which are interposed between the arteries and veins and which replace the normal capillary bed.

These vascular malformations are of three distinct types: (*a*) telangiectases or capillary formations (cavernomas); (*b*) venous angiomas (entirely venous); (*c*) arterial or arteriovenous angiomas.

We are concerned with the second group,—the venous angiomas,—of which there are the following distinct anatomic varieties: (1) the simple varices; (2) the serpentine, referring to a single, much enlarged vascular channel. This at times is associated with (3) a small racemose variety, forming a mass of interwoven, at times pulsating, blood-vessels.

Where the brain is involved, the racemose type either course on the surface of the brain or extend beneath the cortex in a wedge-shaped manner, like an inverted cone with its apex toward the ventricle, in which a terminal hemorrhage not infrequently occurs.

In general the venous system is not so regular in arrangement as the arterial, hence variations in it are much more frequent. When a new-formed blood sac furnishes communication between artery and vein, a varicose aneurysm is formed, but when the blood from an artery is forced di-

rectly into a vein which yields from pressure of the arterial blood, an aneurysmal varix follows. Without any weakening of its force the arterial blood passes directly into the vein, and it is not surprising that the vein becomes tortuous and distended. The force of the blood current also explains the great tortuosity of the smaller venous branches which give rise to the tangle of vascular loops. These branches, designed to carry back venous blood from capillaries to heart, now convey arterial blood to the heart.

The pallor of the blood in the varix is due in part to the mixture of arterial and venous blood and in part to the great thickening of the vessel wall. The vein takes on a darker color the nearer it approaches the disc, as more venous blood is added to the mixture.

Virchow's[2] classic, "Die krankhaften Geschwülste," published in 1863, gives a bibliography of the subject of angiomatous tumors in general prior to that date, with an admirable account of the effort to come to some determination as to the nature and proper classification of these lesions. Little was added to our knowledge until Lindau's[3] paper in 1926, followed two years later by the monograph of Cushing and Bailey,[4] and by Dandy's[5] paper in that same year.

Cushing and Bailey's contribution continues Virchow's review of angiomatous tumors, as it is a survey from 1863 to 1928, with a bibliography of 261 references covering this period. These investigators divide the cerebral blood-vessel tumors into two large groups—the hemangioblastomas and the vascular malformations. The former, which are true neoplasms, and, as shown by Lindau, are usually cystic, have their favorite seat in the cerebellum and in other parts of the central nervous system, such as the brain stem and the spinal cord. These growths are apt to be associated with benign, simple multiple cysts of the pancreas and kidney. Hypernephroma or adrenal adenoma may coexist.

Of the vascular malformations, Cushing and Bailey point

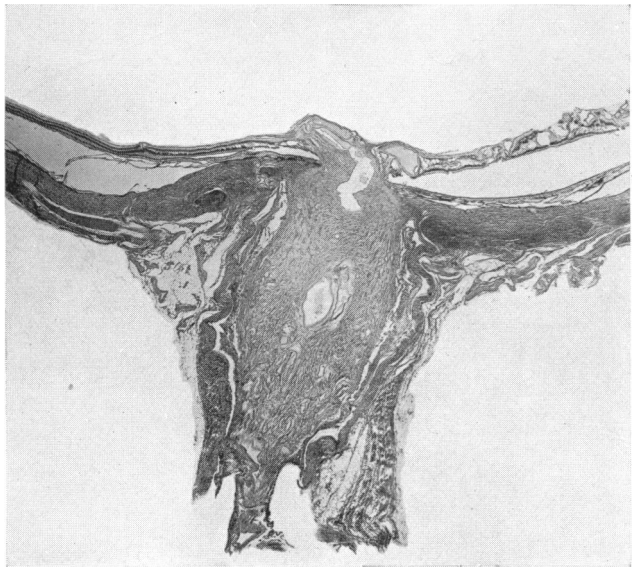

Fig. 3.—Enlarged and dilated central blood-vessels in nerve and retina. Cystic degeneration of retina.

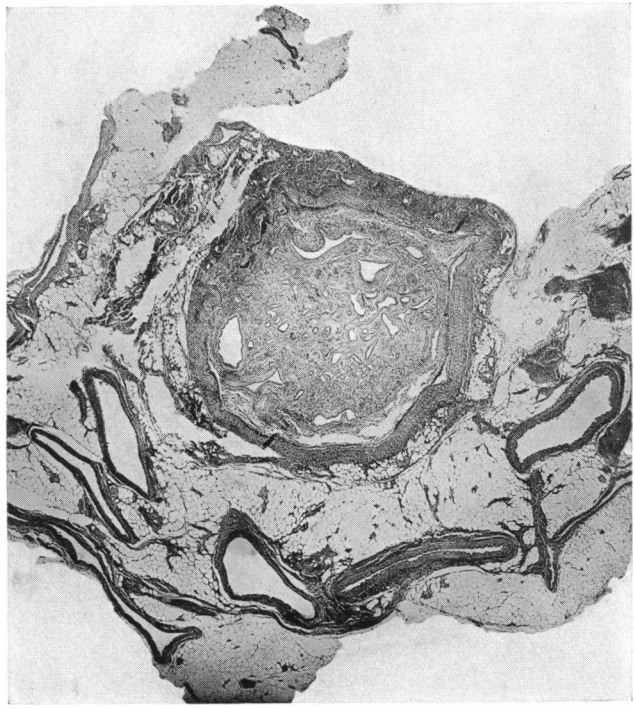

Fig. 4.—Cross-section of nerve near optic foramen showing stem of nerve to be composed almost entirely of blood-vessels. Orbital vessels are larger than normal.

out that it is most important to distinguish between a superficial variety, which is found on the surface of the brain, and a deep form, which extends into the cerebral substance in a wedge-shaped manner.

Dandy devotes his entire paper to this latter, more frequent, and more important group. The presence of a bruit in this group is indicative of the arterial participation in the malformation. The difficulties encountered in distinguishing the venous from the arteriovenous formations, the nature of the histologic changes, whether arteriovenous angiomas are venous lesions which later become arterialized—all these are questions which have been thoroughly discussed by Cushing and Bailey and others.

Cushing and Bailey also state that the presence of an interstitium of gliosed brain tissue between the abnormal blood-vessels is evidence of the essential non-neoplastic nature of the process.

In our case the presence of the enlarged, throbbing right carotid and temporal arteries, with the systolic bruit, indicates an arterial component entering into the vascular complex. In all probability the angioma was undergoing a process of arterialization before becoming an arteriovenous angioma.

The absence of choked disc, we believe, shows definitely that the venous element was the predominating one, as choked disc rarely accompanies venous angioma, although there may be exceptions to this rule.

The tinnitus in the right ear was due to the auditory perception of the systolic bruit in the adjacent enlarged temporal artery.

The presence of vessel abnormalities in the retina always demands a careful study of the visual field. Unless associated with trigeminal nevi, venous angiomas are more often overlooked than when an arterial component enters into the picture, with the development of an audible bruit.

HISTOPATHOLOGY (SAMUELS)

Tracing the structures of the eyeball backward, the cornea and sclera were found to be normal. There was nothing remarkable about the iris or ciliary body. The angle of the anterior chamber was free and normal.

The retina showed an unusually well-developed system of blood-vessels anteriorly, back of the ora serrata. The retinal blood-vessels in this region were larger than normal.

In the posterior segment the vessels attained so great a size and became so tortuous that they occupied more or less the entire thickness of the retina—they touched at points the pigment epithelium or the lamina vitrea of the choroid. They protruded far into the vitreous cavity. Between the walls of the blood-vessels the attenuated retinal tissue was in a high state of cystic degeneration (fig. 3), and here and there the pigment epithelium had propagated into the stroma.

The choroid showed nothing remarkable except some condensation under the vascular growth.

At the optic-nerve entrance many large, thin-walled vessels were found distended with blood. Most of them had the structure of veins rather than that of arteries. The central vessels as they passed through the cribriform fascia were unusually large. Posterior to the fascia they were accompanied by so many other vessels that the stem of the nerve appeared to be converted into a tangle of blood channels lined by endothelial cells and having mostly thin, atypical walls, similar to cavernous tissue (fig. 4). Occasionally a few vessels showed a more fully developed stage. In general, the blood spaces bore the characteristics of sinuses.

Between the vascular structures there was the regular septum system of the nerve, interspersed with islands of glial cells, among which were a few naked axis-cylinders. Aggregations of cells resembling "foam cells" were present at many points (fig. 5). No myelin could be seen throughout the length of the nerve. The vascular spaces grew larger as the nerve approached the brain. The diameter of the orbital division of the nerve was not increased in the sections, although in life, when the spaces were filled with blood, there must have been some increase in caliber over the normal. The intervaginal space revealed many typical psammomas. In the orbital tissues surrounding the optic nerve there were many blood-vessels of larger caliber than are commonly seen (fig. 6).

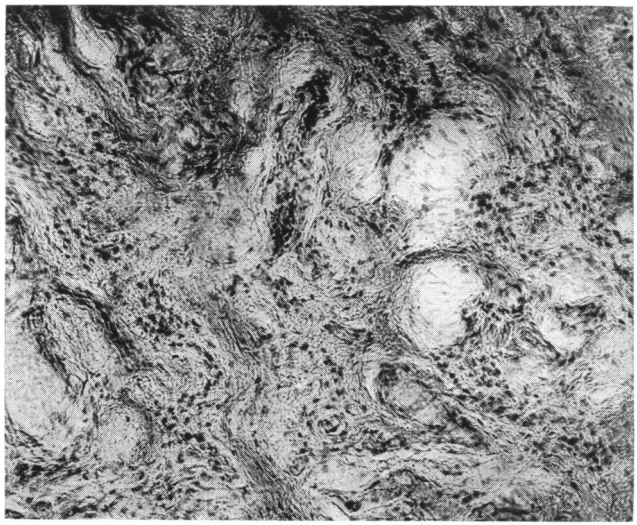

Fig. 5.—Islands of "foam" cells here and there.

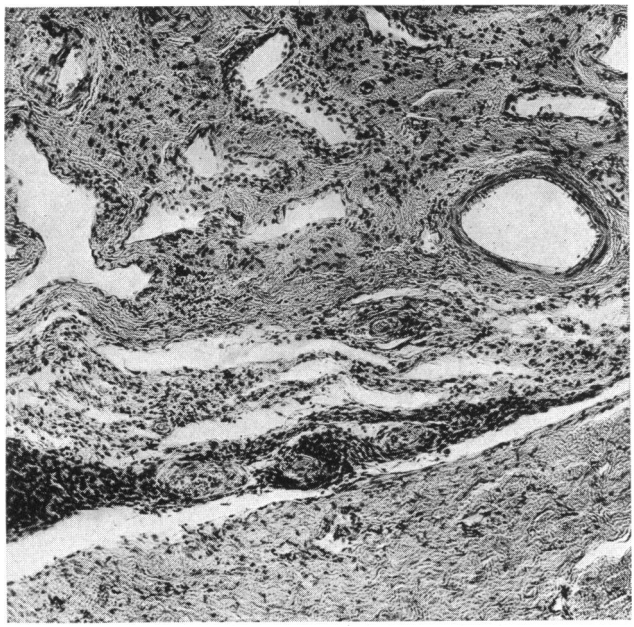

Fig. 6.—Detail of blood-vessels and interstitial tissue.

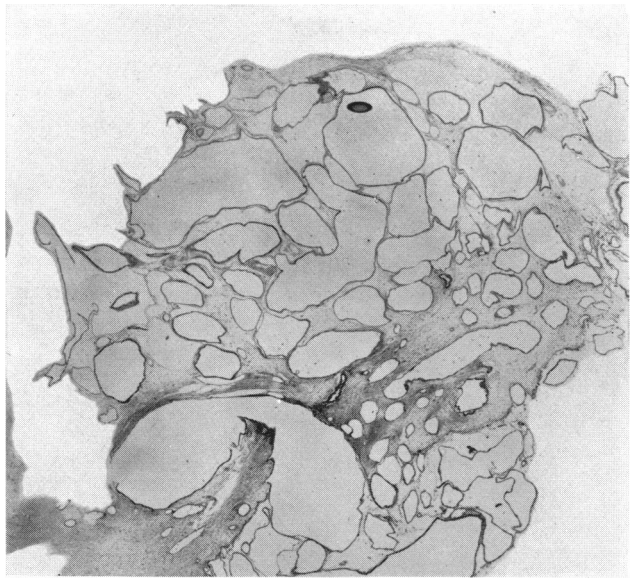

Fig. 7.—Large vascular channels in optic chiasm.

In the optic chiasm a still more unusual picture was noted. Here the vascular channels were exceedingly large (fig. 7). Some of them, by communicating, gave rise to large vascular labyrinths. Deep in the substance of the chiasm the vessels were surrounded by a fairly dense glial network. In one area of the chiasm the glial tissue appeared to be more dense, and gave the impression that the nerve fibers were here preserved; on careful examination, however, no fibers could be detected.

Diagnosis.—Venous angioma of the retina, optic nerve, chiasm, and brain.

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