CONGENITAL ARTERIO-VENOUS ANEURYSM OF THE RETINA*†

A POST MORTEM REPORT

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THERE are very few reports in the literature of arterio-venous malformation in the retina and mid-brain, and of these only one (Brock's Case 3) gives autopsy findings relating to the retina, optic nerve and tract (Krug and Samuels, 1932), and mid-brain (Brock and Dyke, 1932). The following additional autopsy report of an arterio-venous aneurysm of the right retina, optic nerve, optic tract, and mid-brain should, therefore, be of interest; the clinical findings during life in the patient, a 23-year-old female, have already been reported (Cameron, 1958). Briefly the main findings at that time were: a right temporo-parietal bruit; left hemiplegia; right ptosis with external ophthalmoplegia sparing abduction of the left eye; inactivity of the pupils to light and accommodation; left temporal hemianopia; normal left fundus, but arterio-venous aneurysms in the right; right optic foramen twice the diameter of the left; non-perception of light in the right eye, but visual acuity of 6/6 in the left.

Angiography showed a large vascular tumour (7×4 cm.) at the base of the brain, mainly on the right side, with displacement of the left anterior and middle cerebral vessels; ventriculography showed dilation of the lateral and third ventricles.

Early in 1960 the patient's condition gradually deteriorated; she became uncooperative and incontinent and died in April, 1960.

Pathological Material

The brain and eyes were removed *in toto*. Sagittal and transverse sections of the brain were made and photographed, and the brain preserved in 10 per cent. formalin for future dissection. It was intended at a later date to inject the blood vessels and make an accurate dissection, but 7 years later when time permitted, the brain was found to be grossly shrunken and in poor condition. However, the main features are clearly visible and, together with the photographs taken post mortem, permit a reasonable account to be made of the relevant pathology.

The lesion is seen to be a malformation of the right middle cerebral vessels which, by the hypertrophy of their walls and their extraordinary convolutions, occupy most of the third ventricle. The invasion of the upper end of the aqueduct was no doubt responsible for the partial internal hydrocephalus, and mid-brain involvement in this region also explains the ocular and pupillary pareses (Fig. 1, opposite).

Further laterally the widespread involvement of the right basal nuclei is shown by the great number of blood vessels on this side as compared with the left (Fig. 2, opposite).

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FIG. 1.—Close-up view of tumour. Note uninvolved right anterior cerebral artery coursing around the genu, and thickness of the right optic nerve.

FIG. 2.—Widespread involvement of the right basal nuclei (to the left of the photograph) compared with the left side.

On the surface of the right hemisphere the blood vessels were more numerous than on the left side and in the right parieto-occipital region the skull was very thin. However, the blood vessels were not nearly so numerous on the surface as at their origin.

The preparation for histological examination of the eyes, optic nerves, tracts, and geniculate bodies was begun immediately post mortem, when they were fixed in 10 per cent. formalin for 6 months.

The first specimen, consisting of the right eye, optic nerve, and optic tract as far back as the lateral geniculate body, was divided through the optic nerve and the two parts blocked separately.

The second specimen, consisting of the normal left eye, optic nerve, optic tract, and part of the lateral geniculate body, was divided, blocked, sectioned, and stained in exactly the same way as the first specimen, thus providing normal tissue for comparison.

The mid-brain was blocked, sectioned, and stained after 7 years' fixation in 10 per cent. formalin.

Results

(1) Right Eye

Macroscopic Appearances.—The globe was opened by a parasagittal section on the temporal side. The retina was in place and had the thick, grey, rugous appearance characteristic of eyes taken at autopsy. Grey, tortuous, bloodless, supernumerary retinal vessels extended upwards and downwards from the disc in the vertical plane. The globe was blocked in celloidin and sections were stained with haematoxylin and eosin, Masson's trichrome, and phosphotungstic acid haematoxylin (PAH).

Section of the globe showed retinal detachment due to processing and distortion of the retinal structure due to post mortem swelling. Large, abnormal, supernumerary retinal vessels were evident, running from the disc toward the equator. It was not possible to distinguish arteries from veins since all these vessels had fibro-muscular medial coats of variable thickness and wide, almost acellular, fibro-hyaline adventitial coats. Some of the abnormal vessels were so large that they occupied the whole thickness of the retina extending from the internal limiting membrane through to Bruch's membrane, to which they were adherent (Fig. 3). In all sections examined the retina exhibited a marked loss of nerve fibres and some diminution in the number of ganglion cells compared with the normal left eye. The other retinal layers and remaining ocular structures were normal.

FIG. 3.—Right eye. Supernumerary thick-walled vessels distorting the retina near the disc. One vessel is fused to the choroid below. Masson's trichrome. $\times 100$.

(2) Right Optic Nerve, Optic Tract, and Geniculate Body

The specimen was blocked in paraffin and sectioned transversely throughout its length at 1 mm. intervals. The long fixation in formalin made possible satisfactory preparations by Häcgvist's modification of the Alzheimer-Mann method for demonstrating axons and myelin sheaths. Sections were also stained with haematoxylin and eosin, Masson's trichrome, and PAH.

(a) *Optic Nerve and Optic Tract*.—The sections showed gross distortion of the nerve and tract by very numerous abnormal blood vessels. These were either venous channels consisting only of an endothelial lining and a thick fibrohyaline wall or large, thick-walled muscular arteries (Fig. 4a). The latter were also present in the surrounding dura and in the orbital fat outside it. Some of these arteries showed nodose thickenings of their medial muscle coats. No aneurysms were seen.

The glial compartments throughout were compressed and distorted by these aberrant vessels and the Hæcqqvist preparations revealed only very occasional structures resembling axons (Fig. 4b). By contrast, comparable sections from the normal left nerve and tract stained in the same way showed many hundreds of axons with their myelin sheaths per oil immersion field.

FIG. 4 (a).—Photomicrograph of right optic nerve 7 mm. behind the globe, showing marked excess of blood vessels. Some have thick, muscular walls separated by spongy neuroglia. Haematoxylin and eosin. $\times 100$.

FIG. 4 (b).—Right optic nerve, showing one of the better preserved glial compartments. No axons are recognizable. (Oil immersion) Hæcqqvist's method. $\times 1060$.

(b) *Lateral Geniculate Body*.—Multiple sections through the lateral geniculate body showed complete disorganization compared with the normal tissue from the left side. The geniculate body was grossly shrunken and consisted almost entirely of glia and large blood vessels of the type seen in the optic nerve and tract. Neurones and axons were present only in small numbers and were confined to one end of the tissue. The entering optic tract, easily recognizable in the normal geniculate body, could not be identified.

(3) Right Mid-brain

The characteristic large, thick-walled blood vessels are well seen. With Masson's trichrome (Fig. 5, overleaf) the walls stained blue, indicating a high content of fibrous tissue.

FIG. 5.—Right mid-brain, showing large dilated blood vessels. Masson's trichrome. $\times 24$.

Comment

The histological findings support the diagnosis of a congenital angiomatous malformation involving the right eye, optic nerve, optic tract, geniculate body, and mid-brain.

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