ANEURYSMS OF THE GREAT VEIN OF GALEN AND MID-LINE CEREBRAL ARTERIOVENOUS ANOMALIES*

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Since the first reported arteriovenous malformation of the brain in 1895,13 many types of midline intracranial vascular anomalies continue to be reported as aneurysms of the vein of Galen. In reviewing the reports on this subject1-12,14 it is apparent that failure to differentiate between the singular aneurysm of Galen and other racemose malformations in this region leads to considerable confusion.

The authors' experiences in the investigation and management of patients with this entire group of lesions suggest that at least three categories are separable both clinically and pathologically. As a result of this classification, not only is the pathogenesis of the lesions evaluated more readily, but they may also be divided into those that may or may not lend themselves to surgical management.

Three categories of midline arteriovenous anomalies may be defined as follows:

(A) Aneurysm of the great vein of Galen—A singular dilatation of the great cerebral vein, contiguous with a dilated sinus rectus and torcular and fed directly by anomalous branches of the carotid and/or basilar circulation.

(B) Racemose conglomerations of blood vessels deep in cerebral structures with dilated deep venous drainage—Vermiform clusters of anomalous arteries and veins (angiomas, hemangiomas, etc.) residing in midline or deep cerebral structures and draining centripetally into dilated deep veins and sinuses.

(C) Transitional types of midline arteriovenous shunts—
1. Singular vascular dilatations other than the great vein of Galen, draining into dilated sinuses and deep veins.
2. Combinations of midline angiomas accompanied by one or more aneurysmally dilated vessels.
3. Direct arterial shunts to deformed and dilated venous sinuses.

The lesions in group C, transitional between the true aneurysms of the great vein of Galen and the racemose angiomas, no doubt include numerous other possibilities.

A few illustrative cases from each group are presented.

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Group A—Aneurysm of the great vein of Galen.

Case 1. W.A.F., a 7-month-old female, was admitted on June 4, 1958 with a 3-month history of enlarging head and prominent veins of the scalp. A loud bruit was heard over the entire head and there was increased tone in all extremities. Carotid arteriography demonstrated a dilatation (3×4 cm.) of the vein of Galen (Fig. 1).

At operation, the aneurysm was exposed, and all demonstrable feeding arteries were clipped. Repeated arteriography revealed the aneurysm to be smaller but still filling well. A second operation was performed 2 months later, and the aneurysm was completely resected by clipping and separating arterial feeders, then transecting proximal to the torcula.

Postoperative seizures, hydrocephalus, bronchopneumonia and fluctuations in blood pressure all contributed to difficulty in management. Pre- and postoperative spasticity of all extremities is now resolving slowly, and a ventriculopleural shunt, after two revisions, is functioning well. It is noteworthy that pre-operative cardiac dilatation seen on roentgen-ray studies of the chest has entirely receded.

Case 2. J.E., a 9½-month-old female, was admitted on July 8, 1957 with a 1-month history of enlarging head and inability to sit alone. A loud bruit was heard over the head, which measured 51.5 cm., and prominent veins were noted over the forehead. Carotid angiography demonstrated an aneurysm (5.5×6 cm.) of the vein of Galen (Fig. 2).

On July 26, 1957, the right posterior communicating artery was exposed and clipped and, 2 months later, the left posterior communicating artery was clipped. In another 7 months, the aneurysm was exposed via a transventricular approach and two large arteries on its antero-inferior aspect were clipped under hypothermia. During this operation, the sagittal sinus was entered inadvertently.

The patient never regained consciousness, and significant amounts of air were found in the circulation at autopsy, suggesting that death was probably caused by air embolism. Figs. 3 and 4 illustrate the lesion and some of the pathological cerebral alterations caused by its presence.

Group B—Arteriovenous clusters of vessels with dilated deep cerebral venous drainage.

Case 3. Thalamic cluster of vessels with deep midline drainage. C.L., a 29-year-old