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
woman, was admitted on Dec. 27, 1953, with a history of subarachnoid hemorrhage and one seizure 4 years prior to admission. On admission to another hospital at that time arteriography was read as negative and a clot was aspirated from a lateral ventricle at attempted ventriculography. Arteriography, repeated by us, revealed a large midline cluster of anomalous vessels below the posterior portion of the pericallosal artery which was drained by dilated deep cerebral veins.

This patient was not operated upon, and was discharged to the care of her private physician.

Case 4. Deep right parietal cluster of vessels with deep midline venous drainage. B.G., a 16-year-old female, was admitted on March 26, 1954, with a 14-month his-
tory of intermittent headache, nausea and vomiting, associated with numbness of the left arm and toes. There was a history of weakness of the left arm following birth, and influenzal meningitis at age 8. On examination, there was mild weakness of the extensor muscles of the left arm and wrist and early bilateral papilledema.

At arteriography a cluster of deep right parietal vessels was seen which drained into a large internal cerebral vein and a markedly dilated (5 mm. diameter) vein of Galen (Fig. 5).

This patient was not operated upon, and is being followed by her private physician.

![Image](image.jpg)

**Fig. 5. Case 4.** Right common carotid arteriogram demonstrating deep parietal arteriovenous malformation and parts of its vascular supply and drainage.

**Case 5.** Deep right occipital cluster of vessels with deep midline venous drainage. A.R., a 21-year-old female, was admitted on July 1, 1958 with a 4-month history of intermittent headaches, abdominal pains, vomiting and loss of vision in the right eye. On examination, there was pallor of the right optic disc and a temporal defect in the right visual field with a central scotoma. Carotid and vertebral arteriography revealed a deep right occipital cluster of anomalous vessels supplied mainly by the right posterior cerebral artery and a branch from the posterior temporal and angular branches of the right middle cerebral artery. The area drained via the straight sinus (Fig. 6).

Ligation of the feeding vessels has been recommended for this patient.

**Case 6.* Deep left temporal cluster of vessels with dilated deep midline venous drainage.** F.O., a 54-year-old man, was admitted on Sept. 10, 1957 with a 14-year history of intermittent right hemiparesis, unsteady gait and right facial weakness. For 6 years he suffered headaches and attacks of unprovoked laughter. On examination, there were minimal spastic right hemiparesis, slurring of speech, coarse bilateral nystagmus and a constant inappropriate grin. Arteriography revealed a cluster of

* We are indebted to Dr. L. A. Mount for allowing us to include this case.
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left temporal vessels draining into a venous sac ventrolateral to the brain stem (Fig. 7).

After considerable debate a partial left anterior temporal lobectomy with removal of the contained angioma was performed under hypothermia.

The patient failed to regain consciousness and died 2 days postoperatively. Surgical and autopsy studies revealed an arteriovenous angioma of the left hippocampal and fusiform gyri draining into a dilated left basilar vein (Fig. 8) which had become imbedded in the midbrain, pulvinar and subthalamus on the left side.

Fig. 7. Case 6. Left common carotid arteriogram revealing left temporal arteriovenous malformation and massively dilated draining basilar vein. (Anteroposterior view is reproduced on the wrong side.)

Group C—Transitional types of malformations.

Case 7. Dilated midline vessel other than great cerebral vein. A.A., an 11-month-old female, was admitted in March 1952 with a history of left-sided seizures and dilated veins of the scalp on the right side of the head that measured 47 cm. in diameter. There was a left-sided hemiparesis. Arteriography revealed a single large vascular communication joining the right middle and posterior cerebral arteries with a markedly dilated left transverse sinus and deep cerebral veins.

She was discharged from the hospital without surgery.
Fig. 8. Case 6. Ventral view of cerebrum and section through midbrain showing bed of partially excised left temporal lobe and contained angioma and massively dilated basilar vein almost replacing midbrain.

Case 8. Combination of midline aneurysm and angioma. E.M., a 27-year-old woman, was admitted on Nov. 28, 1957 with a 2 1/2-year history of episodic headaches, nausea, vomiting and diplopia which began after mild head trauma. During three admissions to another hospital, she had xanthochromic cerebrospinal fluid although carotid and vertebral arteriograms revealed no significant pathology. Papilledema and seizures were observed prior to admission to the Neurological Institute. Examination revealed tremulous extremities and minimal long-tract signs. On ventriculography (Fig. 9) a well circumscribed mass was demonstrated posteriorly and superiorly in the roof of the third ventricle.

Craniotomy was performed on Nov. 29, 1957 by a right transventricular approach which revealed a mass bulging into the posterior medial wall of the right lateral ventricle. The ependyma was incised, revealing a thickly encapsulated sacular structure which yielded mushy rust-colored material on aspiration. The sac was partially amputated at the point where it dipped into the posterior thalamus.

Continuing signs of subarachnoid hemorrhage postoperatively prompted repeated carotid arteriography which demonstrated no significant vascular abnormality and a normal-looking vein of Galen.

Surgery was repeated on Jan. 3, 1958 in an attempt to arrest the slow hemorrhage. The partially amputated vascular sac was approached once again, ligated at its base and removed.

Death followed signs of repeated hemorrhage and decompensation of the central nervous system in spite of supportive measures including hypothermia.

Necropsy revealed that the amputated vascular sac was actually a thick-walled aneurysmal dilatation overlying a maze of partially thrombosed angiomatous vessels which occupied the posterior portion of the third ventricle and contiguous lateral ventricles as well as the medial thalamic nuclei (Fig. 10). Signs of prolonged hemor-
rhaging were manifested by the infiltration of blood pigments within the spinal cord and brain stem, fibrous thickening of the meninges and a granular ependymitis.

Case 9. Major arteries entering dilated venous sinuses. J.A.R., a 15-month-old female, was admitted on Nov. 24, 1953 with a 5-month history of right-sided seizures. A bruit was heard over the skull, which measured 48 cm. in diameter and had a bony depression over the confluence of the sinuses. The veins were prominent on the left side of the face and there was motor and mental retardation. Carotid and vertebral arteriography revealed massive dilatation of the torcula and adjoining dural sinuses which appeared to be fed by cerebellar, posterior communicating and a dilated left internal carotid circulation (Fig. 11). Other vascular malformations were detected in hemispheres adjoining the floor of the middle fossa.

No surgical therapy was attempted in this case.

Fig. 9. Case 8. Ventriculogram demonstrating mass arising from posterosuperior roof of third ventricle which proved to be an aneurysm associated with arteriovenous malformation of posterior thalamus and choroid plexuses.

Fig. 10. Case 8. Median sagittal section demonstrating arteriovenous malformation in the posterior part of the third ventricle and defect in corpus callosum where aneurysm was surgically removed. The dural reflection of falx and tentorium is entered by a dilated vein of Galen.
Str. 11. Case 9. Left common carotid arteriogram demonstrating massive dilatation of torcular and adjoining venous sinuses and direct arterial feeders.

Case 10. Temporal and midline arteriovenous cluster with dilated great vein and sinuses. J.P.R., a 14-year-old boy, was first admitted in 1937 with a history of tremor and clumsiness of the right arm since age 6. There was a mild right hemiparesis with increased right-sided deep tendon reflexes. Plain roentgenograms of the skull revealed a curvilinear calcification in the region of the left thalamus.

Six years after this admission he suffered a subarachnoid hemorrhage and was managed in another hospital.

Three years later (at age 23) he was admitted after a second subarachnoid hemorrhage, and the left internal carotid artery was ligated.

There were no further episodes until age 28 when another subarachnoid hemorrhage occurred. The right-sided hemiparesis, hyperreflexia and tremor persisted. Carotid arteriography at this time revealed an abnormal cluster of vessels in the left posterior cerebral distribution as well as a large arteriovenous cluster in the left temporal lobe and marked dilatation of the inferior longitudinal and straight sinuses, and the vein of Galen.

No further therapy was attempted.

DISCUSSION

In considering first the aneurysms of the great vein of Galen, it is noted that the lesions were composed of a thick-walled sac which grossly and microscopically resembled dura mater fed directly by arteries from dilated carotid and/or basilar arterial branches. Drainage took place via a continuation of the sac into a wide straight sinus and torcular. The mass of the aneurysm pressed upon the dorsal midbrain, resulting in aqueductal reluctance and dilatation of the ventricles. Clinically the history began in the neonatal period with enlargement of the head, prominent veins of the scalp and a loud cranial bruit which could not be obliterated by carotid compression. Cardiovascular defects and some degree of cardiac decompensation were
variable findings. Demonstration of the intracranial lesion by angiography usually was not difficult.

Our experiences with this type of lesion suggest that total excision is possible under hypothermic anesthesia and should be attempted when the diagnosis can be established. The surgical approach to these malformations is greatly simplified by the presence of concurrent hydrocephalus. Once the ventricles are drained at operation the arterial feeders can be exposed, clipped and severed, thus allowing one to roll the aneurysm gently out of its bed. The venous stalk, which enters the torcula, can then be ligated and sectioned and the aneurysm delivered. The use of hypothermic anesthesia aids in protecting the patient against the effects of surgical manipulation in the region of the third ventricle. The untreated malformation, we feel, is probably incompatible with normal development, manifesting itself as it does in infancy and leading to hydrocephalus and compression of the brain stem, as well as cardiac decompensation.

In contrast to the foregoing lesions, the arteriovenous mass of vessels with dilated deep venous drainage occurring in and about the same region (Group B) may not include the great vein of Galen itself. However, the great vein may be somewhat displaced or engorged. These malformations occur within or about the posterior thalamus and midbrain, and may extend to or from contiguous cerebral areas, choroid plexus, etc. The arterial supply and venous drainage as well as the histologic character of the clustered vessels in the lesion are quite variable. Clinically these patients are often asymptomatic until the second to fifth decades when signs of subarachnoid hemorrhage first occur and, like arteriovenous angiomas elsewhere, are likely to bleed repeatedly. A bruit is not often audible. Ventricular dilatation is not common, but has been noted secondary to subarachnoid hemorrhage. Seizures are common and usually reflect the location of the anomaly or result from hemorrhagic cerebral damage. We do not believe that these malformations are surgically resectable as they commonly involve such vital neural structures. While direct ligation of feeding vessels may be practicable in some instances, these lesions may not curtail normal life expectancy.

Lesions that are herein considered transitional cannot be categorized with any degree of certainty by arteriogram or autopsy because of their overwhelming ramifications. They are demonstrated, for example, as combinations of racemose vascular conglomerations with aneurysmal dilatations, or singular vascular dilatations aside from the great vein of Galen, or dilated and malformed venous sinuses fed directly by arteries, etc. The very nature of lesions of this type makes them surgically inaccessible because of technical limitations and their integral association with vital neural structures.

CONCLUSIONS

1. Three major categories of arteriovenous malformations of midline cerebral vessels are described as a means of facilitating improved classification and therapeutic management.
2. Illustrative examples of each category are presented.
3. Indications for therapeutic management are discussed.

REFERENCES


