Case Reports and Technical Notes

Aneurysm of the Great Vein of Galen

Report of Two Cases

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In 1967 we reported a case of aneurysm of the great vein of Galen with the intention of establishing an association between it and coexistent venous drainage abnormalities. Recently, another case has given us the opportunity to reestablish a relationship between these two rare phenomena since a chronology of the evolution of the pathology is available on angiograms.

Complete background data and review of the literature can be obtained from our preceding publication on the subject,* and to fully appreciate the pathophysiology involved only the barest outline of postulated events need now be reviewed. An intracranial arteriovenous malformation, usually in a position to be fed by a posterior cerebral artery, is the original developmental anomaly setting the stage for subsequent, secondary venous abnormalities. The greatly increased volume of shunted blood flowing through the draining deep venous system results in aneurysmal dilatation of the great vein of Galen, a thin-walled distensible structure. Finally, through a mechanism that is not understood (perilesional hemorrhage, thrombophlebitis within the malformation, etc.), a significant number of these cases show lesional thromboses, probably initially within the small veins draining the malformation. Extension of clot, either directly or by embolization, may then occlude more distant, larger venous structures, including both the aneurysmally dilated vein of Galen and draining dural sinuses. Such an occurrence then necessitates unusual collateral pathways for venous drainage, a phenomenon which may be clinically as well as angiographically apparent, and depending upon the final arrangement attained, may either result in an untenable hemodynamic status, or alternately, spontaneous cure of the original lesion as in our second case.

From a clinical standpoint, these patients characteristically show rather predictable age dependent sets of symptoms. In infancy, they manifest high output cardiac failure if arteriovenous shunting is extreme; in childhood, obstructive hydrocephalus secondary to aqueductal compression by the adjacent aneurysm is the rule; and in adolescence and adult life, the patient generally presents with intracranial hemorrhage. In those patients with secondary venous obstructions who manifest externally apparent venous collaterals, roughly one-fifth of the 51 cases in the literature, we would add another complaint, namely, cosmetic difficulty from facial varicosities.

To emphasize the course of these patients and their radiographic findings, we present two cases. For a more complete review of Case 1, the previously cited work may be consulted.

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Case 1. An 11-year-old boy was sent to the University of Florida Teaching Hospital for cosmetic repair of prominent, superficial forehead veins. These had prompted medical appraisal with bilateral carotid arteriography by the time the patient was 20 months old, and, with the recognition of the accompanying intracranial component of his pathology, the varicosities were deemed surgically unremediable. Carotid angiograms repeated at 7 and 11 years of age showed substantially

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* Since publication of our article, two additional cases (see refs. 2 and 3) of venous occlusions in association with aneurysms of the great vein of Galen have been described in the literature.
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the same findings. Other than occasional mild, poorly-localized headaches of brief duration, the patient was asymptomatic.

Examination. The patient showed compressible greatly-engorged veins extending from the forehead down over the nose and onto the cheeks. A continuous bruit with systolic accentuation could be heard over this area, while a venous hum was audible over the calvarium more posteriorly. During lumbar puncture, the opening pressure of 300 mm of water did not rise with compression of either or both jugular veins, but on compression of dilated veins over the forehead, a rapid and significant rise of cerebrospinal fluid pressure was observed. The fundi showed venous engorgement and indistinctness of the disc margins without hemorrhage or exudate.

Roentgenographic Examination. Skull x-ray films showed two types of findings, one related to lack of flow through normal channels (small jugular foramina), and the other to increased venous flow through collateral routes (large superior orbital fissures). There was evidence that the sigmoid sinuses had developed (sinus plates in mastoid areas) and presumably were at one time functional. Right carotid arteriography disclosed no change in the arteriovenous malformation, aneurysm of the great vein of Galen, or in venous drainage when compared to earlier studies (Figs. 1 and 2). The left side mirrored the same abnormalities.

The deep arteriovenous malformation was felt to be inoperable, and, since the facial varicosities represented the only route for drainage of intracranial blood, it was thought inadvisable to attempt plastic repair of the forehead region.

Case 2. A 37-year-old white man was referred to the Gainesville Veterans Administration Hospital because of diplopia, decreased visual acuity, nausea, and vomiting.

Past History. The patient was said to be hydrocephalic by the age of 6 months, but this became arrested within another 6 months, and he experienced no further difficulty in childhood. Upon completion of the eleventh grade, he enlisted in the Air Force where he remained until 1962. At that time

Fig. 1. Case 1. Right carotid arteriogram, lateral views. Left: Arterial phase. The enlarged right internal carotid artery delivers most of its blood to the dilated right posterior communicating and posterior cerebral arteries. The posterior cerebral artery branches into a racemose network of vessels which drain promptly into a dilated and deformed 16 x 45 mm great vein of Galen. There is early filling of the dilated straight sinus and angiographic evidence of minimal ventricular dilatation. Right: Early venous phase. The great vein of Galen has almost emptied, and there is opacification of the confluence. transverse sinuses, and, faintly seen, the superior petrosal sinuses (arrow). There is no opacification of the sigmoid sinus. (Reprinted by permission of the editors of Radiology, from the April, 1967 issue, vol. 88, pp. 725-729.)