Arteriovenous Malformation Involving the Inferior Sagittal Sinus in an Infant*

Case Report

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Although there has been an increasing interest in the problem of intracranial venous aneurysms, case reports dealing exclusively with them have been relatively infrequent. In 1964, Gold, et al., were able to collect only 34 cases from the literature and in addition reviewed eight of their own. For the most part these congenital intracranial venous aneurysms have demonstrated a predilection for the region of the vein of Galen and invariably they have been associated with an arteriovenous malformation that has involved branches of several of the major intracranial vessels. It is believed that the pathogenesis of these venous aneurysms is related to the shunting of arterial blood under high pressure through a maldeveloped capillary system. This basic developmental defect permits blood under arterial pressure to enter thin-walled veins which, under these circumstances, dilate and become aneurysmal in appearance. It is then not unreasonable to expect that any of the intracranial veins or venous sinuses could be involved.

In general, the surgical approach to aneurysms of the vein of Galen has been to attempt to occlude the arterial supply and thus "shrink" the aneurysm and at times relieve the patient's symptoms. Obviously, because of the location of the lesion, as well as the general nature of the problem, a direct surgical attack is fraught with a high degree of morbidity and mortality. Nevertheless, several case reports have indicated that these lesions should be excised surgically.

This case report is concerned with the successful surgical excision of a venous aneurysm of the inferior sagittal sinus, which in all probability developed secondary to a congenital midline arteriovenous malformation in the frontal region.

Case Report

This 5½-month-old male infant came to the University of Minnesota Hospitals with a 3-week history of progressive enlargement of the head, complicated for the last few days by a number of left-sided focal seizures. Prenatal and neonatal histories were not contributory except that the parents had noted that his development had seemed somewhat "slow" in comparison to that of his siblings.

Examination. The patient was a well-developed, somewhat lethargic child in no obvious distress. The head circumference was 46 cm. There was a prominent venous pattern in the scalp of the frontal region and the fontanel was firm. Auscultation revealed a moderately loud bruit, which was best heard in the left frontal area. There was no specific clinical evidence of focal deficit, but there was a moderate degree of developmental retardation.

From a clinical standpoint, it was believed that the most likely diagnosis was that of an arteriovenous malformation, possibly in association with an aneurysm of the great vein of Galen, although the distinct possibility of a cerebral neoplasm or other intracranial space-occupying lesion was also considered.

Skull roentgenograms revealed an early diastasis of the sutures, and a technetium-99 brain scan was consistent with an area of increased uptake in the midline region anteriorly (Fig. 1). A routine chest roentgenogram demonstrated cardiomegaly. With the suspicion of an arteriovenous anomaly more firmly established, a right retrograde brachial angiogram was performed under local anesthesia (Fig. 2). This showed the presence of a large frontal midline arteriovenous malformation, draining directly into a venous aneurysmal dilatation of the inferior sagittal
sinus. There was also a moderate degree of dilatation and tortuosity of the transverse and straight sinuses, the vein of Galen and the posterior aspect of the inferior sagittal sinus. The superior sagittal sinus appeared normal. The malformation was fed principally from a dilated right anterior cerebral artery; the right middle and posterior cerebral arteries appeared to be uninvolved. A left percutaneous carotid angiogram demonstrated a significant contribution from several large branches of the left middle cerebral artery which coursed over the cerebral convexity (Fig. 3). The left anterior cerebral artery was poorly visualized partly because of technical problems and partly because of its configuration.

Considering the progressive symptomatology and the known propensity for these lesions in this age group to enlarge gradually and progressively, particularly after they have once become symptomatic, we concluded that surgical excision of the lesion was the only advisable form of therapy.

Operation. A standard coronal skin incision was made and a bifrontal craniotomy