Excision of a cirsoid arteriovenous malformation of the corpus callosum in a 16-year-old boy

Case report

Thomas H. Milhorat, M.D.
Branch of Surgical Neurology, National Institute of Neurological Diseases and Stroke, National Institutes of Health, Bethesda, Maryland, and the Department of Neurology and Neurosurgery, The Children's Hospital of the District of Columbia, Washington, D.C.

Cirsoid arteriovenous malformations, although rare, seem to occur most commonly in the region of the corpus callosum. Nine cases have been reported and of these six have been operated on. A review of the literature suggests a number of common features: 1) in contrast to most arteriovenous malformations, cirsoid lesions of the corpus callosum are well-circumscribed and consist of a tight coil of anomalous vessels that form a localized midline mass; 2) the malformation is usually nourished by the pericallosal arteries from both sides and is drained by the inferior sagittal sinus or great vein of Galen; and 3) the clinical picture is generally that of recurrent subarachnoid hemorrhage without localizing signs.

The current case is of interest because the patient is the youngest to be reported to date and because the case is only the second in which a complete excision of the lesion has been demonstrated.

Case Report

A 16-year-old Negro boy was apparently well until he was 14 years old, when he experienced the first of seven attacks of headache, fever, and stiff neck. The attacks were characterized by an abrupt onset of severe throbbing bitemporal headache unrelated to activity or other precipitating factors. Within 3 to 6 hours, fever and stiff neck were usually prominent, and these complaints would persist for 3 to 4 days. With each attack the patient was forced to bed for several days, but he complained of no other symptoms and was otherwise well. Following each attack he recovered fully and returned to school. During the interval between attacks (the shortest being 3 weeks, the longest 5 months), he maintained his average scholastic record and participated actively in sports (the patient stands 6 ft 5 in., weighs 230 lbs, and has hopes of playing professional basketball). Two days following the patient's seventh and most recent attack of headache and stiff neck, he was admitted to the Children's Hospital of Washington D.C.

Examination. The general physical examination on admission was unremarkable. The neurological examination was also normal except for moderate neck rigidity and bilateral retinal hemorrhages. No bruits were heard. A lumbar puncture showed grossly bloody cerebrospinal fluid which was found
to be faintly xanthochromic. A bilateral carotid arteriogram demonstrated a midline cirsoid arteriovenous malformation that was richly supplied from both sides (Figs. 1 and 2). A fractional pneumoencephalogram revealed a 3 x 3 x 2 cm mass in the area of the corpus callosum, projecting into the frontal horn of the right lateral ventricle (Fig. 3).

Operation. Under routine general endotracheal anesthesia, a free bone plate was removed from the right parasagittal area, and the corpus callosum was approached by retracting the right frontal lobe away from the falx (Fig. 4). At the base of the longitudinal fissure a racemose arteriovenous malformation was identified overlying the corpus callosum and embedded within it. The lesion was compact and circumscribed and consisted of a tight coil of vessels fed by the pericallosal arteries and drained by a number of large and small veins to the inferior sagittal sinus. The vascular mass was surgically isolated by systematically obliterating the arterial supply with multiple silver clips. This resulted in a modest collapse of the malformation which was then further isolated by clipping the larger venous channels. By this means it was possible to excise the lesion uneventfully and to deliver it in one piece from its bed in the corpus callosum. The callosal defect measured 1 x 3 cm at the conclusion of the procedure and permitted direct examination of the frontal horn of the right lateral ventricle. No vascular communications to the thalamostriate vein, choroidal vessels, or deep cerebral veins were seen.

Postoperative Course. Recovery was uneventful. The patient was up and walking on the 7th postoperative day and was found to have no neurological deficits. On the 21st postoperative day a brachial arteriogram was performed which indicated that the vascular malformation had been completely excised (Fig. 5). The patient has since returned to school where his record remains average. Preliminary psychological and intelligence tests reveal no significant deficiencies.

Discussion

Although the 1966 Cooperative Study on 545 cases on craniocerebral arteriovenous malformations reports a grouping of 15

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FIG. 1. Left: Arterial phase of right carotid arteriogram demonstrates a deep (3 x 3 x 2 cm) arteriovenous malformation just to the right of the midline. Right: Left carotid arteriogram demonstrates the bilateral arterial supply of the lesion.