Familial Intracranial Aneurysms

Report of Four Cases

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The pathogenesis of intracranial aneurysm remains controversial. Most writers on the subject believe that cerebral aneurysms are of congenital origin. Aneurysms have been found in very young, even newborn, children. Unusual examples have been reported by Garcia-Chavez and Moosy,7 Jones and Shearbun,8 Newcomb and Munns.11 The concurrence of cerebral aneurysm with other congenital lesions, including coarctation of the aorta also suggests congenital origin.

Although there are few reports of intracranial aneurysm in members of the same family, their occurrence suggests a relationship that is more than fortuitous and lends further support to the concept of their congenital nature. Ullrich and Sugar16 reported intracranial aneurysms occurring in 4 family groups and referred to others described by Chambers et al.,3 Eck,5 and Aserjno et al.1 Familial aneurysm was suggested by O'Brien,12 who, at autopsy, found a middle cerebral artery aneurysm in a man whose twin brother had died suddenly from "intracranial hemorrhage or rupture of a brain abscess." Neither diagnosis, however, was proved clinically or at autopsy. Walton,17 in his comprehensive monograph, referred to 6 cases of intracranial bleeding in various families, but specific information was not given. Krayenbühl and Yasargil18 described the case of a man with a cerebro-vascular malformation, who had 2 brothers and a son with saccular cerebral aneurysms. Ross14 found a woman with a left parietal arteriovenous malformation and left middle cerebral artery aneurysm. Her mother had died of subarachnoid hemorrhage from a ruptured right internal carotid-posterior communicating artery aneurysm.

These examples suggest that hereditary factors may play a role. Dunger,4 in discussing the hereditary implications of polycystic disease of the kidney, refers to cerebro-vascular lesions occurring in several members of a family with this disease. Some probably had ruptured cerebral aneurysms, a relationship which is now well known. That genetic factors do have a specific role in some examples of familial aneurysm8 is shown by the occurrence of aneurysm in patients with the hereditary connective tissue disorders (Ehlers-Danlos syndrome and pseudo-xanthoma elasticum).

Because the correlation of autopsy findings in members of a family, one of whom has had an aneurysm, is obviously difficult, it is uncertain how common familial aneurysms may be. In those who died of causes other than subarachnoid hemorrhage, aneurysms may have been overlooked because they were not specifically sought. The finding of intracranial aneurysms at autopsy is notoriously related to the diligence of the search. The real incidence of aneurysms in more than one member of a family is therefore likely to be higher than is commonly believed.

Because of the rarity of detailed studies of familial aneurysms, we are reporting two family histories of aneurysms in siblings. In each family the aneurysms were strikingly similar in anatomical location and angiographic appearance. We have previously reported a brother-sister pair with Ehlers-Danlos syndrome and spontaneous carotid cavernous fistula.8

Case Reports

Case 1. D.D., a 28-year-old woman, was admitted to Buffalo General Hospital on May 1, 1938. Earlier that day she had been seized with intense right retro-orbital, then generalized, head pain. She vomited, had a poorly described convulsion, and lost consciousness. She had been taken to an outlying hospital where she had several attacks of extensor rigidity. Lumbar puncture revealed a bloody cerebrospinal fluid. She was then referred to this hospital.

Examination. She was rational, but restless. Her neck was supple. Babinski's sign was suggested bilaterally. Bilateral carotid angiograms on May 16 showed filling of both anterior cerebral arteries from the left internal carotid artery, but not from the right. No aneurysm was demonstrated. Right carotid angiogram made on May 21 (Fig. 1) demonstrated a saccular aneurysm 1 cm. in diameter at the trifurcation of the middle cerebral artery.

Operation. On May 28, through a right fronto-temporal craniotomy, an old subdural hematoma 1 cm. thick was evacuated, and the aneurysmal neck was clipped.

Postoperative Course. The patient had a postoperative left hemiparesis which was receding at the time of discharge from the hospital on June 17. Repeat right carotid angiography on July 30 showed no evidence of aneurysm. She remains well.

Case 2. R.W., 28-year-old brother of D.D., was ad-
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Fig. 1. Aneurysm at the trifurcation of the right middle cerebral artery in Case 1.

Fig. 2. Aneurysm at the trifurcation of the right middle cerebral artery in Case 2 (sibling of Case 1).