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Multiple Cerebral Aneurysms in Identical Twins

T. Hagen, K. Neidl, and U. Piepgras

Summary: We report the finding of multiple cerebral aneurysms in a pair of identical twins. One twin had subarachnoid hemorrhage and intracerebral hematoma; her sister had subarachnoid hemorrhage and intraventricular bleeding. Angiography in both cases showed multiple cerebral aneurysms. It is appropriate to recommend a cerebral angiographic examination of an asymptomatic twin after the other twin has suffered an aneurysmal subarachnoid hemorrhage.

Index terms: Aneurysm, cerebral; Familial conditions

The familial occurrence of cerebral aneurysms implies the presence of such aneurysms in two or more first- to third-degree family members. The highest association occurs among siblings. We present a pair of monozygotic twins with subarachnoid hemorrhage. Angiography, performed after subarachnoid hemorrhage, showed multiple cerebral aneurysms.

Case Reports

Case 1

A 39-year-old woman reported acute onset of severe headache and emesis in November 1990. Examination upon arrival at the hospital showed the patient to be somnolent (Hunt and Hess grade IV). No risk factors, such as smoking, hypertension, or other associated vascular disease, were known. Computed tomography (CT) revealed a large right-sided temporal intracerebral hematoma with mass effect and a right-sided hemorrhage. Four-vessel angiography showed a 7-mm right supraclinoid internal carotid artery aneurysm pointing posterolaterally (between the anterior choroidal artery and the posterior communicating artery). In addition, a 5-mm bilobular aneurysm projecting posteriorly and a 2-mm aneurysm pointing inferoposteriorly at the trifurcation of the right middle cerebral artery were found. The measurements were corrected for magnification. Arterial vasospasm was present in the middle cerebral artery (Fig 1).

After external ventricular drainage, the patient's level of consciousness improved slowly. On day 24, rehemorrhage occurred. The three aneurysms were clipped, and the patient's postoperative course was uncomplicated.

Case 2

The identical twin sister of the patient in case 1 was admitted to the hospital with severe headache and rapid deterioration of her level of consciousness in August 1992. As with her sister, no risk factors for vascular disease were known. A CT scan showed intraventricular and subarachnoid hemorrhage. Four-vessel cerebral angiography revealed a 5-mm aneurysm pointing posteriorly at the trifurcation of the right middle cerebral artery, a 4-mm right posterior communicating artery aneurysm projecting laterally, and a fusiform enlargement of the infraclinoid internal carotid artery (Fig 2). Additionally, a 3-mm left posterior communicating artery aneurysm pointing posteriorly was found. To prevent repeat hemorrhage, we performed an early clipping of the two right-sided aneurysms. Postoperatively, the patient's neurologic status deteriorated progressively. She died 3 days after admission and surgical clipping. An autopsy revealed the aneurysms and a general vasospasm, with multiple infarcted regions of the brain stem.

Discussion

Only a few cases of identical twins with intracranial aneurysms have been reported (Table). The highest rate of familial aneurysms seems to be in twins, but they do not necessarily have a higher frequency of aneurysmal occurrence than that of the general population (1). Thirty-two (6.7%) of 485 patients with subarachnoid hemorrhage have a first- to third-degree relative who had a subarachnoid hemorrhage. On the basis of chance alone, one would expect the frequency of such an occurrence to be 5.6% (2). Hence, familial aneurysms can be explained by accidental aggregations. However, familial aneurysms have to be distinguished from sporadic, nonfamilial aneurysms, because they show some specific differences. Like aneurysms in twins, they appear to rupture at an earlier age, 41.9 years for the eight reported cases and 42.3 years for familial aneurysms (3), in contrast to an average age of 51.4 years

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Fig 1. Case 1.

A, Right carotid angiogram, anteroposterior view, shows a 7-mm supraclinoid internal carotid artery aneurysm (*arrow*) and a 2-mm aneurysm pointing inferoposteriorly (*arrowhead*) at the trifurcation of the right middle cerebral artery.

B, Lateral view shows the internal carotid artery aneurysm between the anterior choroidal artery and the posterior communicating artery (*arrow*) and a 5-mm bilobular aneurysm projecting posteriorly (*arrowhead*). Vasospasm is present in the middle cerebral artery.

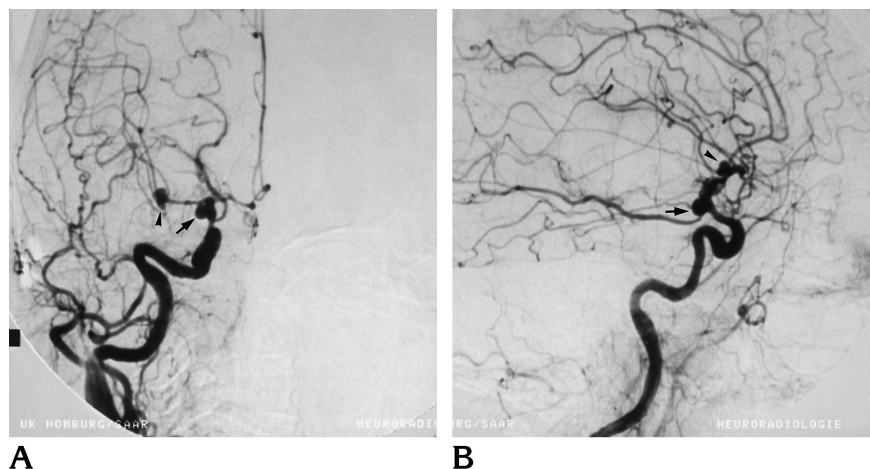
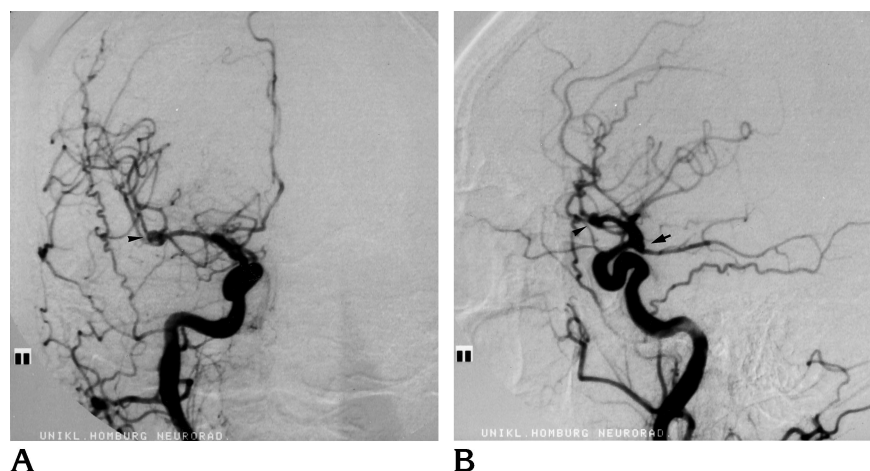


Fig 2. Case 2.

Cerebral angiograms in anteroposterior (A) and lateral (B) views show a 5-mm aneurysm pointing posteriorly (*arrowheads*) at the trifurcation of the right middle cerebral artery, a 4-mm right posterior communicating artery aneurysm (*arrows*), and a fusiform enlargement of the infraclinoid internal carotid artery.



for the rupture of sporadic nonfamilial aneurysms. The difference in age at presentation among the pairs of twins reported ranges from 2 to 8 years. Familial aneurysms seem to rupture at a smaller size and to occur less frequently in the anterior communicating artery (3).

In identical twins, aneurysms are located more frequently at the same site or at mirror sites than in randomly selected patients. Andrews (4) found that the frequency of occurrence at the same site was over twice the expected frequency. In a retrospective study of familial intracranial aneurysms, 15 of 38 pairs of siblings had at least one aneurysm in the ipsilateral vessel and 26 of 40 pairs had aneurysms at mirror sites (5). Among the sets of twins listed in the Table, aneurysms occurred in the same location in four pairs (6–8), including ours, and in mirror sites in two pairs (9, 10).

The findings of aneurysms in identical twins make a genetic predisposition probable. An au-

tosomal mode of inheritance is most likely (11); however, polygenic multifactorial transmission may be possible (3, 12, 13). Cerebral aneurysms have been reported in association with Ehlers-Danlos syndrome type IV (14, 15), Marfan syndrome (16, 17), pseudoxanthoma elasticum (18–20), Anderson-Fabry disease (21), polycystic kidney disease (22–25), and coarctation of the aorta (26, 27). Arterial hypertension may contribute to the development of aneurysms in the latter two diseases. Besides arterial hypertension, arteriosclerosis has been implicated as an acquired factor in the pathogenesis of cerebral aneurysms. Ostergaard et al (28) postulated an association of the *C3F* gene with arteriosclerotic vascular disease. This gene was suggested to be a risk factor for early aneurysmal rupture.

Collagen type III deficiency results in an abnormal extensibility of the arterial wall (29). Such deficiency has been reported in patients

Eight pairs of identical twins with cerebral aneurysms

Study	Year	Age, y/Sex	Signs and Symptoms	Site of Aneurysm
O'Brian (34)	1942	26/M	Sudden death	Unknown
		34/M	SAH, fatal	L MCA
Brisman and Abbassioun (33)	1971	30/F	SAH, aneurysms clipped	L and R MCA
		35/F	SAH, fatal	L and R ICA
Wilson and Cast (7)	1973	42/F	SAH, no surgical treatment	L MCA
		45/F	SAH, fatal	L MCA
Fairburn (9)	1973	44/F	SAH, ligation of ICA	R ICA
		46/F	SAH, no surgical treatment	L ICA
Schon and Marshall (6)	1984	39/M	SAH, fatal after clipping	ACoA
		43/M	SAH, fatal	ACoA
Weil et al (8)	1988	43/F	SAH, two aneurysms clipped	L and R ICA, L PCoA, basilarA
		43/F	Headaches, one aneurysm clipped	L and R ICA, L PCoA, basilarA
Parekh et al (10)	1992	36/F	Epilepsy, two aneurysms clipped	L MCA, L PICA
		37/F	SAH, fatal	R MCA
Present report	1997	39/F	SAH, three aneurysms clipped	R ICA, two in R MCA
		41/F	SAH, two aneurysms clipped	R ICA (fusiform), R PCoA, R MCA

Note.—ACoA indicates anterior communicating artery; basilarA, basilar artery; ICA, internal carotid artery; MCA, middle cerebral artery; PCoA, posterior communicating artery; PICA, posterior inferior cerebellar artery; and SAH, subarachnoid hemorrhage.

with sporadic aneurysms, but we found no abnormalities of collagen type III reported in patients with familial cerebral aneurysms.

Not only are the genetics and pathogenesis of intracerebral aneurysms controversial but also the screening of relatives in families with two or more members with subarachnoid hemorrhage. Ter Berg et al (11) devised a decision tree, taking into consideration the age of the patient, the risks of intraarterial digital subtraction angiography (IA-DSA) (30), and the postoperative morbidity related to the size of the aneurysm(s) (31). However, the risks of diagnostic procedures play only a minor part in the resulting gain for the patient. Use of magnetic resonance (MR) angiography as the screening method for calculations based on the decision tree and the assumption of a sensitivity of 100% would only result in a slight increase in the benefits. In conclusion, Ter Berg et al recommended screening of asymptomatic relatives only for those between the ages of 35 and 65 years (11). Clinical factors, like familial history or associated medical conditions, and morphologic parameters, such as the size, shape, and location of an aneurysm, are subjects of intense interest to those engaged in refining the characteristics of patients with unruptured aneurysms who are at risk for subarachnoid hemorrhage. The risk of hemorrhage has to be compared with the total risk of diagnostic screening and treatment of the aneurysm, such as surgery or endovascular intervention.

Less invasive imaging studies such as MR angiography or CT angiography may be used for the screening of relatives of patients with intracerebral aneurysms. Owing to its higher sensitivity, IA-DSA remains the definitive study for the diagnosis and preinterventional delineation of aneurysms.

Whether the risk of subarachnoid hemorrhage in an asymptomatic twin after the other twin has had an aneurysmal subarachnoid hemorrhage is increased or decreased, we believe that an angiographic examination should be offered to the asymptomatic twin. We found one pair of monozygotic twins who did not have identical intracranial aneurysms reported (32). In this case, one twin had aneurysmal subarachnoid hemorrhage and MR angiography and IA-DSA could not detect intracranial aneurysms in his asymptomatic brother. In the set of twins reported by Brismann and Abbassioun (33), one sister had normal angiographic findings after her twin sister died of a subarachnoid hemorrhage. However, 4 years later, cerebral angiography of the same vessels revealed bilateral middle cerebral artery aneurysms, which have been clipped. Thus, normal findings at cerebral angiography do not preclude later development or visualization of an aneurysm. The development of aneurysms in arteries that were previously reported to be normal is not unusual. MR angiography may be used as a tool to study the natural course in patients with risk factors for vascular disease.

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