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Subarachnoid haemorrhage in identical twins

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SUMMARY A pair of identical twins both of whom died of subarachnoid haemorrhage from ruptured anterior communicating artery aneurysms are reported. These twins are compared to the three other reported twins with ruptured cerebral aneurysms.

The lesion underlying a cerebral aneurysm is a defect in the medial coat of the artery usually at a point of bifurcation. Despite the fact that this defect is believed to be present from birth, the vast majority of subarachnoid haemorrhages due to the rupture of intracranial aneurysms are not associated with any identifiable genetic predisposing factors. However, families have been reported[1–4] in which multiple cases have occurred, suggesting the possibility that a genetic factor may play a part, at least in this minority group. This view is further supported by the well known association of certain rare hereditary disorders with an increased incidence of cerebral aneurysms; these include Ehlers-Danlos syndrome,[5] pseudo-xanthoma elasticum[6] and polycystic renal disease.[7 8] It has been suggested that the mechanism underlying such a predisposition to aneurysm formation is a deficiency of type 3 collagen.[9]

In this paper a pair of identical twins is reported who died of subarachnoid haemorrhage from identically sited aneurysms. There are three other twin pairs reported in the literature of which only two had proven cerebral aneurysms.

Case reports

A 39-year-old man was admitted to hospital in 1978 with acute onset of headache, nausea and photophobia. He had no focal neurological signs but uniformly bloodstained cerebrospinal fluid. Angiography demonstrated an anterior communicating artery aneurysm (fig A) which was successfully clipped. He made an uneventful recovery but collapsed and died suddenly on the seventeenth postoperative day. Post mortem examination only revealed subarachnoid blood in the skull base region.

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His 43-year-old identical twin brother was admitted to hospital in 1983 following the acute onset of headache, vomiting and rapid loss of consciousness. On examination he was comatose with neck stiffness and no response to painful stimuli. Fundoscopy revealed extensive subhyaloid haemorrhages. Over the subsequent six hours he lost all brain stem responses. A computed tomographic brain scan revealed extensive blood throughout the ventricular system. The patient died the following day. Post mortem examination revealed an anterior communicating artery aneurysm (fig B) in addition to subarachnoid blood.

The mother of these twins died aged 76 years of peripheral vascular disease and the father aged 65 years of gastric carcinoma. There were no other siblings and no history suggestive of subarachnoid haemorrhage in any other members of the family. Both patients were known to share the same O negative blood group.

Discussion

The details of the other three twin studies as well as this pair are summarized in the table. All cases presented before the age of 50. The age at presentation within each twin pair was remarkably similar, the difference ranging from 2–8 years. It has been suggested that the distribution of intracranial aneurysms differs between sporadic and familial cases.[13 14] In particular anterior communicating artery aneurysms were found twice as frequently in sporadic cases as compared to familial ones. This difference may however only be due to the relatively small number of familial cases.

Perhaps the most difficult clinical problem is whether it is justified to carry out angiography on the asymptomatic twin of a patient with a cerebral aneurysm. In the cases of Brisman *et al*,[12] the first sister initially had normal angiographic studies when she originally presented at the age of 30 with a subarachnoid haemorrhage. She was only reinvestigated after her twin sister died from a subarachnoid haemorrhage and on this occasion her two aneurysms were detected and successfully treated. In the

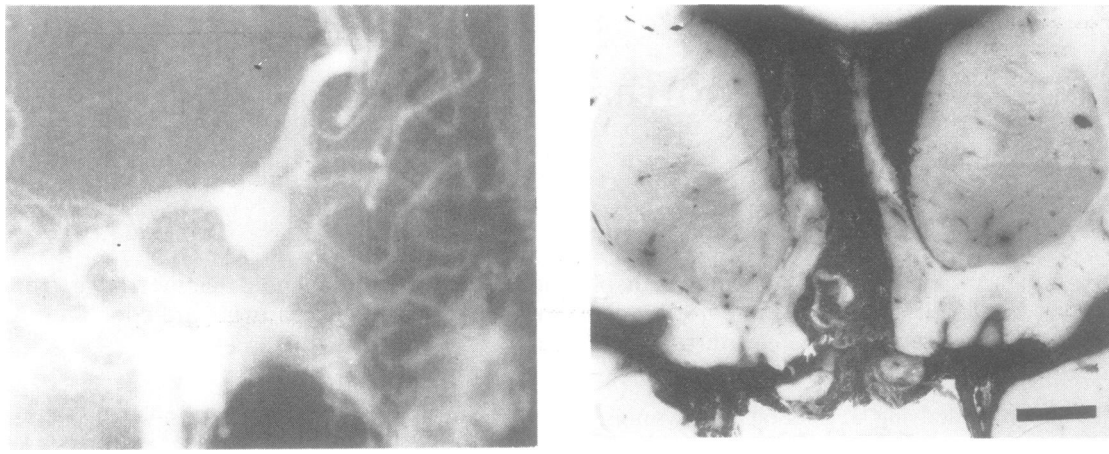

Fig *(A) Right carotid angiogram demonstrating a moderate sized anterior communicating artery aneurysm. (B) Coronal section through the hemispheres showing the anterior communicating artery aneurysm lying in front of the optic nerves surrounded by subarachnoid blood. Calibration bar is 1 cm.*

family reported by Fox *et al*,[15] after three siblings out of a sibship of 13 amongst which there were no twins, had presented with subarachnoid haemorrhage, the other 10 were offered angiography; of the eight who agreed to be investigated, three had aneurysms detected and treated with excellent results. However, there are at present no prospective data on the risks of asymptomatic twins developing subarachnoid haemorrhage after one twin has had such a bleed. Nevertheless, in view of these case reports there would appear to be reasonable justification for offering angiography to an asymptomatic twin. It is possible that with the further development of digital subtraction angiography such patients will be able to be investigated without significant risks.

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Table *Summary of information on 4 sets of Twins with subarachnoid haemorrhage*

Authors	*Age at Presentation*	*Sex*	*Clinical detail*	*Aneurysm site*
O'Brien JG[10]	26	Male	Sudden death of unproven cause	Not known
	34	Male	SAH, fatal outcome	Left MCA
Fairburn B[11]	44	Female	SAH, survived with left hemiparesis after right Carotid ligation	Right ICA (Supraclinoid)
	46	Female	SAH, No treatment, survived with expressive dysphasia	Left ICA (Supraclinoid)
Brisman R *et al*[12]	30	Female	SAH, Both aneurysms clipped. Survived with minimal deficit	Left MCA and Right MCA
	35	Female	SAH, Left hemiparesis Sudden death	Right and Left ICA
Schon F *et al*	39	Male	SAH, died 17 days post surgical clipping	ACA
	43	Male	SAH, sudden death	ACA

MCA—Middle cerebral artery. ICA—Internal carotid artery. ACA—Anterior communicating artery. SAH—Subarachnoid haemorrhage.

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