

Intracranial Dissecting Aneurysms in Childhood

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SUMMARY Children presenting after trauma with headache, seizures, hemiplegia and coma may have an intracranial dissecting aneurysm. Specific angiographic findings provide confirmation of this diagnosis. The dissection occurs subintimally and differs clinically and pathologically from dissecting aneurysms of extracranial arteries. The course in children beyond infancy is catastrophic, justifying consideration of potentially life saving surgical intervention.

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ARTERIAL OCCLUSIVE DISEASE accounts for approximately 20% of acquired hemiplegias in childhood.¹ Several vascular syndromes have now been defined. Intracranial dissecting aneurysms are rare in childhood — only sixteen cases fully reported.²⁻¹⁶ The peak incidence is in the third and fourth decades.¹⁷ The combined clinical and radiologic profile is relatively specific. Intracranial dissecting aneurysms should be added to the list of arterial occlusive syndromes of childhood. Antemortem recognition of this generally catastrophic entity may allow life saving intervention.

Case Report

In April 1978, an eight year old boy developed a sudden left hemiplegia which had been preceded by two days of right sided headache. There was no history of trauma. Family history was remarkable for a paternal great aunt with migraine. Laboratory studies including CBC, routine chemistries, ESR, ANA, LE prep, EKG, Echocardiogram, Chest X-ray, and IVP were unremarkable. CT scan showed a non-enhancing lucent area in the right basal ganglia. An arteriogram demonstrated a supraclinoid stenosis of the right internal carotid artery and an occlusion of the posterior parietal branch of the right middle cerebral artery. He recovered uneventfully. One month later only a minimal left central facial paresis and drift of the extended left arm were apparent on exam.

Studies of platelet functions several weeks after the ictus revealed hyperaggregability. He was treated with aspirin and Persantin. Six months later repeat determinations showed a further increase from the previous values and the medications were discontinued.

The patient had no further problems until June of 1979 when he had a generalized seizure. Thereafter, a dense right hemiplegia was apparent. Examination several hours later revealed an agitated, obtunded child with a normal habitus who showed no response to verbal stimuli, but occasionally opened his eyes spontaneously and responded with appropriate avoidance to painful stimuli. He was treated with decadron and an anticonvulsant. His course was complicated by marked hypertension (diastolic blood pressures ranging from 100-120) and hyperpyrexia (to 106). Three days after the ictus in association with an acute rise in blood pressure to 250/140, his pupils

became fixed and dilated. Treatment with mannitol, hyperventilation and antihypertensive agents were unsuccessful and he expired two days later.

A non contrast CT scan done on the evening of the ictus showed edema of the entire left hemisphere with compression of the lateral ventricle. A lumbar puncture performed that same evening revealed a normal opening pressure and cerebrospinal fluid profile.

Post Mortem Examination

Post mortem examination was limited to the brain. Both cerebral hemispheres were swollen, left greater than right, and there were signs of bilateral transtentorial uncal herniation. A large recent hemorrhagic infarct was present in the territory of the left middle cerebral artery. An old cystic infarct was found in the right basal ganglia.

Microscopic examination showed a dissecting aneurysm in the supraclinoid portion of the left internal carotid artery. The lesion extended into the middle cerebral artery and its main Sylvian branch. In transverse sections, the "aneurysm" consisted of a rounded mass of blood between the media and internal elastic lamina on one side of the artery (fig. 1A). The elastic membrane and intima were displaced by the mass resulting in marked narrowing of the lumen (fig. 1A). The hematoma began in the distal few millimeters of the internal carotid artery; no communication between the lumen and the blood in the vessel wall was found, even though several successive sections near the origin of the aneurysm were examined. In a section just proximal to the lesion, the vessel had a redundant fold of internal elastic lamina projecting into the lumen. Recent thrombus occluded the internal carotid and anterior cerebral arteries.

A dissecting aneurysm with histologic signs of healing involved the right internal carotid and middle cerebral arteries (fig. 1B). The lesion occupied a similar region between the internal elastic membrane and the media, but the lumen was not greatly narrowed. The blood in the wall communicated with the lumen in the distal few millimeters of the internal carotid artery. The inner layer of the vessel wall was discontinuous at this site, and the severed ends of the internal elastic lamina were coiled. A section of the artery just proximal to the lesion was normal except for mild focal fibrous thickening of the intima. In some areas of the right middle cerebral artery the dissecting aneurysm was surrounded by a thin layer of smooth muscle and an elastic membrane, clearly separate from the media and internal elastic lamina of the artery. Although this portion of the lesion resembled a second arterial

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lumen, it was not lined by endothelium. The right anterior cerebral artery did not have a dissecting aneurysm, but the elastic membrane was abnormally convoluted and small deposits of hemosiderin were

found between the elastica and media. The artery also had signs of a recanalized thrombus. Sections of posterior cerebral, basilar, and vertebral arteries were normal except for slight focal thickening of the intima and splitting of the internal elastic lamina.

Discussion

Although the number of patients with intracranial dissecting aneurysms is small, a rather consistent clinical history is apparent (table 1). Males predominate (10/16) in contrast to the equal sex incidence in adulthood. A prodromal and often localizing headache occurs for from hours to weeks before an acute hemiplegia develops. Trauma is often mentioned in the history, though it appears trivial for the most part. Seizures are common, both heralding the hemiplegia and occurring during the course of the illness. The course has generally been catastrophic, with coma and death supervening in days to a few weeks. The early deaths are generally due to massive cerebral edema with herniation.

The previously reported cases of Adelman² and of Chang³ had bilateral disease, but unlike the present case the interval between dissections was on the order of weeks, rather than months, and interval recovery was minimal. As the causes of multivessel disease in childhood are few, the clinical story along with a suggestive radiologic picture (see below) should alert the physician to the possibility of dissecting aneurysms, even though they are rare.

The three youngest patients aged three to twelve months^{11, 14, 16} were exceptional in that they survived the ictal event (the diagnosis being made years later at hemispherectomy for seizure control¹⁶ and at post mortem after death from another cause¹¹). The long survival of a fourth patient¹⁸ (not reported in detail) aged nine months corroborates the suggestion that infants fare considerably better than children 5–16 years. Therefore spontaneous cessation of dissections do occur.

Treatment with anticoagulants,⁷ corticosteroids,⁸ and decompressive craniotomy^{10, 13} proved unsuccessful. In Shillito's patient¹⁴ an arteriotomy was performed. Whether this was the decisive factor in the one year olds' survival is not clear, as the followup arteriogram showed reocclusion of the involved vessel and the development of collateral circulation. Neurosurgical reviews of predominantly adult patients with intracranial dissecting aneurysms suggest that microvascular surgery may be life saving.^{9, 17} A recent

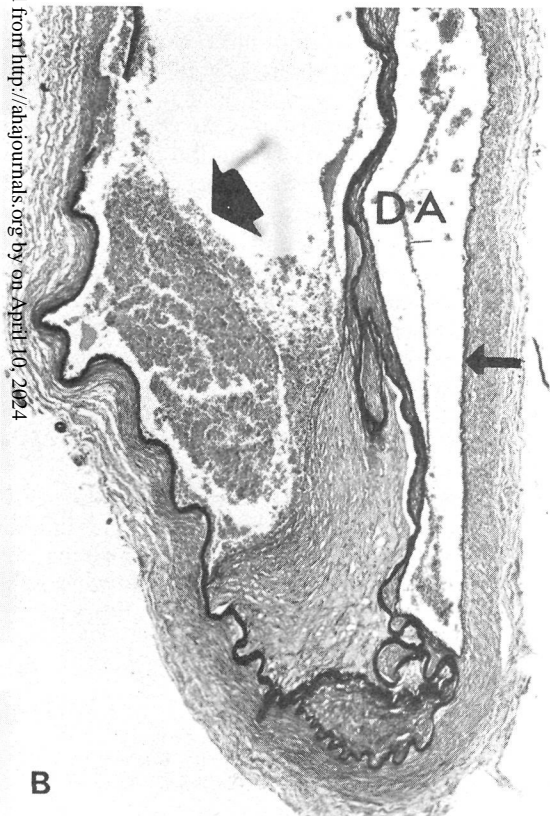
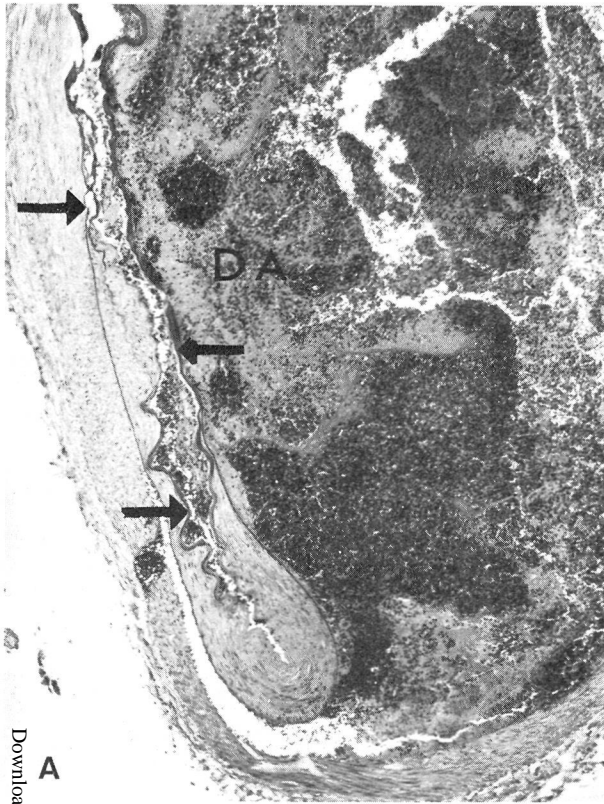


FIGURE 1. *Dissecting aneurysms.* 1A. Recent dissecting aneurysm (DA) in left internal carotid artery. The lesion consists of an intramural hematoma located between the internal elastic lamina and the media. The lumen of the artery (arrows) is markedly narrowed. H&E, $\times 220$. 1B. Old dissecting aneurysm (DA) in right middle cerebral artery. A thin layer of elastica (small arrow) surrounds the lesion. The lumen of the artery contains blood (large arrow). The internal elastic lamina is reduplicated in the vicinity of the lesion. Verhoeff stain, $\times 220$.

TABLE 1. *Clinical Findings in Sixteen Patients with Intracranial Dissecting Aneurysms*

History of trauma	8
Prodromal headache	7
Seizures	7
Coma and death	13
Acute < one week	8
Subacute several weeks	5

review of extracranial carotid dissecting aneurysms¹⁹ details several effective treatment modalities including surgery, anticoagulation, aspirin, corticosteroids, and rehabilitation, and reports a high survival rate from angiographically documented lesions in this location.

Radiology

A number of angiographic characteristics of dissecting aneurysms have been described^{20, 21} — the string sign, the rosette sign, and the pearl reaction, the latter two a result of total occlusion of the lumen with proximal distention. These radiographic signs are also seen in atherosclerotic vascular disease which is so rare in childhood that it can be disregarded in the differential diagnosis. The pathognomonic picture of the double lumen consisting of a narrow strip of greater contrast between broader bands of lesser contrast — the narrow strip being the incompletely occluded true lumen, the broader band the subintimal false lumen — has been occasionally reported.

Of the above signs only the string sign is specifically alluded to by name in the case reports of Hockberg⁷ and Johnson.⁹ The angiograms illustrated in several reports show a rosette sign. The most frequent finding (see table 2) was an occlusion of the middle and/or anterior cerebral arteries or their branches. Stenosis of the supraclinoid segment of the internal carotid, a recognized syndrome of arterial occlusive disease in childhood, was a prominent but never a singular finding in several of the patients. In no patient was the arteriogram normal. Pilz' patient¹² is most interesting in that the patient had both moya moya-supraclinoid stenosis with basal ganglia telangiectasia — and beading of many distal vessels, the latter correlating pathologically with multiple discontinuous dissecting aneurysms.

Pathology

The gross pathologic picture in those cases where death occurred acutely was one of hemispheric softening and swelling with herniation. Hemorrhagic infarction was the exception, perhaps because the dissecting

TABLE 2. *Angiographic Findings in Eleven Patients*

String sign	2
Probable rosette sign	4
Supraclinoid stenosis plus	3
Major vessel occlusion only	4
Major vessel occlusion plus	2

aneurysm usually caused complete rather than partial occlusion of the involved vessels.

Unlike dissecting aneurysms of the aorta which characteristically occur in the middle layers of smooth muscle in the media, all but Hayman's⁶ patient, involving the posterior circulation, were located between the internal elastic lamina and media, so called subintimal dissections. Only in Wisoff's patient¹⁵ was medial necrosis present, the cystic variety of which is the most common pathologic finding in extracranial carotid dissections. Associated systemic arterial dysplasia was rarely noted, but most post mortems were limited to the brain. Syphilitic arteritis and periarteritis nodosa have been reported in adults.¹⁷

In most patients absence, fraying, splitting, and reduplication of the internal elastica was mentioned. Whether these changes reflect congenital abnormalities of vessel wall, or whether they are within the range of normal, or even secondary to the dissection itself has been much debated. While intimal cushions, particularly at bifurcations and mild medial defects are generally accepted as normal, intimal tears are more frequent in patients with aneurysms.²²⁻²⁵ The most compelling argument for ascribing dissections to congenital wall defects is Wolman's finding¹⁶ of a minute saccular aneurysm at the origin of the dissection.

Trauma is an historical feature in half the patients, and Scott has suggested that the shearing forces and blood pressure fluctuations after trauma could open a minor congenital defect to dissection.²⁶

Similar pathologic changes of the internal elastic lamina have been described in Trichopoliodystrophy,²⁷ experimental transplacental infection with rubella,²⁸ moya moya,²⁴ and homocysteinuria.²⁹ A dissecting aneurysm involving both extra and intracranial carotid has been reported in a patient with homocysteinuria.³⁰

Chronic chemical injury to the endothelial wall leading to platelet thrombi formation have been held responsible for the occlusive vascular disease in homocystinuria.²⁹ Experimentally produced platelet aggregates have caused pathologic changes in vessel wall similar to that seen with dissecting aneurysms, possibly via the release of elastolytic enzymes.³¹ Thus the platelet hyperaggregability demonstrated in this patient may have played a role in his disease.

The child presenting possibly after trauma with headache, hemiplegia, seizures, and progressive obtundation, whose arteriogram demonstrates any of the previously mentioned signs should be suspected of having a dissecting aneurysm. In the face of the nearly uniform fatal outcome in children beyond infancy heroic treatment may be justified and life saving, but can only be instituted if the diagnosis is considered antemortem.

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