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Dolichoectasia of the Anterior Cerebral Arteries in an Adolescent

Stephen E. Doran, John P. Deveikis, and William F. Chandler

Summary: We report a case of bilateral dolichoectasia of the anterior cerebral arteries in a 14-year-old girl as documented by MR imaging and cerebral angiography. Concurrent thickening of the adjacent medial frontal lobes was found, suggesting either a neuronal migrational abnormality or secondary changes attributable to vascular insufficiency.

Index terms: Arteries, abnormalities and anomalies; Arteries, cerebral, anterior; Pediatric neuroradiology

Dolichoectasia (elongation and fusiform dilatation) of the intracranial arteries is a rare condition found almost exclusively in adults. We report a case of bilateral dolichoectasia of the anterior cerebral arteries in a 14-year-old girl as documented by magnetic resonance (MR) imaging and cerebral angiography.

Case Summary

The patient is a 14-year-old girl in good health other than having mild childhood asthma, a delayed onset of puberty, and short stature. She presented with a 1-year history of feelings of dissociation from her surroundings lasting several seconds. She denied any olfactory, gustatory, or visual hallucinations or any focal or generalized seizures. She had no history of drug abuse, head trauma, neurofibromatosis, connective tissue disorder, or homocystinuria. She was neurologically intact on physical examination.

Because her symptoms were suggestive of partial complex seizures, an MR image was made (Fig 1 A, B). This study showed prominent serpentine structures with flow-void in the region of the interhemispheric fissure, raising the suspicion of an arteriovenous fistula or arteriovenous malformation. These dilated vascular structures could be seen to distort the contour of the adjacent corpus callosum. There also was apparent thickening of the cortex of the medial frontal lobes, suggesting a neuronal migrational anomaly in addition to the vascular anomaly. Three-dimensional phase-contrast MR angiography suggested

that the dilated structures were the anterior cerebral arteries, but an arteriovenous fistula could not be excluded.

On the basis of the MR findings, a cerebral arteriogram was made (Fig 1 C, D). This revealed fusiform dilatation and ectasia of the precollosal and supracollosal segments of both anterior cerebral arteries, sparing the infracollosal segments. There was no evidence of arteriovenous shunting, and, in fact, there was some delay in washout of contrast from these ectatic vessels. The remaining intracranial vessels appeared entirely normal. A diagnosis of a giant aneurysm was excluded by the absence of characteristic findings on either MR or angiography.

Because an arteriovenous malformation was not found, no specific therapy for the vascular abnormality was recommended. A trial of carbamazepine was suggested to the patient to treat her probably partial complex seizures, but the patient's family refused.

Discussion

We report a case of dolichoectasia of both anterior cerebral arteries extending distally to involve the pericallosal and callosomarginal branches in a 14-year-old girl. Dolichoectasia is found almost exclusively in adults, with three reported pediatric cases (1–3). Two of these cases involved the basilar artery (1, 2); the third involved a single anterior cerebral artery (3).

Dolichoectasia of the cerebral vessels is characterized by elongation and fusiform dilatation (3, 4). Grossly, the vessels are soft and flexible, but the arterial wall is thickened. Histologically, the media demonstrates areas of smooth muscle atrophy and replacement with fibrous tissue showing hyaline changes, whereas the intima is concentrically thickened. Multiple defects in the internal elastic lamina also are found. Arteriosclerosis is frequently but not universally found (3, 5).

Dolichoectasia of the intracranial arteries is rare and most frequently affects the vertebro-

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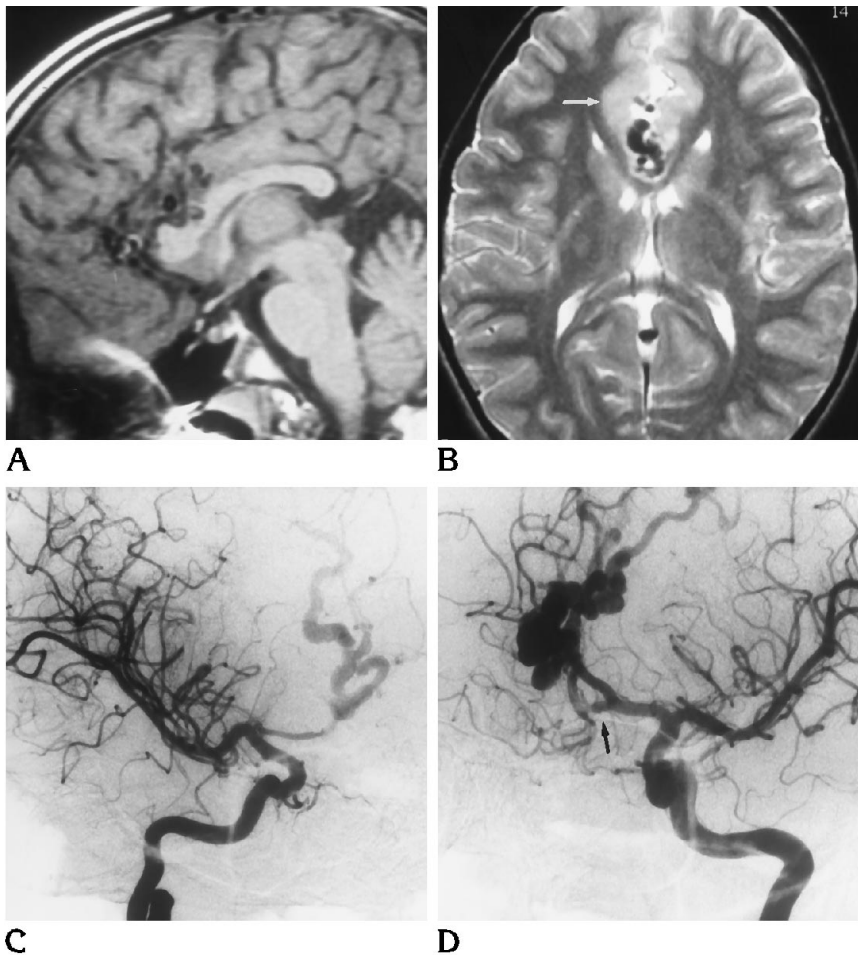


Fig 1. Sagittal T1-weighted (A) and axial T2-weighted (B) images reveal serpentine structures with flow-void in the interhemispheric fissure, suggesting arteriovenous malformation. Thickening of the medial frontal lobes on T2-weighted images (B) suggests a neuronal migrational anomaly (arrow).

Oblique views of selective right (C) and left (D) internal carotid artery arteriograms reveal fusiform dilatation of the precollosal and supracollosal segments of both anterior cerebral arteries as well as a patent anterior communicating artery that fills from the left (arrow) (B), eliminating the possibility of an azygous anterior cerebral artery. No arteriovenous shunting is demonstrated.

basilar system followed by the internal carotid artery. Of approximately 40 000 carotid and 10 000 vertebral angiograms performed at the National Hospital, Queen Square, London, between 1959 and 1980, 31 cases of ectasia of the vertebrobasilar or carotid system were retrospectively identified (6). Fusiform dilatation and elongation of the branches of the carotid artery are even less common (7).

The clinical picture of dolichoectasia of the intracranial arteries is varied and is usually attributable to the mass effect of the dilated vessels. Cranial nerve palsies (8, 9), chiasmal compression (9), hydrocephalus (9), trigeminal neuralgia (10), hemifacial spasm (11), nystagmus (12), ataxia (9), and dementia (9) all have been reported in association with dolichoectasia of the vertebrobasilar system. Dolichoectasia of the anterior circulation has been associated with seizure (4, 7), headache (7), visual field defects (13), and dementia with hydrocephalus (14).

Both congenital and acquired causes have been suggested for this condition. Support for

an acquired cause lies in the pathologic dilatation of the basilar artery found in elderly patients with extensive and advanced atherosclerosis (5). In addition, hypertension, diabetes mellitus, hyperlipidemia, coronary artery disease, and smoking have been associated with ectasia of the basilar artery (6). Evidence for a congenital origin of dolichoectasia lies in the occurrence of the condition in children and young adults in the absence of other potential predisposing health problems (1-3, 7). In our case, the thickened cortex of the medial frontal lobes suggests a migrational abnormality, which would provide additional evidence for a developmental cause. Other possible causes for this cortical thickening would be chronic changes attributable to vascular insufficiency or an in utero injury resulting in polymicrogyria. In reviewing their own cases and those reported in the literature, Araki et al concluded that there are two distinct causes of arterial ectasia, namely, arteriosclerosis and congenital malformation (15). In the arteriosclerosis group, ectasia appears in per-

sons generally over 40 years of age, more frequently male, and involves the proximal vertebrobasilar and carotid arteries. In the congenital group, affected persons are under 40 years of age of either sex and more frequently have involvement of distal branches of cerebral arteries.

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