Cerebral arterial dolichoectasia with seizure

Case report

JOSEPH R. THOMPSON, M.D., PHILIP R. WEINSTEIN, M.D., AND CHARLES R. SIMMONS, M.D.
Loma Linda University Medical Center, Loma Linda, California

Cerebral angiography, performed after a seizure in a patient with a life-long history of typical hemiplegic migraine, disclosed markedly dolichoectatic anterior and middle cerebral arteries. No abnormality of the adjacent capillary or venous structures was present. A positive brain scan was attributed to ischemia induced by vaso-spasm rather than to the corresponding large tortuous anterior and middle cerebral arteries. There were no permanent sequelae and the patient has been free of seizures on Dilantin and phenobarbital over a 3-year follow-up period. Angiographic demonstration or description of a similar ectatic set of anterior and middle cerebral arteries could not be found in the literature. The concurrence of seizures and hemiplegic migraine adds to the peculiarity of this case.

KEY WORDS  *

Cerebral arteries  
Dolichoectasia  
Ectasia  
Migraine  
Seizures

Arterial dolichoectasia (elongation-distension) is an uncommon condition that may involve intracranial arteries, but rarely causes gigantic enlargement. When severe enlargement does occur, it involves the arteries at the base of the brain. This entity usually presents symptoms like those of an intracranial tumor. Our search of the literature has not revealed a previous case involving cerebral arteries with the exclusion of the carotid or basilar arteries. The occurrence of migraine headache complicated by hemiplegia and the presentation with a seizure adds interest to this unusual case.

Case Report

This 39-year-old right-handed man was admitted for neurodiagnostic evaluation 1 month after his first grand mal seizure. He had not slept for 24 hours when he developed his typical prodromata of migraine and took ergotamine tartrate (Cafergot) which brought relief. About 30 minutes later he experienced nausea and dizziness which progressed to loss of consciousness and generalized seizure.

The patient had a history of two or three typical episodes of hemiplegic migraine per year since the age of 11 years. The attacks
began with right arm weakness and progressed to the face and palate causing speech difficulty. Within 10 minutes, retro-orbital tension and burning pain occurred followed by a severe bilateral, pounding headache that was disabling for several hours. He had not been evaluated for his migraine previously and had been placed on medications only recently. He had taken chlorpheniramine maleate (Chlortrimeton) and Cafergot at the onset of migraine prodromata and had twice aborted an attack.

Similar attacks of hemiplegic migraine were experienced by his mother throughout her life but no other incidence of migraine or seizure was present in the family. Other medical history, and physical and neurological examinations were negative. There were no cranial bruits. Skull x-ray films and electroencephalogram were normal. Lumbar puncture did not suggest subarachnoid hemorrhage and cerebrospinal fluid examination was normal. Brain scan showed diffuse, but definitely increased activity in the left frontal lobe with most prominence in the anterior cerebral distribution.

Bilateral common carotid and left vertebral angiograms were performed (Fig. 1) because of the recent seizure and positive brain scan. Right carotid and vertebrobasilar systems were completely normal including both posterior cerebral arteries. A diffuse luminal enlargement of profound degree began at the left internal carotid artery bifurcation and involved selectively the entire anterior cerebral artery and most branches of the middle cerebral artery (Fig. 2). No unusual capillary staining was present and the cortical and deep venous configuration of the left cerebral hemisphere was completely normal. No mass effect was present.

The patient has remained free of seizures on diphenylhydantoin (Dilantin) and phenobarbital during the three years of follow-up.
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He has remained on active duty in the Air Force.

Discussion

Although the angiographic findings in this case suggest a vascular malformation, a purely arterial malformation has never been verified angiographically.\(^1,2,6,7\) If, as Bergstrand, et al.,\(^1\) claimed, pure arterial angiomas do not exist, the dilated cerebral arteries in our patient would have to have resulted from some acquired process. The appearance most closely resembles that of arterial dolichoectasia, although the unilateral, luminal uniformity, and cerebral hemispheric location are atypical. Patients with dolichoectasia have not been reported to present with seizures but characteristically have shown cranial nerve deficits,\(^8,9\) and occasionally hydrocephalus,\(^9\) findings which suggest the presence of an underlying intracranial tumor.

The cause of dolichoectasia has not been established. Histological changes occur in the elastic tissue and the muscle fibers of the media. Arteriosclerosis when present is believed to be incidental. Examples of the entity are reported in children\(^8\) and are unassociated with arteriosclerosis. Because other diseases may cause ectatic arterial change, syphilis, arteriosclerosis, Ehlers-Danlos syndrome, pseudoxanthoma elasticum, and nonsyphilitic vasculitis should be considered in the gamut of possibilities.

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References


Address reprint requests to: Joseph R. Thompson, M.D., Department of Radiology, Loma Linda University Medical Center, Loma Linda, California 92354.