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Computed Tomographic Findings in Cerebral Arterial Ectasia

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The computed tomographic findings in 15 patients with angiographically documented cerebral arterial ectasia are reported. In addition to demonstrating the majority of pathologic arterial segments, computed tomography also documents the presence and extent of associated intracranial abnormalities. In this study these included cerebral atrophy (10 patients), infarction (three patients), and subarachnoid hemorrhage (one patient).

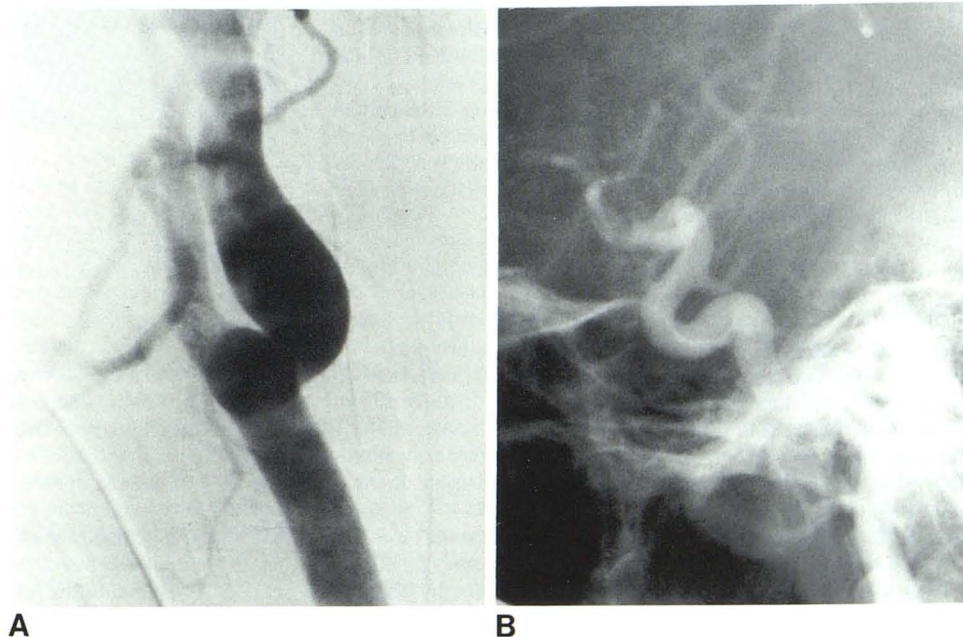
Cerebral arterial ectasia is an unusual arteriopathy. Its diagnosis is based on the radiographic findings of pathologic elongation, dilatation, and tortuosity of the arteries comprising the circle of Willis. The intracranial internal carotid arteries and vertebrobasilar system are most frequently involved, followed by the proximal segments of both the middle and anterior cerebral arteries [1, 2]. Most patients present with either cranial nerve palsies secondary to compression by the tortuous vessels, or with symptoms of cerebral ischemia [1, 3-5].

Until recently, the diagnosis of cerebral arterial ectasia was established by angiography or, occasionally, pneumoencephalography. With the introduction of computed tomography (CT), it has become possible to noninvasively identify and characterize this vascular disorder and its associated intracranial complications. This report describes the CT findings in 15 patients with angiographically documented cerebral arterial ectasia.

Materials and Methods

Between April 1980 and June 1982, 512 cerebral arteriograms were obtained at University Hospital, University of Kentucky. Based on the arteriographic findings of excessive tortuosity and elongation of the intradural portions of the internal carotid and basilar arteries, the diagnosis of cerebral arterial ectasia was established in 14 patients. The other patient in the series was evaluated at the University of Cincinnati Medical Center. Most of the patients in this

Fig. 1.—Case 3, 10-year-old boy with pulsatile neck mass. Right common carotid artery angiography. **A**, Cervical carotid system is markedly enlarged and tortuous. **B**, Intracranial carotid artery, lateral view. Intrapetrous, cavernous, and intradural parts of right internal carotid artery are dilated to almost twice diameter of normal contralateral internal carotid artery (not shown). No evidence of luminal irregularity to suggest atherosclerosis.



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TABLE 1: Clinical and CT Findings in Angiographically Documented Cerebral Arterial Ectasia

Case No. (Age, Gender)	Signs and Symptoms	Abnormal Arteries	Arterial Calcification	Ventriculomegaly
1 (58, M)	Ischemia	BA, ICA	Yes	...
2 (68, M)	Ischemia	BA	Yes	Moderate, atrophic
3 (10, M)	Neck mass	ICA-b	No	...
4 (87, M)	Ischemia	ICA-L	Yes	Mild, atrophic
5 (60, M)	Ischemia	ICA-R	Yes	Moderate, obstructive
6 (39, M)	Ischemia	BA, ICA-b, ACA	No	Mild, atrophic
7 (73, F)	Ischemia	BA, ICA-b	Yes	Mild, atrophic
8 (78, F)	Ischemia	ICA-b	No	Mild, atrophic
9 (77, M)	Ischemia	ICA-b	Yes	Mild, atrophic
10 (68, F)	Tic douloureux	BA, ICA-L	Yes	Moderate, atrophic
11 (47, F)	Ischemia	ICA-b, MCA	No	...
12 (40, M)	Subarachnoid hemorrhage	ICA-b, MCA-R, ACA-b	Yes	Moderate, communicating
13 (64, M)	Ischemia	BA	Yes	...
14 (66, M)	Bilateral optic atrophy	BA, ICA-L, MCA-L, ACA-L	Yes	...
15 (68, M)	Ischemia	ICA-b	Yes	Mild, atrophic

Note.—Infarction was demonstrated in three cases: cases 4 and 9, left parietal lobe; case 5, cerebellum. BA = basilar artery; ICA = internal carotid artery; MCA = middle cerebral artery; ACA = anterior cerebral artery; b = bilateral; R = right, L = left.

series (11 cases) had angiography performed because of signs and symptoms of cerebral ischemia. Subarachnoid hemorrhage, a pulsatile neck mass, the sudden onset of bilateral visual loss, and tic douloureux were the indications for angiography in the other four individuals.

Except for a 10-year-old boy who was evaluated because of pulsatile neck mass (fig. 1), all patients were at least 39 years of age at the time of presentation (range 10–87 years, average 61 years). The series included 11 men and four women. Twelve of the 15 patients were hypertensive by history or physical examination at the time of CT scanning.

Noncontrast axial cranial CT scans were obtained before contrast administration in all but one patient. All patients underwent cranial CT following a bolus intravenous injection of 42 g of iodine. Scanning was performed in the axial position only, utilizing a scan angle of 25° from Reid base line with either 8 mm or 10 mm collimation.

All patients underwent transfemoral bilateral cerebral angiography using Seldinger technique. Arch and vertebral angiograms were obtained as clinically indicated.

Results

The clinical and radiographic details of this series are presented in table 1. The diagnosis of cerebral arterial ectasia was based on the angiographic findings of arterial elongation, dilatation, and excessive tortuosity with or without superimposed atherosclerotic disease. The right internal carotid artery was involved most frequently (11 patients), followed by the left internal carotid artery (nine patients), and the basilar artery (six patients). Pathologic changes involving the middle cerebral and the anterior cerebral arteries were identified in five cases each. In 11 of the 15 patients multiple vessel involvement was identified. Angiography also revealed the presence of a clinically silent anterior choroidal artery aneurysm.

The unenhanced CT scans demonstrated evidence of cerebral arterial ectasia in 12 of 14 patients. The ectatic arteries, located predominantly in the suprasellar and interpeduncular cisterns, appeared as serpiginous, tubular structures on the unenhanced scan. Prominent vessels were occasionally noted in the sylvian and inter-

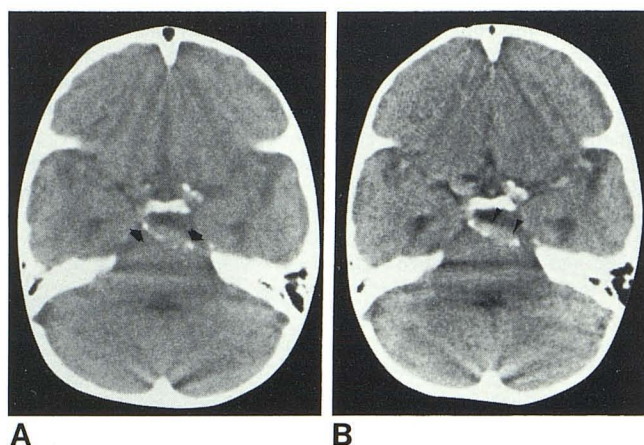


Fig. 2.—Case 7, 73-year-old woman with vertebrobasilar ischemic episodes. **A**, Unenhanced CT scan at level of dorsum sellae. Hyperdense tubular structure coursing across midpons. Margins are moderately well defined by linear calcifications (arrows). **B**, Enhanced scan, same level. Central part of calcified mass enhances homogeneously, confirming identity as basilar artery (arrowheads).

hemispheric cisterns as well. Vascular calcifications were identified on the noncontrast scans in 10 patients (fig. 2). The unenhanced scan of the 10-year-old boy was normal.

Ventricular enlargement was diagnosed on the basis of the CT scan in 11 patients. In nine of these cerebral atrophy was manifest by the prominence of the basal cisterns and cortical subarachnoid spaces. The atrophic changes were graded as severe in two patients, moderate in three, and mild in the other six patients. In one case enlargement of the lateral and third ventricle associated with nonvisualization of the fourth ventricle suggested the diagnosis of obstruction at the level of the aqueduct secondary to compression by an enlarged basilar artery. In the final case ventriculomegaly was attributed to recurrent subarachnoid hemorrhage resulting in communicating hydrocephalus.

The contrast-enhanced CT scans demonstrated dense, sharply defined, homogenous intraluminal enhancement in all 15 patients,

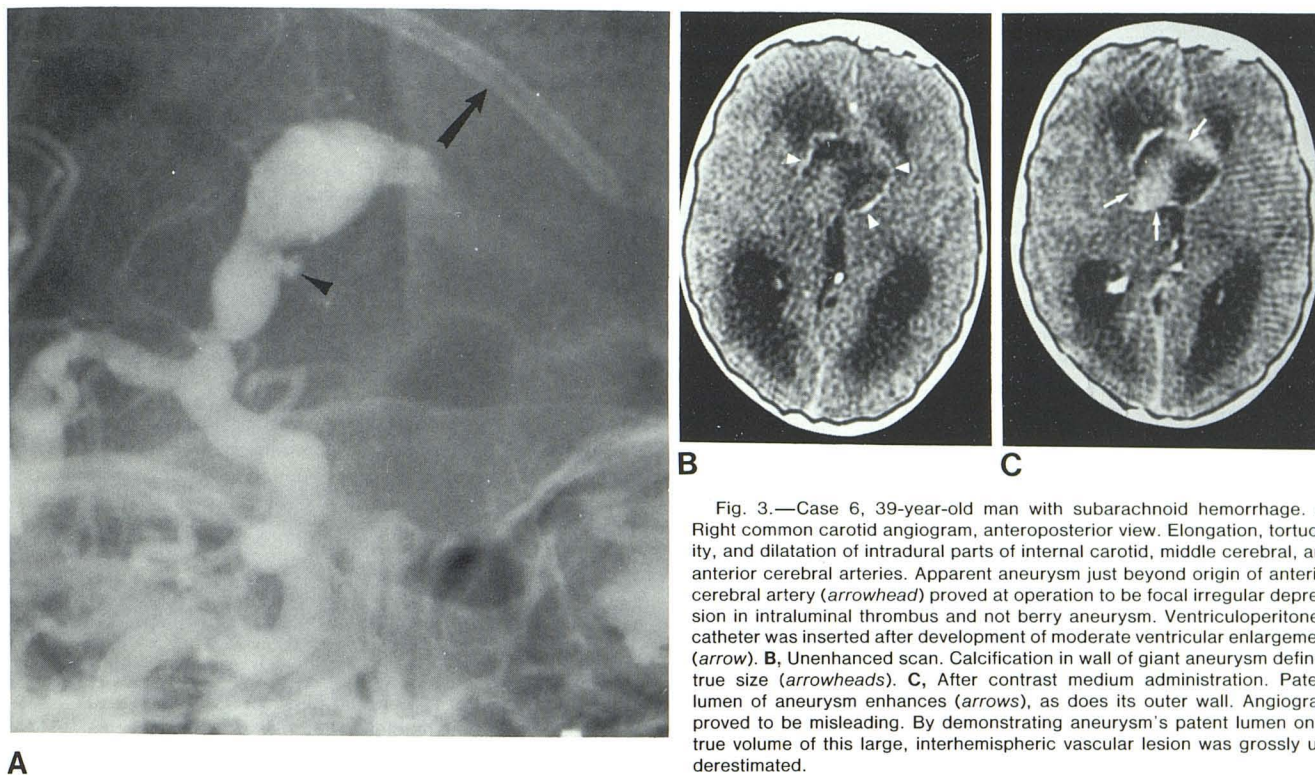


Fig. 3.—Case 6, 39-year-old man with subarachnoid hemorrhage. **A**, Right common carotid angiogram, anteroposterior view. Elongation, tortuosity, and dilatation of intradural parts of internal carotid, middle cerebral, and anterior cerebral arteries. Apparent aneurysm just beyond origin of anterior cerebral artery (arrowhead) proved at operation to be focal irregular depression in intraluminal thrombus and not berry aneurysm. Ventriculoperitoneal catheter was inserted after development of moderate ventricular enlargement (arrow). **B**, Unenhanced scan. Calcification in wall of giant aneurysm defines true size (arrowheads). **C**, After contrast medium administration. Patent lumen of aneurysm enhances (arrows), as does its outer wall. Angiogram proved to be misleading. By demonstrating aneurysm's patent lumen only, true volume of this large, interhemispheric vascular lesion was grossly underestimated.

corresponding to the abnormally dilated arteries at the base of the brain. The computed tomographic scan failed to demonstrate convincingly arteriographically proven changes involving the anterior cerebral artery in one case and dilatation of the middle cerebral artery in two other patients. In all three of these cases, however, the scans revealed excessive dilatation of the basilar and of at least one carotid artery.

CT demonstrated the presence of three infarcts in three different patients. Two of these were located in the middle cerebral artery distribution and one was in the cerebellum. The contrast-enhanced scans demonstrated cortical enhancement in one parietal lobe infarct, confirming its acute nature. This patient had cerebral arterial ectasia in association with atherosclerosis of the ipsilateral internal carotid artery. Vertebral angiography demonstrated marked ectasia of the basilar artery with evidence of intraluminal thrombus in the patient who presented with cerebellar infarction.

CT was superior to angiography in the evaluation of the individual who presented with recurrent subarachnoid hemorrhage. While the arteriogram did demonstrate pathologic changes involving the right carotid, anterior, and middle cerebral arteries, it did not reveal the true nature or extent of the giant fusiform aneurysm involving the A₁ segment of the anterior cerebral artery on the right. The CT scan displayed not only the patent lumen of the aneurysm, but also the presence of mural calcification and the extent of mural thrombus formation (fig. 3). The actual size and resulting mass effect of the giant aneurysm was convincingly depicted by CT scan alone.

Discussion

Vascular dysplasias have been associated with a variety of familial, hereditary, and connective tissue disorders including Marfan

syndrome, Ehler-Danlos syndrome, pseudoxanthoma elasticum, and Menkes syndrome [1, 6, 7]. While a review of the literature concerning cerebral arterial ectasia does not reveal a consensus regarding its etiology, it is likely that this arteriopathy probably results from extensive deficiencies involving the muscularis and the internal elastic lamina [1, 8–10]. Cerebral arterial ectasia has been reported in the very young, including a patient in the first year of life, suggesting that the pathologic deficiencies in the arterial wall are, in part, congenital in nature [8, 9]. The diffuse arterial manifestations of this disease was emphasized by Sacks and Lindenberg [8], who reported a high incidence of coexistent abnormalities of the noncerebral circulation. In a series of 34 pathologically proven patients with cerebral dolichoectasia, 12 patients had documented aortic aneurysms, seven of whom died from aortic rupture [9].

While cerebral arterial ectasia is probably secondary to congenital defects in the arterial wall, symptoms do not usually develop until the patient reaches middle or old age. In those individuals, the arteries dilate and elongate under the prolonged stress of often elevated systemic blood pressure and superimposed atherosclerotic disease. Arterial compression of adjacent neural structures is one of the more common causes of symptoms and neurologic deficits in these patients [1–5, 8–11]. While involvement of all the cranial nerves has been reported, paresis of the oculomotor nerve is the most common [3]. Cerebral ischemia, especially in the vertebrobasilar territory, has also been frequently reported in patients with cerebral arterial ectasia [1–4, 10, 11]. Embolization from thrombus within the dilated vessels appears to be the likely cause of the ischemic episodes. Luminal encroachment by clot and atheroma with impaired flow distally may also account for diminished perfusion [1, 9]. Subarachnoid hemorrhage and intracerebral hematoma occur infrequently in this group of patients [12, 13]. The dilated vessels are probably not liable to rupture because of the

gross thickening of the arterial wall secondary to hyaline degeneration of the muscular layers and sclerosis of the intima [1, 2].

The major cerebral vessels can be readily visualized with CT scans as they course through the basal subarachnoid cisterns. Serpiginous tubular configurations and the presence of mural calcifications help to differentiate these pathologic vessels from neoplasm on the CT scan. The contrast-enhanced CT appearance of dolichoectasia is almost pathognomonic [10, 11, 14]. Central arteriovenous malformations and giant fusiform aneurysms are the only entities that may be confused with cerebral arterial ectasia by CT scanning.

CT scanning successfully identified pathologic changes in all of our cases, although angiography sometimes revealed more extensive disease. Although the need for preoperative angiography remains undeniable, CT scanning, both before and especially after contrast infusion, is sufficiently diagnostic of cerebral arterial ectasia to preclude the need for more invasive studies in the patients for whom conservative therapy is best.

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