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MR Angiography of Cervical Fibromuscular Dysplasia

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Summary: The authors present examples of cephalic fibromuscular dysplasia (involving five vessels in two patients) using 2-D time-of-flight MR angiography and conventional angiography. Artifacts that can mimic the MR appearance of fibromuscular dysplasia will tend to decrease the sensitivity and specificity of MRA in its detection.

Index terms: Arteries, carotid; Arteries, vertebral; Magnetic resonance angiography (MRA)

Fibromuscular dysplasia (FMD) is an uncommon arteriopathy of unknown etiology that affects small- and medium-sized arteries. Although the renal arteries are most commonly involved, the cervical internal carotid and vertebral arteries can also be affected; the intracranial arteries are rarely involved. FMD is often an incidental discovery during cerebral angiography; however, patients can also present with focal neurologic deficits. The angiographic appearance of the lesion is usually characteristic, with alternating areas of concentric narrowing and dilatation of the affected vessel, often described as having a "string of beads" appearance.

Conventional magnetic resonance (MR) and MR angiography (MRA) were performed in two patients with five involved cervical vessels allowing comparison of the two techniques in visualization of the lesions. The details of the two-dimensional time-of-flight MRA technique utilized have been described elsewhere (1). The studies were done on a 1.5-T MR imaging system. The imaging time for the MRA portion of the examination was 12 min 40 sec.

Patients

A 49-year-old woman (patient 1) presented with a complaint of recurrent right amaurosis fugax of a few minutes' duration. A right carotid bruit was present on examination. The patient was mildly hypertensive. MR examination of

the brain was unremarkable. Antiplatelet therapy resulted in resolution of visual symptoms.

A 62-year-old woman (patient 2) presented with recurrent episodes of left-hand numbness and of right amaurosis fugax, in each case lasting a few minutes. Bilateral carotid bruits were present. The patient was not hypertensive. MR of the brain was unremarkable. The patient ultimately underwent a right superficial temporal artery to middle cerebral artery bypass, with improvement of symptoms.

In Figure 1, lateral views of the right carotid artery from the conventional angiogram and MRA examinations of patient 1 are compared. In the conventional image, segmental concentric dilatations of the middle portion of the cervical internal carotid artery are seen in an appearance characteristic of FMD. The MRA examination demonstrates areas of apparent dilatation alternating with areas of signal loss. Signal intensity is regained more distally, where the caliber of the vessel returns to normal.

Figure 2 compares right and left carotid and vertebral arteries in patient 2. Images from a lateral conventional angiogram are compared to the MRA study in anteroposterior and off lateral projections chosen to best visualize the affected vessels. In Figure 2A, a long segment of mildly irregular concentric narrowing is present in the proximal right cervical internal carotid artery, followed by several prominent segmental dilatations. The internal carotid artery bulb and the petrous internal carotid artery are unaffected. In Figure 2B, the right vertebral artery demonstrates marked focal outpouching at the C1-C2 level. The MRA in Figures 2C and 2D also shows the narrowing of the proximal internal carotid artery. In the distal cervical portion of the vessel, segments of signal dropout alternate with areas of more normal signal intensity. In the vertebral artery, focal dilatation similar in configuration to the findings on conventional angiography is visualized. Signal dropout is present along the posterior aspect of the carotid bulb on the MRA because of nonlaminar recirculating flow commonly present in this area (2).

Similar but less dramatic changes are present in Figure 2E, where proximal narrowing of the left internal carotid artery is followed by segmental dilatations. These areas are also visualized on MRA (Fig. 2G) as regions of relatively decreased signal intensity and narrowing alternating with more normal appearing segments. Irregularity of the ver-

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Fig. 1. Conventional (A) and MR (B) angiograms of the right carotid artery in the lateral projection in a patient with recurrent right amaurosis fugax. The segmental dilatations seen in the midcervical internal carotid artery in the conventional study are characteristic of FMD. Changes seen in the corresponding location on the MRA include regions of apparent luminal narrowing and signal loss. For the MRA, TE = 10 msec.

tebral artery at the C1-C2 levels (Fig. 2F) is seen as a subtle irregularity on the MRA.

Discussion

An extensive review of cervical FMD has been published by Heaton (3). The cervical vessels, particularly the internal carotid arteries, are the second most common site of involvement after the renal arteries. Nearly 90% of patients with cervical FMD are women, most commonly in the fourth or fifth decade; FMD is very uncommon in children.

Three histologic types known as the medial, intimal, and periarterial or subadventitial forms are described, with the medial form found in 90% of cases. The etiology is unknown, although an autosomal dominant mode of inheritance has been described for certain cases (4).

Although intracranial involvement of FMD is rare, some investigators report a 20% to 50% incidence of intracranial aneurysms in patients with cervical FMD (3). Other less commonly associated pathologies include spontaneous carotid

artery dissection, carotid aneurysm formation, and spontaneous arteriovenous fistulas (5). Clinically, carotid bruits are present in 70% to 100% of patients with cervical FMD and are sometimes audible to the patient. Patients in whom FMD is demonstrated angiographically often present with focal neurologic complaints (6), although a causal relationship has been established only in a minority of patients (3).

Three angiographic patterns are associated with cervical FMD (7), the most common of which consists of segments of constriction alternating with normal appearing or dilated segments, present in approximately 80% of cases. This pattern is usually associated with the medial histologic type, and the appearance is considered to be pathognomonic, although angiographic standing waves and circular spastic contractions can present a similar pattern. A smooth concentric tubular lesion is seen in less than 10% of cases, associated with any histologic type. A similar pattern is also associated with other entities including Takayasu arteritis, arterial hypoplasia, and catheter-induced spasm. A third uncommon appearance, described as atypical FMD, consists of eccentric outpouchings that may progress to frank aneurysm formation. Differential diagnosis for this appearance includes atherosclerotic aneurysm and traumatic pseudoaneurysm.

Cervical FMD is also characteristic in location (3). Lesions are present in the internal carotid artery in 95% of cases. In about one fourth of cases, the vertebral artery is affected. The C2 level is the most common location in both the carotid and vertebral arteries. The origins of the arteries are uncommonly affected, which is helpful in differentiating these lesions from atherosclerosis.

Although a single description of the sonographic appearance of carotid FMD using gray scale ultrasound has been reported (8), the modality is not widely used because of the inaccessible location of most carotid and essentially all vertebral FMD.

Two-dimensional time-of-flight MRA is a useful, noninvasive modality for the evaluation of narrowing of the carotid artery bifurcation (2, 9). The cervical internal carotid and vertebral arteries are clearly defined in a cervical MRA examination. In the five affected vessels in two patients in this report, changes in signal intensity associated with FMD are present that correlate in location with the characteristic changes of FMD seen on the conventional angiograms. The nature of the MRA

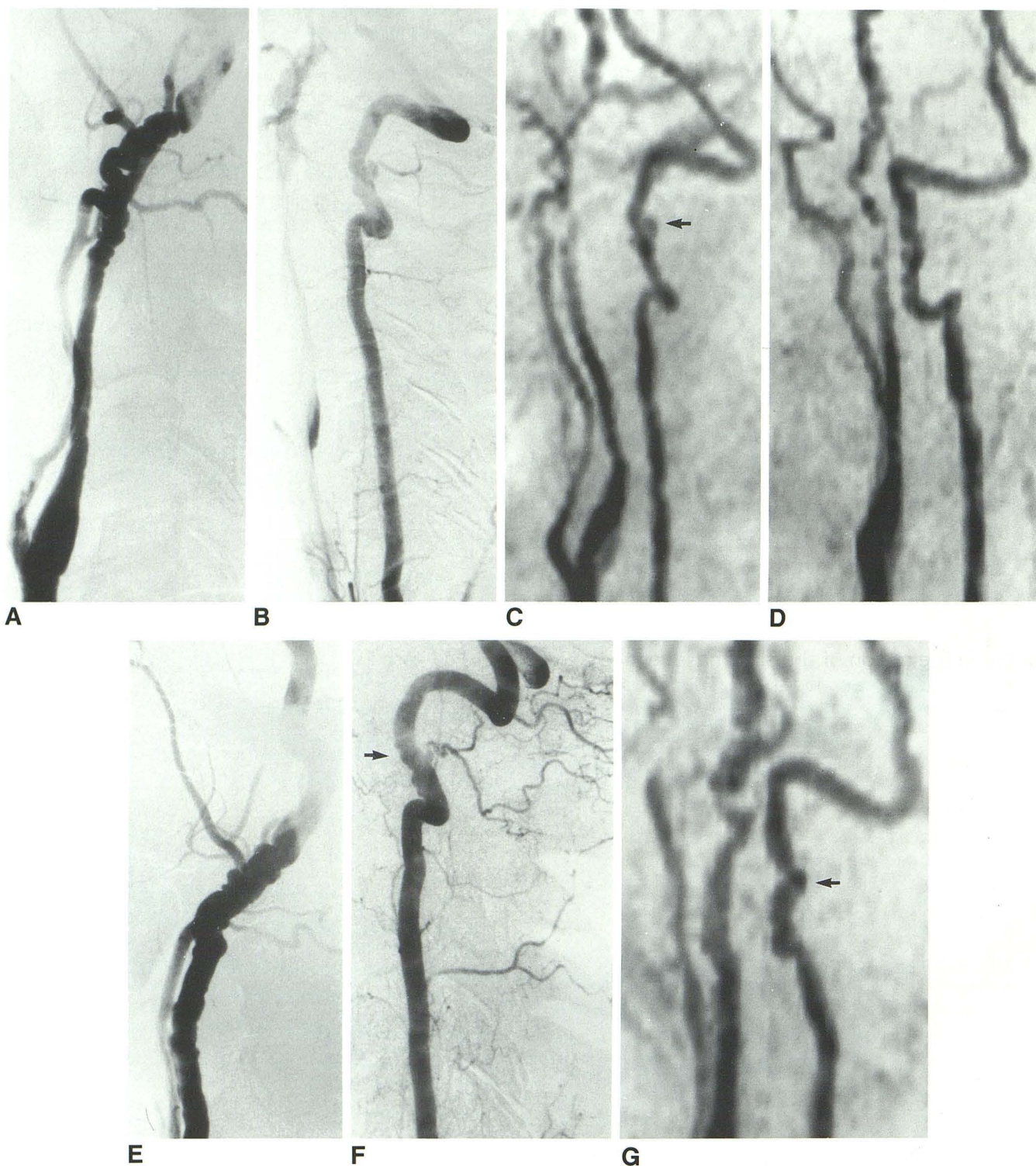


Fig. 2. Conventional angiograms of the right carotid (A) and vertebral (B) arteries in the lateral view in a patient with left hand numbness and right amaurosis fugax. The changes of FMD are present in both vessels, centered at the C1-C2 level. Narrowing and luminal irregularity are also present within the proximal cervical internal carotid artery beginning at the C3 level. Lateral (C) and anteroposterior (D) views from an MRA study of the same vessels demonstrate regions of decreased signal intensity and narrowing, approximating the appearance in the conventional study. Focal outpouching of the vertebral artery at the C1-C2 level seen on the conventional study is reproduced on the lateral projection from the MRA (arrow). For the MRA, TE = 9.5 msec. Conventional angiograms of the left carotid (E) and vertebral artery (F) in the lateral projection. Findings characteristic of FMD are seen involving the cervical internal carotid artery and are present subtly in the vertebral artery at the C1-C2 level (arrow). A lateral projection from the MRA (G) demonstrates irregular narrowing of the vessel lumen involving the cervical internal carotid artery and a focal luminal irregularity at the C1-C2 level in the vertebral artery (arrow). Subtle changes of FMD may be simulated or masked on MRA examinations by patient motion artifact.

appearance reflects mechanisms responsible for changes in signal intensity on two-dimensional time-of-flight MRA images (2, 10). In a vessel with a rapidly changing luminal diameter, non-laminar flow is expected—resulting in intravoxel dephasing and loss of signal intensity due to spin saturation related to increased residence time within the excitation slice. These phenomena lead to areas of decreased signal intensity, with recovery of signal further downstream when laminar flow is reestablished.

Several artifacts known to affect the two-dimensional time-of-flight MRA technique utilized in these examinations may lead to an appearance similar to that described here for FMD. Patient motion and swallowing may produce focal regions of apparently abnormal signal intensity within the carotid artery that may mimic FMD. The involvement of both carotid and vertebral arteries at the affected level may provide a clue to the artifactual nature of these changes. Motion artifacts may also mask the changes of FMD.

More subtle and often unilateral artifacts may occur in the region of the skull base related to in-plane flow and susceptibility gradients. These often occur near the point of entry of the internal carotid artery into the petrous bone. Such artifacts may mimic the appearance of FMD and will tend to decrease both the sensitivity and specificity of MRA for the detection of FMD.

Artifacts caused by susceptibility changes at the skull base are reduced by shorter echo times (TE) and thus the minimum achievable TE should be used. Three-dimensional time-of-flight MRA (11), which can be implemented using shorter echo times, may also be helpful in further evaluation of suspicious areas encountered near the skull base on two-dimensional time-of-flight examinations.

Conventional angiography is the examination of choice for the evaluation of FMD and exclusion of associated intracranial aneurysms. However, the noninvasive nature of cervical MRA suggests a role in the screening of patients with transient ischemic attack symptomatology, the follow-up of known FMD with or without therapy, and the evaluation of potentially affected relatives. Most importantly, changes suggestive of FMD may be recognized on MRA examinations performed for other indications, such as the routine evaluation of carotid stenosis.

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