ABNORMAL RADIONUCLIDE ANGIOGRAM IN PROVEN INTRACRANIAL FIBROMUSCULAR DYSPLASIA: CASE REPORT

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Left internal carotid occlusion with collateral perfusion was shown by radionuclide and contrast angiography in a 22-year-old woman. Fibromuscular dysplasia, intimal fibroplasia type, was found at autopsy in the intracranial carotid, vertebral, and basilar arteries.

Although fibromuscular dysplasia (FMD) (*1*) of the extracranial carotid and vertebral arteries is recognized with increasing frequency (*2*), only two cases of histologically proven FMD of the intracranial arteries have been described in print (*3,4*). This report describes radionuclide and conventional contrast angiography in a patient with FMD of the intracranial segments of the internal carotid, vertebral, and basilar arteries documented at autopsy.

CASE REPORT

A 22-year-old white college student was admitted with increasing headache and difficulty with speech. She had experienced episodes of nonspecific headache for 10 years. Ten days before admission she developed left-sided headache and sore throat. Tonsillitis was diagnosed and she was treated with an antibiotic. The headache increased in severity. She noted the onset of right facial numbness, difficulty with use of her right hand, and occasional difficulty in verbal expression. She was admitted for evaluation. Pertinent past history included long-standing menstrual irregularity treated with oral contraceptives for the past 10 years. Positive physical findings included harsh bruits over both orbits, expressive aphasia, right hemiapraxia, and a right plantar extensor response. No sensory abnormalities were noted. Normal laboratory findings included complete blood count, urinalysis, SMA-14, VDRL, and monospot test. Hemoglobin A was found present by electrophoresis. Skull and chest radiographs were normal. Electroencephalogram was compatible with diffuse cortical dysfunction. The radionuclide angiogram showed markedly decreased activity in the left middle cerebral artery distribution during the arterial phase, with increasing activity late in the venous phase (Fig. 1). This was thought to represent partial obstruction of the internal carotid with filling through collateral circulation. The static brain images 30 min later were normal. Two days later contrast arteriography showed occlusion of the left internal carotid artery with filling of the intracranial internal carotid segment through large ophthalmic collaterals (Fig. 2). The right internal carotid artery also showed marked

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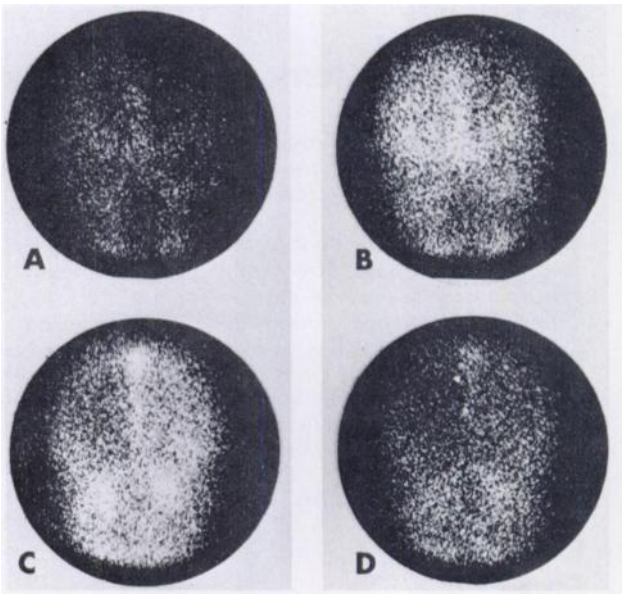

FIG. 1. Technetium-99m-pertechnetate angiogram. Early decreased perfusion of left middle cerebral distribution is evident in frames A and B, with subsequent increasing perfusion in frames C and D.

FIG. 2. Left common carotid arteriogram, lateral projection, showing complete tubular occlusion of proximal internal carotid (lower arrow) with collateral flow to intracranial segment through external carotid–ophthalmic collaterals (upper arrow).

narrowing at the level of the siphon. Twenty-four hours after the angiogram she developed right hemiplegia, increasing lethargy, and rapidly became decerebrate. She was declared neurologically dead 4 days later. At autopsy fibromuscular dysplasia, intimal fibroplasia type, was found in the intracranial segments of both carotid, vertebral, and basilar arteries, all of which were thrombosed (Fig. 3). The kidneys were taken by the transplant team, but were discarded.

DISCUSSION

Fibromuscular dysplasia is most common in adult women but has been reported in both sexes of all ages (*5,6*). Multiple arterial sites of involvement have been described (*5,6*). Most reported examples of FMD have been the medial fibroplasia type. Histologically, areas of medial hyperplasia and luminal narrowing alternate with areas of medial thinning and mural aneurysm. This appears on contrast angiography as the typical "string of beads" (*1*). Intimal fibroplasia with luminal obstruction, as in the present case, is rare (*1,6*).

Several instances have been reported of cerebrovascular accidents in young women with FMD of the cervical arteries complicated by oral contraceptives (*3,7*). Arterial thrombosis secondary to the contraceptive may obstruct collateral arteries, thus causing the acute episode of infarction (*3*). In the present case, there was thrombosis of the carotid, vertebral, and basilar arteries. Vascular proliferative lesions have been reported in patients on oral contraceptives but these are not identical to the lesions of FMD (*8,9*). Intracranial "berry" aneurysms may be associated with FMD (*2,6*). During the acute phase of cerebrovascular accidents, both radionuclide and contrast angiography often show early decreased perfusion in the involved area, with later increasing perfusion in the ischemic area through collateral channels (*10*). Static brain images are commonly normal during the acute phase (*10*). These findings occurred in the present case.

Although the scan findings are nonspecific, cephalic FMD must be suspected in young women presenting with cerebrovascular accidents. While not always apparent at gross inspection, the renal arteries are commonly involved in FMD. Since young patients expiring from cerebrovascular accidents or other brain trauma are often a source of organs for transplantation, the clinician must be alerted to the possibility of FMD to prevent the transplantation of a kidney or other organ already compromised by FMD.

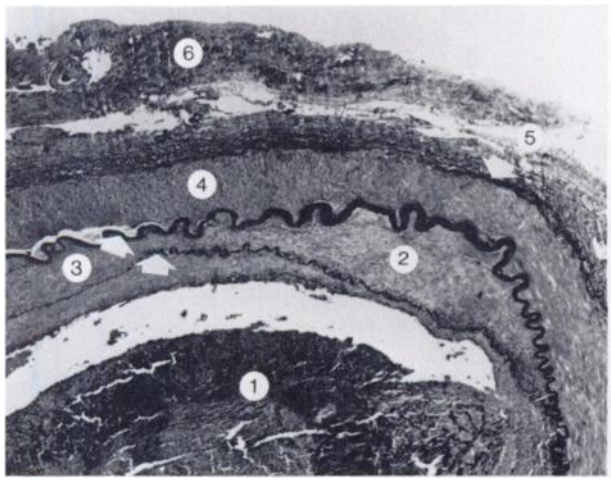

FIG. 3. Intracranial segment of left internal carotid artery, elastic stain. Lumen is filled with thrombus (1). Marked hypertrophy of internal elastica (3) with intimal thickening and fibrosis (2) is evident. Media (4), external elastica (5), and adventitia (6) are normal. Intima is normally much thinner than media.

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