Fibromuscular Dysplasia of the Posterior Cerebral Artery: Report of a Case and Review of the Literature

BY DAVID B. FRENS, M.D., JACK H. PETAJAN, M.D., PH.D., ROBERT ANDERSON, M.D.,* AND HAROLD J. DEBLANC, JR., M.D.

Abstract:
Fibromuscular dysplasia (FMD) of the posterior cerebral artery with classic presumptive angiographical findings is presented, and the literature pertaining to FMD of the cervical arteries is reviewed. The focal neurological findings and characteristic changing pattern seen on sequential brain scans clearly associate the presence of this lesion to cerebral infarction. Various proposed etiologies and the characteristic pathological and radiological appearance of FMD are discussed. Emphasis is placed on the potential for this lesion to produce significant clinical sequelae, and on the relationship between FMD and the presence of intracranial aneurysms. The natural history and proper treatment remain uncertain; however, the general impression is for slow progression of existing lesions associated with development of new lesions in other locations.

Additional Key Words: cerebral infarction in young adult, arterial occlusion, intracranial aneurysms.

Introduction
Fibromuscular dysplasia (FMD) of the internal carotid and vertebral arteries is a relatively rare lesion which was first recognized in 1964. Since that time at least 109 cases of FMD involving these arteries have been reported (table I). Involvement of the intracranial portion of these vessels has been recognized even less frequently, being reported in only 7 of 109 cases with FMD of the cervical arteries. The lesion in five of the seven cases was limited to the intromuscular portion of the internal carotid artery. A sixth case showed involvement of the intracranial portion of the internal carotid artery up to its bifurcation, and a seventh case showed clear involvement into the middle cerebral artery. The purpose of this paper is to report the first case of FMD involving the posterior cerebral artery and to review the literature dealing with this lesion in the cervical arteries.

Case Report
A 28-year-old right-handed white man was referred to the University of Utah Medical Center on May 27, 1973, with a 24-hour history of right-sided neurological deficit. Approximately 36 hours prior to admission he had a moderately severe, persistent, throbbing, bifrontal headache. Twenty-four hours prior to admission he suddenly became lightheaded, developed numbness and weakness of his entire right side, had diminished vision to the right, and noted difficulty with his speech. On initial evaluation he was mildly confused and inappropriate. There were a right visual field defect, diminished sensation of light touch, pin prick, position and vibration on the right, mildly diminished strength in both right extremities, symmetrical tendon reflexes, bilateral flexor plantar responses and a supple neck. Skull films and routine chemistries were normal. A lumbar puncture revealed clear fluid with an opening pressure of 130 mm H2O. The CSF protein was less than 10 mg %, and no cells were seen.

When he was seen at our hospital the speech difficulty, as well as the right-sided numbness and weakness, had disappeared. His major complaints were inability to see to the right and impaired memory. The blood pressure was 120/75 mm Hg, pulse rate 72 per minute, and temperature 98°F (36.7°C). General physical examination was unremarkable. The carotid and peripheral pulses were equal and full without bruits. The neck was supple. The patient appeared fully alert with a seeming indifference about his condition. Recent memory appeared mildly impaired. Cranial nerves were normal except for the presence of a right visual field defect. Sensation on the right was mildly decreased to light touch and pin prick. Motor and cerebellar functions were normal. Tendon reflexes were symmetrical, and the plantar responses were flexor.

During the hospitalization the complete blood count,
TABLE 1

Summary of Reported Cases of Fibromuscular Dysplasia of the Cervical Vessels

<table>
<thead>
<tr>
<th>Author</th>
<th>Number of cases</th>
<th>Cervical vessel involved</th>
<th>Number with intracranial aneurysms</th>
<th>Number with histological verification</th>
</tr>
</thead>
<tbody>
<tr>
<td>Palubinskas and Ripley</td>
<td>1</td>
<td>Internal carotid (bilateral)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Hill et al.</td>
<td>1</td>
<td>Internal carotid (bilateral)</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Houser et al.</td>
<td>52</td>
<td>Internal carotid (48)</td>
<td>10</td>
<td>3</td>
</tr>
<tr>
<td>(60% bilateral), vertebral (7)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Anderson</td>
<td>4</td>
<td>Internal carotid (3), vertebral (1) (bilateral)</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Bergan et al.</td>
<td>5</td>
<td>Internal carotid (3), vertebral (1), both (1)</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Ehrenfeld et al.</td>
<td>1</td>
<td>Internal carotid (bilateral)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Ennis et al.</td>
<td>3</td>
<td>Internal carotid (3) (bilateral in 2)</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Handa et al.</td>
<td>1</td>
<td>Internal carotid</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Huber et al.</td>
<td>3</td>
<td>Internal carotid (3)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Kaufmann</td>
<td>1</td>
<td>Internal carotid</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Kramer</td>
<td>1</td>
<td>Internal carotid</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Polin</td>
<td>3</td>
<td>Internal carotid (3)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Morris et al.</td>
<td>13</td>
<td>Internal carotid (12) (bilateral in 9), vertebral (1) (bilateral)</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Rainier et al.</td>
<td>1</td>
<td>Internal carotid</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Wylie et al.</td>
<td>6</td>
<td>Internal carotid (6) (bilateral in 4)</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>Galligioni et al.</td>
<td>3</td>
<td>Internal carotid (3)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Hartman et al.</td>
<td>1</td>
<td>Internal carotid (bilateral)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Lamis et al.</td>
<td>1</td>
<td>Internal carotid</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Pollack et al.</td>
<td>1</td>
<td>Internal carotid</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Kishore et al.</td>
<td>1</td>
<td>Internal carotid</td>
<td>0</td>
<td>1</td>
</tr>
<tr>
<td>Hooshmand et al.</td>
<td>1</td>
<td>Internal carotid (bilateral)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Perry</td>
<td>5</td>
<td>Internal carotid (5)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>109</td>
<td>Internal carotid (109) (bilateral in 48), vertebral (11) (bilateral in 2)</td>
<td>23</td>
<td>12</td>
</tr>
</tbody>
</table>

erythrocyte sedimentation rate, lupus erythematosus preparation, urinalysis, blood glucose, blood urea nitrogen, blood and CSF serology, skull films and electrocardiogram were normal. An electroencephalogram showed marked slowing in the left temporal and occipital regions. A lumbar puncture revealed clear fluid without xanthochromia and an opening pressure of 185 mm H₂O. The CSF protein was 43 mg %. The first tube contained 84 RBCs per cubic millimeter, while the third tube showed 117 RBCs per cubic millimeter. Four mononuclear cells were seen. An initial brain scan using 99m Tc-pertechnetate was negative four days after the onset of symptoms (fig. 1A). Ophthalmological evaluation showed normal visual acuity with a nontotal, relative right homonymous hemianopia.

A left vertebral angiogram was performed, which revealed an abnormal proximal left posterior cerebral artery. At least six discrete, sharply localized constrictions compromising the arterial lumen were seen. The arterial diameter between these areas of constriction was normal or greater than normal (fig. 2). Near the distal end of the irregular segment an outpouching of the arterial wall was seen, which was first thought to represent an aneurysm. On the AP view, however, an obvious lack of distal filling was seen on the left, indicating that this appearance of an "aneurysm" may in fact represent the proximal stump of an occluded arterial branch (fig. 3). Left carotid artery study demonstrated retrograde filling of a posterior cerebral branch via a distal anterior cerebral artery communication. The left carotid circulation was otherwise normal.

A second brain scan performed 12 days after the onset of symptoms was abnormal, demonstrating a well-defined triangular zone of increased activity in the left lower occipital region adjacent to the lower end of the Sylvian fissure, torcular and left transverse sinus (fig. 1B). This region corresponds precisely to the distribution of the left posterior cerebral artery. The abnormality also corresponded to the region of decreased vascularity in the region of the left optic cortex noted on the arteriogram (fig. 3).

Psychological testing revealed a full scale I.Q. of 127. All subtest scores were above normal except two. Both of these were concerned with immediate memory, one in the verbal half and one in the performance half of the test. The hospital course was stable, and the patient was discharged without treatment. A right visual field defect and a mild deficit in recent memory persisted.

Discussion

FMD is a nonatherosclerotic angiopathy of small-sized and medium-sized arteries first recognized in 1938. Various theories, including vascular stretch,
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congenital malformation, metabolic or hormonal derangement, trauma, inflammation and an abnormality of the vasa vasorum, have been proposed but not proved to be etiologically significant. There is no known familial tendency. No convincing association with Marfan-like syndromes, healed arteritis, or collagen vascular disease has been recognized.

The pathological features of this angiopathy have been extensively reviewed. Three histological patterns (intimal fibroplasia, medial fibromuscular dysplasia, and periadventitial fibroplasia) are recognized, based on the location of the major lesion within the arterial wall. The most frequently observed pattern is that of medial fibromuscular dysplasia. Histologically this lesion most often results in multifocal areas of arterial stenosis secondary to hyperplasia of the fibrous and muscular components of the arterial wall, alternating with areas of marked mural thinning and aneurysmal dilatation due to absence or thinning of the smooth muscle and disruption of the internal elastic membrane. Other patterns of arterial stenosis, including unifocal and tubular types, are occasionally found. FMD has been identified histologically in many different arteries. A marked female preponderance is widely recognized, young and middle-aged adult females representing 85% to 90% of the reported cases.

Although it has been reported in the cervical arteries in 109 cases, FMD has been histologically verified in only 12 cases (table 1). In one of these cases the lesion was noted to extend into the interosseous and intracranial portions of the internal carotid artery, demonstrating clearly that FMD can occur in vessels within the cranial cavity.

The characteristic roentgenographical finding which allows a presumptive diagnosis of FMD is a "string of beads" appearance of the arterial lumen. This pattern is produced by multiple, discrete constrictions of the lumen of the vessel with normal or dilated segments between. This appearance is apparently caused by alternating areas of medial thinning and thickening in the arterial wall. Involvement of the cervical arteries has been reported characteristically to affect that portion of the vessel immediately adja-

A: Posterior and left lateral views from brain scan done four days after insult. No abnormality is seen. B: Posterior and left lateral views from brain scan done 12 days after insult. Triangular-shaped lesion characteristic of left posterior cerebral infarction is clearly seen in both views.
Lateral view of mid-arterial phase of vertebral angiogram shows multiple, discrete constrictions of the lumen of the left posterior cerebral artery proximally (small arrows) and an apparent obstructed branch farther distally (large arrow). The distal vertebral arteries, basilar artery, and all other branches appear normal.

FMD of the cervical vessels is a relatively rare lesion which has been reported as an incidental finding in asymptomatic individuals and which has been associated with many vague clinical symptoms. FMD of the left posterior cerebral artery proximally (small arrows) and an apparent obstructed branch farther distally (large arrow). The distal vertebral arteries, basilar artery, and all other branches appear normal. Evidence that this lesion can cause clinical symptoms is further illustrated by the changing pattern seen in the patient’s brain scans. Approximately 50% of patients suffering ischemic cerebral infarction initially manifest normal brain scans within two weeks after insult. Most of these patients (75% to 80%) develop abnormalities corresponding to the distribution of the occluded artery in later scans. Though the demonstration of posterior cerebral artery infarction by brain scan is uncommon, probably because of the low incidence of this lesion, the changing pattern seen in this patient’s scans, and the classical shape and location of the lesion noted on the second scan, document that he suffered cerebral infarction related to FMD of the left posterior cerebral artery. FMD of the cervical vessels is a relatively rare lesion which has been reported as an incidental finding in asymptomatic individuals and which has been associated with many vague clinical symptoms. Despite its rarity, the lesion has important clinical significance because of its potential for producing severe neurological impairment in an individual of any age. Transient ischemic attacks and cerebral infarction have been clearly related to both arterial stenosis and occlusion caused by this lesion. Also of clinical importance is the association between intracranial berry aneurysms and FMD of an artery anywhere in the body. Twenty-three intracranial aneurysms were diagnosed angiographically in 109 reported cases of FMD of the cervical arteries (table I). In addition, three cases of subarachnoid or intracranial hemorrhage, without identification of the source of bleeding, were found. The large number of intracranial aneurysms found in these 109 cases adds increasing support to the contention that the association of FMD and intracranial aneurysms is more than
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AP view shows absence of filling of branches of the posterior cerebral artery distally on the left (arrows). Compare with AP brain scan, figure 1B

merely chance. It also suggests that the discovery of this lesion in any artery should be followed by a cerebral arteriogram to rule out intracranial aneurysm.

The proper treatment of FMD of the cervical arteries remains uncertain. When a lesion is clearly related to sequential clinical symptoms and is accessible, operation by a variety of techniques seems to be the best approach. Proper treatment of the symptomatic but inaccessible lesion, such as the one found in our case, remains completely unsolved. Only one patient is reported in whom an anticoagulant was given for TIA’s, which were believed to be secondary to FMD of the internal carotid artery. In this case the TIAs disappeared, suggesting that anticoagulant treatment may be at least one method of therapy for the inaccessible symptomatic lesion.

Increasing the problem of selection of proper treatment is the fact that little is known about the natural history of the disease. A report of 16 follow-up renal arteriograms in patients with FMD of the renal artery over a five and one-half-year period showed definite progression and development of new lesions in six cases. No regression of a previously existing lesion was found. Evidence as to the natural history of this lesion in the cervical arteries is even less available. Of the 109 cases of FMD involving the cervical arteries, three had second arteriograms over a period from three to nine months after the initial study. Two patients showed no progression. A third developed a new lesion in the opposite internal carotid artery within three months. The general impression remains that FMD is a slowly progressive lesion of small-sized and medium-sized arteries with a potential for causing serious clinical disease.

References

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