SUMMARY  A young man with cerebral infarction, skeletal, cardiac and renal malformations was found to have on angiography a rare lesion involving the cervical left internal carotid artery, formed by the superposition of two arterial lumina. Anti-platelet therapy did not prevent thrombosis of the lesion and reinfarction. Extracranial-intracranial bypass graft was performed, followed by excision of the arterial lesion. Pathological examination revealed fibromuscular dysplasia and dissecting aneurysm. The few reported cases associating fibromuscular dysplasia of the internal carotid artery with dissecting aneurysm are reviewed. Surgical therapy is recommended in such cases when symptomatology progresses. Fibromuscular dysplasia occasionally coexists with somatic malformations, suggesting a congenital origin of this condition.

FIBROMUSCULAR DYSPLASIA (FMD) involving the internal carotid artery is a disease of unknown natural history in spite of the large number of cases reported. It has been associated with ischemic brain accidents in young people, although it is an incidental finding in many patients. Because of the usual good prognosis, therapy must be individualized. The condition has recently been reviewed.1, 2, 3, 4

Spontaneous dissection is a recognized complication of renal artery FMD.5 However, only a few cases have been recorded in internal carotid artery FMD, most of them without histological proof. The management of this complication is controversial.

We present here a case of internal carotid artery FMD associated with dissecting aneurysm showing double lumen on angiography in a young patient with viscero-skeletal malformations.

Report of a Case

A 22-year-old man was admitted to the hospital in January 1980 because of the sudden onset of right-sided weakness and aphasia. Past medical history revealed a single episode of generalized convulsions in 1978.

General examination showed pectus excavatum, scoliosis, a second heart sound split and fixed, and a grade 2/6 systolic ejection murmur at the left upper sternal border. The neurological findings included a right hemiparesis, hyperactive reflexes on the right, hypesthesia over the right side and non-fluent aphasia.

Routine blood and urine examinations were normal. The electroencephalogram disclosed theta waves over the left temporal area. A right bundle branch block appeared in the electrocardiogram. Computerized tomography of the head demonstrated a low-density lesion in the territory of the left middle cerebral artery.

Transfemoral arteriography was performed. Two centimeters distal to its origin, the left internal carotid artery showed a complex vascular tangle formed by the superposition of two arterial lumina, extending from the level of C3 to C1 (fig. 1). The rest of the angiography, including the contralateral carotid, vertebrals and intracranial vessels was without abnormalities.

Renal arteriography and urography, performed after an episode of frank hematuria, revealed a dysplastic and hydronephrotic left kidney with normal arteries.

The cardiological study was consistent with an atrial septal defect, ostium secundum type.

A treatment with aspirin and dipyridamole was instituted and the patient left the hospital.

Four months after the first admission he suffered a new stroke involving the left cerebral hemisphere. A repeated left internal carotid arteriography showed then a partial thrombosis of the arterial lesion (fig. 2A and 2B).

Anastomosis of the left superficial temporal artery to the left middle cerebral artery was performed. One month later, the cervical left internal carotid artery was surgically exposed: the vessel had a fusiform dilation reaching the base of the skull consistent with dissecting aneurysm. At arteriotomy large amount of thrombotic material with interspersed vascular channels was found, filling the aneurysmatic bed. The arterial wall was very friable so that reconstructive maneuvers were not feasible. In view of the stump pressure (85 mm Hg) and the risk for embolization, excision of the artery was performed.

Pathological study revealed FMD and extensive dissection (fig. 3A and 3B).

Postoperative course was uneventful except for the development of an important soft tissue fibrosis around the areas of previous surgery. Two years after the last operation the patient had not suffered new neurological complaints.

Discussion

Spontaneous dissection in FMD may be due to degenerative changes in the arterial wall. Pollock and Jackson found treatment of the intima and internal
elastic membrane from the media, associated with carotid FMD. They suggested that a cleavage phenomenon would account for the increased risk of spontaneous dissection in certain types of renal artery FMD.

Since this report we have found in the literature six cases of spontaneous dissection complicating internal carotid FMD. Four of them were diagnosed by the finding of changes typical of FMD in the arteriogram of the contralateral carotid, and the rest by pathological examination. None showed double lumen on angiography. Surgery was attempted in three cases; in two arteriotomy was not performed because the dissection extended beyond the limits of surgical accessibility. In one case a saphenous vein graft replaced the resected aneurysm.

Arterial dissection has also been described associated with vertebro-basilar FMD. In absence of pathological proof, some cases of dissection causing stenosis of the lumen or thrombosis may be misdiagnosed if associated with a localized form of carotid FMD. This fact would explain in part the scarcity of reports of this complication in the carotid artery while it is fairly common in other arteries.
affected by FMD, of equal caliber and similar histologic type.7

The optimum treatment for dissecting aneurysms of the internal carotid artery is not established. A conservative management of neurologically stable patients has been recommended since the aneurysms may disappear in some cases without surgery.8,10,12 In these patients anticoagulant therapy may be initiated in order to prevent thrombosis of the lumen or embolization from a mural thrombus.12,13 Anti-platelet agents have been used,10 but they are probably less effective than anticoagulants. In our case, brain ischemia recurred in spite of that therapy.

Surgery seems indicated when a progressive or recidivant neurological deficit takes place. The long extension of dissecting aneurysms in carotid FMD is an obstacle to the usual surgical technique for aneurysms. Because of that, intracranial-extracranial bypass graft may be performed. This procedure has successfully been applied in carotid FMD13 and in spontaneous dissection of the internal carotid artery.14 If required, it may be followed by a direct surgical approach to the cervical internal carotid artery, or by arterial occlusion by ligature, Selverstone clamp, or detachable balloon. In our case, surgical repair of the aneurysm was attempted after the bypass graft, but arterial excision was finally performed because of surgical difficulties and the high risk for embolization.

The etiology of FMD is obscure. The association, in certain cases, of FMD with somatic malformations, hypertension, otosclerosis, and its occasional familial occurrence, suggest a congenital origin.4 Our patient showed pectus excavatum and renal dysplasia, already described in a few cases of FMD.15,16 We have found no previous reports of cardiac septal defects or scoliosis. It has been pointed out that FMD would reflect a hereditary mesenchymal disorder in which an abnormal fibroproliferative response to mechanical or circulatory stimuli exists, rather than a specific arterial defect.4,15,16

Recently Monfort et al have reported one case of dissecting aneurysm of the internal carotid artery associated with FMD, angiographically demonstrated.17 Sato and Hata have described another case of this association with histological verification.18 Both articles cite Manelfe et al19 who reported three cases of dissecting aneurysms of the carotid arteries, one of them with histological study, among 70 patients with FMD of the cervicocephalic arteries.

According to these authors, dissecting aneurysm would complicate FMD of the internal carotid artery more commonly than is in general believed.

Acknowledgments

We are indebted to Dr. J. R. Fernández-Espino, from the Pathology Department, Centro Ramón y Cajal, and to Drs. M. L. Pascual and E. Martín, from the Service of Neurology, Clínica Puerta de Hierro, for their help.

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*Stroke*. 1983;14:815-818
doi: 10.1161/01.STR.14.5.815

*Stroke* is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0039-2499. Online ISSN: 1524-4628

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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