Fibromuscular hyperplasia of the extracranial internal carotid artery

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Fibromuscular hyperplasia is an alteration of the arterial wall, affecting mostly middle-aged women. When localized to the internal carotid arteries, it can give symptoms of intermittent or permanent cerebral ischemia, but is usually asymptomatic. Three cases, all of them accidental angiographic findings, are used as a basis for discussion of the angiographic features, differential diagnosis, and surgical treatment.

KEY WORDS: carotid artery, fibromuscular dysplasia, angiography, arterial occlusive disease, cerebrovascular insufficiency

We have recently seen three cases of a condition which, because of its neurological consequences, deserves more attention in the neurosurgical literature. Reports of similar cases have appeared in American periodicals of general surgery and radiology, but to the best of our knowledge, the problem has not been discussed in a neurosurgical journal.

Like other causes of arterial stenosis, fibromuscular dysplasia can affect the internal carotid artery and thus cause permanent or intermittent cerebral ischemia. It appears most often in 40- to 50-year-old women and is usually an accidental arteriographic finding.

Pathologically it is characterized by “irregularly spaced focal zones of fibrous and muscular hyperplasia of the media, disruption of elastic lamellae, and the occasional appearance of berry aneurysms at points of disruption of the media.” These alterations usually begin in the middle third of the extracranial internal carotid artery, extending upwards for varying distances. The disease produces a characteristic angiogram with a “loose stocking” pattern of irregularities of the arterial lumen, caused by zones of dilatation alternating with irregularly spaced ring-like constrictions that may reduce the caliber of the vessel by as much as 40%. This typical angiographic appearance makes diagnosis easy, and differentiates the disease from other organic or functional causes of stenosis of the internal carotid artery, such as plain arteriosclerotic changes, stationary arterial waves, and filling defects due to improper techniques of puncture, cannulation, catheterization, and injection of the carotid artery.

Fibromuscular dysplasia can be limited to the carotid arteries; more frequently it is the local expression of a more widespread, systemic disease involving other arteries, such as the renal, celiac, mesenteric, iliac, and coronary. Its most frequent site is the renal artery, where it produces the clinical picture of hypertension; a well-defined bypassing surgical procedure is then available.

Fibromuscular dysplasia of the internal carotid artery was first described by Connett and Lansche and then by others.
though the syndrome is usually asymptomatic, it sometimes is characterized by intermittent crises of cerebral ischemia\(^1\) that depend on positional changes.\(^1\) Its asymptomatic course and its obscure natural history possibly account for the lack of a precise therapeutic program.

We found incidental angiographic evidence of fibromuscular dysplasia of the internal carotid artery in three patients. In the first, a carotid arteriogram had been performed for a subdural hematoma (Fig. 1). In addition to the hematoma, the left carotid angiogram demonstrated the characteristic “loose stocking” appearance of the extracranial left internal carotid, caused by alternating regions of dilatation and constriction (Fig. 2 upper left and right). Three months later a right carotid angiogram showed early changes of the same disease in the right carotid (Fig. 2 lower left and right).

In our second case, the patient had had a head injury. The intracranial angiogram was normal but again showed the incidental finding of a “loose stocking” appearance in the extracranial internal carotid (Fig. 3).

The grave clinical significance of fibromuscular dysplasia of the renal arteries as contrasted to that of the internal carotids probably is related to the different circulatory patterns of these arteries. While the renal arteries have a terminal character and possibilities of collateral circulation are poor, defects on one side of the carotid circulation, even of some gravity, can be adequately compensated for by the contralateral carotid or by the vertebral artery, through the communicating arteries. Thus, while certain indications for surgical treatment have by now been established for fibromuscular dysplasia of the renal artery, a similarly specific program of treatment for fibromuscular dysplasia of the internal carotid artery has not yet been formulated. The concepts underlying surgical treatment of cerebrovascular insufficiency have been discussed by De Bakey, et al.\(^5\) This is the reason why we think that patients with this apparently incidental disease should be followed closely to detect the onset of neurological symptoms; at that time a complete clinical, electroencephalographic, and arteriographic reevaluation would be indicated and the possibility of a surgical program entertained.

From a neurosurgical point of view, it would also be worthwhile to keep in mind the association of intracranial aneurysms and fibromuscular dysplasia, which Wylie, et al.,\(^2\) have emphasized as “greater than that of chance alone.” There is a preponderance of women and young adults among patients affected by both diseases. There is a strong histological similarity: both conditions are characterized “by degeneration or absence of
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Fig. 2. Case 1. Details of left arteriogram showing alternating dilatation and constriction of carotid artery characteristic of fibromuscular dysplasia. Upper Left: Anteroposterior view. Upper Right: Lateral view. Lower Left and Right: Right arteriogram, 3 months later, shows incipient alterations of the contralateral internal carotid artery.
the media, disruption of the elastica, and loss of the internal elastic membrane." Furthermore, both aneurysms found in the renal arteries affected by fibromuscular dysplasia and those in the intracranial vessels tend to develop at arterial bifurcations. The association between fibromuscular dysplasia of the renal artery and intracranial aneurysms, however, could also be due to the effect of arterial hypertension in patients with stenosis of a renal artery upon weak points in the wall of predisposed cerebral arteries. This latter factor has been considered responsible for the development of intracranial aneurysms even in the absence of a diseased renal artery or hypertension.

Summary

The angiographic identification of asymptomatic fibromuscular dysplasia of the extracranial internal carotid artery has been discussed. The possibility is raised that this condition could, at some future time, produce neurological symptoms requiring surgical treatment.

References

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FIG. 4. Case 3. Right carotid angiogram, anteroposterior views, showing multiple evidence of a meningioma, plus the “loose stocking” appearance of the extracranial internal carotid artery.

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