The Syndrome of Intracranial Aneurysm Associated with Fibromuscular Hyperplasia of the Renal Arteries

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FIBROMUSCULAR hyperplasia (FMH), as a cause of renal artery stenosis and consequent renovascular hypertension due to ischemia of one or both kidneys, first became recognized as a pathological entity in 1938, although the majority of reports have appeared only recently.1,3,5,8,10,11

By 1965, six patients were reported with a combination of the renal artery abnormality and intracranial aneurysms, none of which had apparently been responsible for a subarachnoid hemorrhage.6

The purpose of this paper is threefold: first, to call attention to the coexistence of intracranial aneurysms in a high percentage of patients with FMH of the renal arteries; second, to present yet another patient with FMH who, following renal artery surgery, developed a subarachnoid hemorrhage due to a middle cerebral artery aneurysm, for which she underwent craniotomy and clipping of the aneurysm, with recovery; and third, to comment on two other angiographic discoveries in this patient, “stationary waves” of one internal carotid artery and a fusiform splenic artery aneurysm.

Case Report

Hypertension was first discovered in this 37-year-old right-handed white woman in 1964 in the last trimester of her tenth pregnancy. She showed mild hypertensive retinopathy and an abdominal bruit. Her blood pressure (190-230/110-140) was easily controlled by diuril and reserpine.

Renal function was moderately impaired on the right as determined by intravenous pyelogram and renogram, and an aortogram revealed the characteristic changes of fibromuscular hyperplasia (FMH) in both renal arteries, more marked on the right. In addition, there was a fusiform aneurysm of the splenic artery (Fig. 1).

A right renal artery reconstruction was attempted with a dacron prosthesis and vein grafts, but postoperatively a renal scan demonstrated non-function of the right kidney.

Postoperative hypertension was well controlled by diuril and apresoline, and the patient remained normotensive until 3 months later, a subarachnoid hemorrhage occurred together with a gradually evolving right hemiplegia and global aphasia. She was discovered to be hypertensive again. Her blood pressure was reduced by drugs; in 5 days her hemiplegia had cleared, and within 3 weeks some expressive and all receptive speech function had returned. Six weeks after the hemorrhage she had normal motor and sensory function, but a moderately severe expressive dysphasia. Bilateral carotid arteriography revealed a left middle cerebral artery aneurysm (Fig. 2 left) and also “stationary waves” of the left internal carotid artery (Fig. 2 right).

Following angiography, the patient had a severe left hemiparesis (ipsilateral to the aneurysm) and complete aphasia. These resolved in 4 days, and when the patient’s condition had stabilized, the aneurysm was dissected free at craniotomy and clipped. Postoperatively, the patient did well except for a transient increase in her expressive speech deficit and recurrent hypertension which was satisfactorily controlled by drugs. A postoperative arteriogram showed proper placement of the clip and obliteration of the aneurysm, and also disappearance of the “stationary waves” of the left internal carotid artery.

Discussion

Fibromuscular hyperplasia is a disease primarily described in the renal arteries.5,10,11 It frequently causes hypertension secondary to the renal artery stenosis, and 85% of cases are seen in young-adult or middle-aged women. It is also observed in other arteries, such as
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Fig. 1. Aortogram of right renal artery. *Left:* Anteroposterior view shows bead-like constrictions characteristic of FMH (arrow). *Right:* Oblique view confirms aneurysmal dilatation of splenic artery (large arrow) and also involvement of the left renal artery by FMH (small arrow).

The celiac axis, external iliacs, superior mesenteric, and internal carotids, as a silent lesion except for the occasional carotid insufficiency syndrome. Its etiology is unknown, although case reports of female siblings affected by it suggest a genetic predisposition in some instances.\(^2,3\) Bilateral involvement of paired arteries is usually found, and characteristically, the abnormality in the renal arteries is seen only in the distal half of the vessel, having a corrugated appearance resembling a string of beads of varying sizes (Fig. 1). Finally, there is an increased incidence of intracranial aneurysm associated with this renal artery abnormality.\(^5\)

Pathologically, there is irregular thickening of the vessel by the proliferation of smooth muscle and fibrous tissue of the media narrowing the lumen; or the stenosis may be caused by the projection of thickened septa, trabeculae, and ridges into the lumen. The arterial wall between these ridges is abnormally thinned, and aneurysmal dilatations are often seen. There may also be marked

Fig. 2. Left carotid angiograms. *Left:* Oblique view reveals middle cerebral artery aneurysm (arrow). *Right:* Subtraction film indicates transient corrugations (stationary waves) in extracerebral portion of internal carotid artery (arrow).