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.1,2,3,5,8,10,11

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. 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.

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).
overgrowth of fibrous tissue and smooth muscle just under the adventitia, as well as proliferation of the intima, and the internal elastic membrane is often fragmented. There is never evidence of inflammation. The basic pathophysiologic abnormality is the hyperplasia of medial smooth muscle, causing stenosis of the renal arteries and secondary hypertension (Fig. 3).

The angiographic appearance of FMH is so characteristic that the diagnosis can be made by arteriography alone. The surgery for correction of the hypertension, in which the involved segment is excised and an end-to-end anastomosis of the renal artery is performed, sometimes coupled with the use of vein grafts, is quite satisfactory; of 42 operated cases, 75% had significantly reduced blood pressure postoperatively.

The association of intracranial aneurysm with genetically determined systemic arterial diseases, such as Marfan's syndrome and coarctation of the aorta, is well-recognized. Like these, FMH is also a disease affecting the smooth-muscle layer of large arteries, and a genetic tendency has been noted in at least two families (five middle-aged females). That it should be associated with intracranial aneurysm, therefore, is not unexpected. Indeed, where carotid arteriograms have been done in patients with (angiographically) proven FMH, 50% had intracranial aneurysms, although none of these had apparently bled. (Hansen, et al., reported two sisters with FMH who died suddenly, each with massive subarachnoid hemorrhages. Even though carotid arteriography had not been done antemortem and aneurysms were not demonstrated at autopsy, the possibility that they were present, though undiscovered, is strong. Since hypertension is usually present in these cases, the danger of rupture of the intracranial lesion would seem to be much higher than usual. Therefore, when FMH is discovered to be the cause of a case of renovascular hypertension, carotid arteriography should be considered. (If the femoral approach has been used on the aortogram, the catheter can be directed into the carotid system at that time.) Conversely, if a subarachnoid hemorrhage occurs in a young woman with hypertension, FMH must be suspected. The importance of this is self-evident since this is one of the few causes of elevated blood pressure amenable to surgical correction.

In our case, "stationary waves" were seen in the internal carotid on the side of the aneurysm (Fig. 2 right). The appearance has been termed "corrugated," as in FMH, and likened to a "string of pearls." These waves are regularly transverse striations seen in the contrast column during arteriography and not associated with narrowing of the arterial lumen. Their etiology is unknown; the theories are that they represent a type of arterial spasm, and are completely ablated by intra-arterial vasodilator drugs, or that they are due to distal obstruction to blood flow and the consequent rippling of the blood-contrast interface during arteriography, when the difference in velocity between the two layers is just sufficient to produce rippling but not mixing. Part of their significance is that the change may closely simulate the appearance of early FMH; thus, in the absence of histologic proof, or without a syndrome of progressive obstruction of the involved vessel, the FMH label may be erroneously applied. Actually, the appearance of advanced FMH is unequivocal, the corrugation being not quite so regular, getting worse with time, and not improving spontaneously (as here); nor can it be eliminated with vasodilators. In early FMH, however, real doubt may exist in differentiating the two lesions. The simultaneous appearance in this patient of FMH and stationary waves was entirely fortuitous.

The finding of a fusiform aneurysm of the splenic artery in this case is of interest in the context of the basic disease present. The artery does not show the beading of the FMH, but is enlarged generally; perhaps this represents the earliest change of FMH in this
vessel, or it may simply be a coincidental finding. Without histologic verification, no definite answer is possible, but we would tentatively add the splenic artery to the growing list of vessels that may be "silently" involved in the FMH process.

Summary

We have briefly discussed the association of fibromuscular hyperplasia, a stenosing disease of the renal arteries causing secondary hypertension, with intracranial aneurysm. This is the first recorded instance of a subarachnoid hemorrhage occurring in such a patient in which an aneurysm was demonstrated as the cause, and with a successful surgical outcome.

The patient we have reported had a subarachnoid hemorrhage with subsequent successful surgical elimination of the aneurysm of the middle cerebral artery. We have also emphasized the possible significance of two other angiographic observations, namely, "stationary waves" in the carotid and a fusiform aneurysm of the splenic artery.

References