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Spontaneous Dissections of the Renal Arteries in a Patient With Previous Spontaneous Dissections of the Internal Carotid Arteries

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SUMMARY An otherwise healthy 35-year-old woman suffered spontaneous dissections of both internal carotid arteries. She made an excellent recovery but was left with occlusion of the left internal carotid artery and a residual subcranial dissecting aneurysm of the right artery — both were asymptomatic. Eight years later, spontaneous dissections of both renal arteries occurred. The exact nature of the underlying arterial disease is not clear. Although fibromuscular dysplasia is suspected, other undetermined arteriopathy cannot be excluded.

To our knowledge, such an association has not been previously reported.

Report of a Case

In early June 1975, at the age of 35 years, an otherwise healthy right-handed woman experienced a deep-seated pain in the right ear and became aware of a small pulsatile mass in the right side of her neck and a loud bruit over the entire right side of her head. Within ½ hour the pain involved the right side of her head and face. Within 12 hours, she noted drooping of the right upper lid. Symptomatic treatment was not helpful. Neurologic evaluation led to transfemoral bilateral ca-
Figure 1. A (top left) Right carotid arteriogram shows subcranial dissecting aneurysm (solid arrow) adjacent to narrowed segment of arterial dissection, which extends into carotid canal (open arrow). B (top right) Right vertebral arteriogram shows luminal irregularities at C-2. C (bottom left) Right carotid arteriogram 11 months after diagnosis shows significant decrease in aneurysm and resolution of intracanalicular dissection. D (bottom right) Left carotid arteriogram 11 months after diagnosis shows occlusion of internal carotid artery distal to its origin. Collateral flow from the accessory meningeal artery (lower arrow) and artery of pterygoid canal (upper arrow) fills cavernous internal carotid artery. E (facing page) Right vertebral arteriogram 11 months after diagnosis shows resolution of luminal irregularities.
rotid and vertebral cerebral angiography on July 2, which demonstrated occlusion at the origin of the left internal carotid artery, a subcranial saccular aneurysm of the right internal carotid artery with an adjacent stenosis of the internal carotid artery extending into the carotid canal (fig. 1 A), and luminal irregularities in the vertebral arteries at C-2, suggestive of (although not typical for) fibromuscular dysplasia (fig. 1 B). Further workup, including investigations for detection of collagen vascular disease, revealed no abnormalities or clues. The patient was advised to stop using oral contraceptives, which she had been taking for a number of years, and to stop smoking cigarettes.

In August 1975, the patient was evaluated at the Mayo Clinic. She reported that the intensity of her symptoms had been decreasing.

Neurologic examination showed a mild ptosis of the right lid, equal and normally reacting pupils, and grade 2/6 systolic bruits over both carotid arteries. There were no cardiac murmurs. The rest of the neurologic examination was normal. Brachial blood pressure was 115/70 mm Hg, and the pulse rate was 60/min and regular. General medical examination and lengthy laboratory investigations disclosed no abnormalities.

In May 1976, the patient returned for follow-up evaluation. Her right-sided headaches had resolved, but she had had fairly frequent occipital bifrontal headaches, with the characteristics of tension headaches. The subjective bruit had decreased and was audible only in a quiet room or after exercise. On examination, the only finding was slight ptosis of the right upper lid. The carotid bruits had disappeared. Transfemoral bilateral carotid and vertebral angiography showed considerable improvement from the earlier findings. Only mild tortuosity and minimal luminal irregularities of the right internal carotid artery persisted in the carotid canal (fig. 1 C). The subcranial aneurysm had significantly decreased in size. The complete occlusion of the left internal carotid artery had remained unchanged (fig. 1 D). The vertebral arteries were now normal (fig. 1 E). An indeterminant arteriopathy with tendency to arterial dissection and subsequent regression was suspected. The patient had remained normotensive.

Over the ensuing years, the patient’s tension headaches became less frequent and the subjective right-sided bruit was noted only rarely or for a brief period after vigorous activity. The patient’s general medical and neurologic status remained stable and satisfactory.

In early June 1983, while watching a ball game, the patient experienced pain in the left flank, followed by nausea. The pain gradually extended to the left groin and anterior aspect of the left thigh. That evening, she was seen by her local physician. Examination of urine revealed microhematuria. The brachial blood pressure was 180/120 mm Hg. Treatment involved bed rest and analgesics, the initial impression being renal stone. By this time, the patient also had a severe generalized headache. After about 3 days, pain also developed in the right flank, extending to the right groin and thigh. She now had severe nausea and frequent vomiting. Transfemoral abdominal aortography and bilateral renal angiography on June 18, 1983, demonstrated dissection involving the main renal artery, extending into the two major primary branches (fig. 2 A). This was accompanied by high-grade stenosis at the bifurcation. In addition, there was irregularity of the right main renal artery, also involving primary and secondary branches.

The patient’s hypertension was well controlled with antihypertensive agents. The headaches, flank pain, nausea and vomiting resolved within 2 weeks. Repeat abdominal aortography and bilateral renal arteriography on October 1983 demonstrated intimal irregularities in the aorta distal to the take-off of the renal arteries. These irregularities were shallow and extended into both common iliac arteries. There was no abnormality in the take-off of the right and left renal arteries. The right kidney had a small 4- by 5-mm aneurysm in the middle portion (fig. 2 B) and a cortically based wedge-shaped area of infarction at the junction of the middle and lower poles (fig. 2 C). The left renal artery was narrowed at the bifurcation of the branches to the upper and lower poles. The patient has remained essentially asymptomatic but has continued taking her antihypertensive medications.

Discussion

Spontaneous or traumatic mural arterial dissection is produced by penetration of the circulating blood into the wall of an artery and its subsequent extension for varied distances along the arterial wall, causing a false
lumen of blood within the arterial wall (usually within the media). Sometimes the hemorrhage ruptures back into the original arterial lumen, creating a double lumen. Among the cervical cephalic arteries, the extracranial segment of the internal carotid artery is the vessel most commonly involved. Dissection is "spontaneous" when no overt trauma is reported. Spontaneous dissections of the cervical segments of the internal carotid arteries are uncommon but not rare. The first case was reported in 1954. Subsequent reports appeared sporadically, until clinicians and radiologists became more familiar with the clinical and angiographic features of this entity. In a recent review, 180 cases were collected from the English language literature. Nonetheless, the disease is still more common than the literature indicates.

Renal artery dissection occurs more commonly as a continuation of an aortic dissection or sometimes as an isolated dissection that usually is attributed to trauma (for example, from a catheter tip). However, isolated spontaneous dissection of the renal artery is rare but has been reported. The first case was described in 1944 and a number of reports followed. The exact cause of spontaneous dissection of the internal carotid artery, as well as spontaneous dissection of the renal artery, is unclear. However, both entities have been observed with higher frequency in association with fibromuscular dysplasia. An underlying disorder of the blood vessel wall is probably a significant factor in the pathogenesis of spontaneous dissection.

Pathologic studies of spontaneous dissection of the internal carotid artery on postmortem or surgical specimens have demonstrated changes of fibromuscular...
dysplasia, cystic medial degeneration, fragmentation, and disorganization of elastic fibers. Spontaneous dissection also has been noted with the Marfan syndrome. The exact nature of the underlying arterial disease in the present case is not clear. Fibromuscular dysplasia is a probability, although the possibility of an undetermined arteriopathy cannot be excluded because of the resolution of the irregularities of the vertebral arteries.

Generally, spontaneous dissection of the renal artery is seen in middle-aged men. The most frequent clinical accompaniments of this entity include hypertension, flank pain, hematuria, and headaches. The involvement is more frequently bilateral than unilateral. The common clinical manifestations of spontaneous dissection of the renal artery include unilateral pain in the head, face, or neck, occlusive symptom complex, subjective and objective bruits, and focal cerebral ischemic symptoms.

The prognosis for most patients with spontaneous dissections of the internal carotid arteries is very good. More than three-fourths of the patients with spontaneous dissections of the internal carotid arteries recover without residuals, and the angiographic abnormalities either resolve or significantly improve, and only infrequently do they progress to complete occlusion. Most patients with spontaneous dissections of the renal arteries also do well and remain asymptomatic on antihypertensive therapy. External rupture of the dissection has not been observed, and, in general, renal function remains relatively stable with adequate control of the hypertension.

Although dissection of visceral arteries, such as hepatic, splenic, and mesenteric arteries, as well as dissection of the vertebral artery, has been noted in association with spontaneous dissections of the renal arteries, the association of spontaneous dissections of the renal and internal carotid arteries has not been previously reported.

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