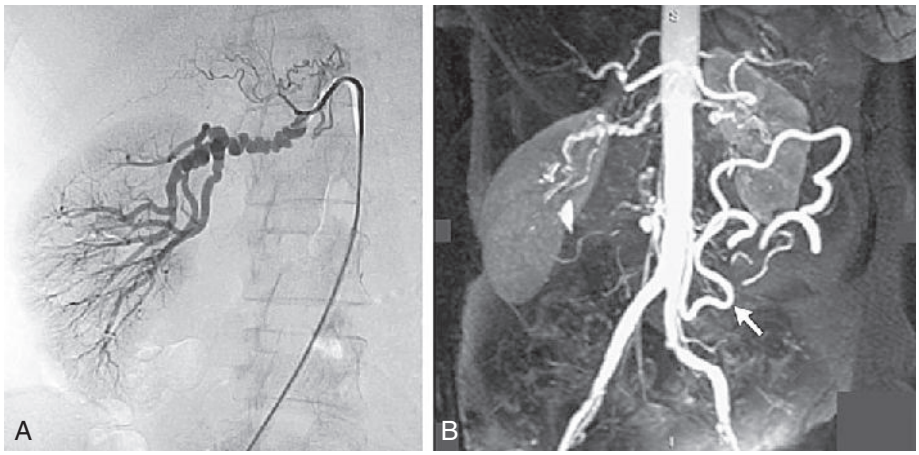

FIGURE 30-3 **A,** Typical medial fibroplasia (i.e., string-of-beads appearance) on an angiogram of a right renal artery. **B,** Gadolinium-enhanced magnetic resonance angiography in the same patient, revealing bilateral medial fibroplasia of the renal arteries and a large marginal artery of Drummond *(arrow)*, indicates that there is disease of the superior mesenteric artery. (From Slovut DP, Olin JW: Fibromuscular dysplasia, N Engl J Med 350:1862–1871, 2004.)

Medial subtypes of FMD usually have a benign course and are responsive to angioplasty. The intimal subtype may have a higher likelihood of ischemic events and multiorgan system involvement. Symptoms are usually precipitated by stenoses, but FMD may rarely cause dissection or macroaneurysms that require intervention. Although CTA or MRA may be useful in the detection of FMD in the main renal arteries and main branch arteries, arteriography is necessary for detection of stenoses in smaller arteries.

Treatment for FMD depends on the severity of complications. Pharmacologic treatment alone may be adequate to control hypertension in many patients. Intervention with angioplasty (with or without stenting) or surgery should be considered for patients with severe or difficult to control hypertension or declining kidney function. Angioplasty without stenting is successful in many patients, but recurrent stenosis is not uncommon, and new stenoses from FMD may develop at other sites. For this reason, regular monitoring of blood pressure and serum creatinine levels is essential, and many patients require imaging studies to detect new or recurrent lesions.

Aortic Dissection

Aortic dissection occurs after disruption of the intimal layer of the aorta and propagation of blood flow that dissects along the wall of the aorta, producing a false lumen and compression of the true aortic lumen. Aortic dissection is classified by the site of origin (i.e., DeBakey classification) or the segment of the aorta involved (i.e., Stanford classification). DeBakey type I and II dissections originate in the ascending aorta, and type III dissections originate in the descending aorta. Stanford type A refers to dissections involving the ascending aorta, and type B refers to all others not involving the ascending aorta.

Major branch vessels of the aorta, including the renal arteries, may become obstructed or occluded as a result of extension of the dissection. Aortic dissection frequently compromises the renal arteries (the left more commonly than the right) when it extends into the abdominal aorta and causes renal failure in approximately 20% of patients with type B dissections. When disease is extensive enough to cause AKI, vascular compromise to the intestinal and cerebral vasculature and severe aortic regurgitation often contribute to the high mortality rate.

Aortic dissection most frequently affects older patients (>50 years of age) with coexistent vascular risk factors such as hypertension, smoking, and atherosclerosis. Men are affected more commonly than women. Occasionally, a genetic connective tissue defect such as Marfan syndrome or Ehlers-Danlos syndrome type IV (about 5% of cases) causes aortic dissection, and these conditions should be considered in younger patients (<40 years of age).

AKI occurs in about 20% of patients diagnosed with acute type B aortic dissection and is an independent predictor of in-hospital mortality. Trauma or procedures (e.g., aortic catheterization) can also cause dissection of the aorta or renal artery. Isolated, spontaneous renal artery dissection may rarely occur, most commonly in the setting of polyarteritis nodosa or FMD. Segmental arterial mediolysis is another uncommon condition of unknown origin that is characterized by vacuolar degeneration of smooth muscle cells in the arterial media, which leads to disruption of the arterial medial layer, vessel dissection, hemorrhage, and ischemia. Segmental arterial mediolysis can affect abdominal visceral arteries and virtually any other arterial system.

The most frequent symptom during aortic dissection is chest pain, which may be described as a ripping sensation. Isolated loss of pulse in one or more extremities may provide a clinical clue, and the number of arteries involved correlates with the severity of dissection. A common clue to the diagnosis on a routine chest radiograph is a widened mediastinum, with or without a pleural effusion (most often on the left).

After the diagnosis of aortic dissection is established, evaluation of renal artery involvement is best undertaken noninvasively to minimize further vascular injury. Contrast-enhanced CT, magnetic resonance imaging (MRI), or MRA usually provide images capable of confirming or excluding renal involvement, although each modality carries the same limitations as outlined for the evaluation of RAS. Transesophageal echocardiography is useful for establishing the diagnosis of aortic dissection, but it does not provide information about the aorta below the diaphragm. Renal duplex ultrasonography may be useful for evaluating renal