Fibromuscular Dysplasia of Renal and Carotid Arteries

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Abstract

We report here two cases of fibromuscular dysplasia (FMD). The first case describes an asymptomatic 75-year-old man with FMD of the right internal carotid artery. The second case reports a 17-year-old man who presented with arterial hypertension caused by FMD of the left renal artery and was subsequently successfully treated by angioplasty. FMD is a rare nonatherosclerotic, noninflammatory angiopathy, which can involve almost every arterial vascular bed. It is a less common cause of stenosis of renal and carotid arteries. FMD can present with arterial hypertension when it involves renal arteries or with ischemic stroke or transient ischemic attack when the disease affects the carotid or vertebral arteries. Many cases are asymptomatic and may be discovered incidentally. Percutaneous transluminal angioplasty should be used in patients with a stenosis of the renal artery causing arterial hypertension. On the contrary, conservative therapy should be chosen in patients with asymptomatic and extensive lesions of the carotid arteries.

Keywords
- fibromuscular dysplasia
- "string-of-beads" sign
- stenosis
- percutaneous transluminal angioplasty
- arterial hypertension

Although the most common cause of stenosis in renal and carotid arteries is atherosclerosis, fibromuscular dysplasia (FMD) should be considered in the differential diagnosis of patients without the classical risk factors for atherosclerosis. We present here two cases of patients with FMD and a favorable long-term follow-up.

Case Report 1

A 75-year-old asymptomatic patient, with a low cardiovascular risk, underwent ultrasonography of the carotid arteries because of a murmur along the right carotid artery. A stenosis of the right internal carotid artery (RICA) was diagnosed. Carotid angiogram revealed a long stenosis of RICA spreading from the origin of the artery with the typical "string-of-beads" sign that is characteristic for FMD consisting of alternative areas of narrowing and dilation (►**Fig. 1**).[1] As the patient was asymptomatic and efficient brain circulation was provided by collaterals, conservative therapy was chosen with a favorable 4-year follow-up.

Case Report 2

A 17-year-old patient newly diagnosed with arterial hypertension underwent magnetic resonance and dynamic scintigraphy of the kidneys. A single left renal artery stenosis was revealed (►**Fig. 2A**). Renal angiogram was performed and confirmed a bifurcation stenosis of the left renal artery. Percutaneous transluminal angioplasty (PTA) of the stenotic lesion was successfully performed (►**Fig. 2B**). Subsequently, during a 4-year follow-up, arterial hypertension was not demonstrated.

Discussion

FMD is a nonatherosclerotic, noninflammatory angiopathy of unknown etiology that may involve any layer of a visceral

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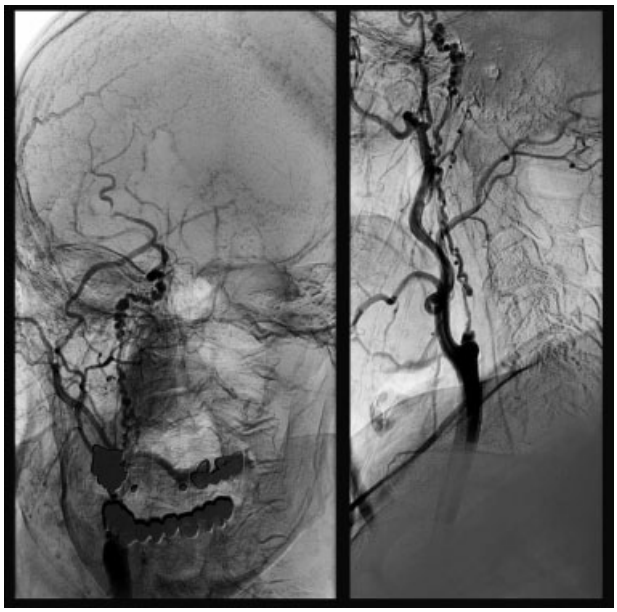

Fig. 1 Angiography of the right carotid artery. Medial fibromuscular dysplasia of right internal carotid artery with the typical "string-of-beads" sign. Diameter of the "beading" is larger than the diameter of the affected artery which is typical for medial fibroplasia. In perimedial fibroplasia, the diameter of the "beading" is generally smaller than the diameter of the artery involved.

cervicocranial arteries (range, 25–30%), visceral arteries (9%), and the arteries of the extremities (5%),[3] rarely also coronary arteries.[4] Clinical manifestations reflect the arterial bed involved. The most common symptoms are resistant hypertension (lesion of a renal artery), headache, tinnitus, dizziness or stroke (lesions of cervicocranial arteries), leg pain (involvement of lower limb arteries), and acute coronary syndrome (lesion of coronary arteries). Noninvasive methods such as computed tomography and ultrasound can help to make the diagnosis. Catheter angiography or computed tomography angiography is considered the gold standard for confirming the diagnosis. The differential diagnoses include atherosclerotic stenosis and stenosis associated with vasculitis or vascular Ehlers–Danlos or Williams syndromes. FMD can be differentiated angiographically from atherosclerosis as it occurs in the middle or distal portion of the vessel whereas atherosclerosis occurs proximally. The treatment of FMD is individually based on the patient's symptoms, location, and significance of the stenosis. The treatment options include medication, PTA, or surgical revascularization. Conservative therapy should be chosen in patients with an asymptomatic stenosis of a carotid artery or when the anatomical structure of the carotid artery is not suitable for revascularization.[5] Although there are no randomized data available, there is an increasing body of evidence that stenting might be the first

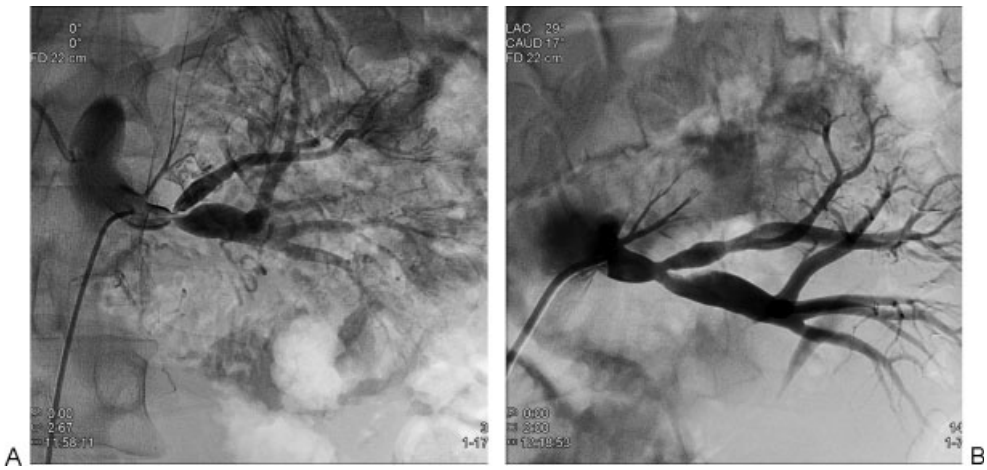

Fig. 2 (A) An angiographic image of the left renal artery before PTA. The image reveals fibromuscular dysplasia of the left renal artery. The smooth focal stenosis is typical of intimal fibroplasia. (B) An angiographic image of the left renal artery after PTA. After PTA of severe stenosis, the left renal artery has a satisfactory appearance on angiography. PTA, percutaneous transluminal angioplasty.

artery. The prevalence of FMD is not precisely known, but according to the CORAL trial the prevalence of renal artery FMD is approximately 2.3%.[2] It is histologically classified into intimal, adventitial, and (the most common) medial fibroplasia. Intimal fibroplasia may be angiographically presented by a smooth focal stenosis or long-tubular stenosis, as opposed to the medial fibroplasia that is usually presented with multiple stenoses which appear as a "string-of-beads." FMD lesions may progress to severe arterial stenosis, occlusion, aneurysm, or dissection, causing organ ischemia or infarction. This generally involves renal arteries (range, 60–75%), therapeutic option in symptomatic patients with FMD and intracranial stenosis.[6] In patients with stenosis of a renal artery causing arterial hypertension, the PTA should be performed. This may lead to normalization of blood flow in the kidneys and a reduction of arterial hypertension.

Conclusion

FMD is ranked among the less common causes of stenoses of carotid and renal arteries. The PTA should be performed in patients with a stenosis of the renal artery causing arterial

hypertension. On the contrary, conservative therapy may be appropriate in patients with asymptomatic and extensive lesions of the carotid arteries.

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