
Percutaneous Transluminal Angioplasty of Subclavian Stenosis from Neurofibromatosis

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Summary: We report a case of successful percutaneous transluminal angioplasty of a stenotic left subclavian artery caused by vascular involvement of neurofibromatosis type 1.

Index terms: Neurofibromatosis; Arteries, transluminal angioplasty

Vascular involvement in neurofibromatosis type 1 occurs in approximately 9% of patients in autopsy series (1). Vascular symptoms are usually from renal artery stenosis or cerebral arterial occlusion. Treatment of symptomatic vascular neurofibromatosis has been by medical and surgical means as well as more recently by percutaneous transluminal angioplasty (PTA) (2, 3). We report a case of stenosis of the left subclavian artery treated successfully with angioplasty.

Case Report

The patient is a 9-year-old white boy with the diagnosis of neurofibromatosis type 1. Because of learning disabilities and recurrent headaches, he underwent cranial magnetic resonance imaging, which demonstrated retrograde flow in the dominant left vertebral artery. The patient was neurologically intact by examination. He underwent cerebral angiography, which demonstrated complete occlusion of the left common carotid artery, with collateral supply from the right external carotid artery and across the anterior communicating artery from the right. There was a high-grade stenosis of the proximal left subclavian artery with slow, retrograde flow in the left vertebral artery (Fig 1A). Given the patient's underlying disease process and involvement of the left carotid system, he returned 1 month later for PTA of the left proximal subclavian artery using the femoral artery route. Initial mean pressure gradient across the stenosis was 40 mm Hg. PTA was first performed with a 4-mm balloon, followed by a 5-mm balloon after angiography demonstrated residual stenosis

after inflation of the 4-mm balloon. Pressure gradients after angioplasty were 4 mm Hg mean. Postangioplasty angiography demonstrated a patent angioplasty site with antegrade flow in the left vertebral artery. There was, however, an intimal flap at the angioplasty site (Fig 1B). Preangioplasty brachial blood pressures were 86/35; postangioplasty pressures were 96/53. No neurologic complications were encountered and the patient remained neurologically intact. Because of several episodes of discrepant blood pressure between arms and the known intimal flap at the angioplasty site, the patient returned to angiography 3 months later and had routine follow-up angiography 8 months after angioplasty. There was excellent flow in the left subclavian artery and the angioplasty site was widely patent in both angiograms (Fig 1C and D). Pressure gradients across the site were unchanged, and discrepant blood pressures greater than 10 mm Hg systolic have not been measured over the 10-month follow-up interval.

Discussion

This case brings to focus two important questions. First, should retrograde vertebral artery flow (ie, vertebral steal) be treated, and if so, when? Second, if treatment is deemed necessary, given the location of the lesion and neurofibromatosis as the cause, should percutaneous transluminal angioplasty be the treatment of choice?

Much has been written about the need for treatment of vertebral steal, particularly when asymptomatic (4, 5). Symptomatic involvement resulting from vertebrobasilar insufficiency is most likely caused by decreased cerebral perfusion resulting from multiple stenotic or occluded arteries rather than an isolated subclavian stenosis (5, 6). In the case presented here, the decision to proceed with treatment

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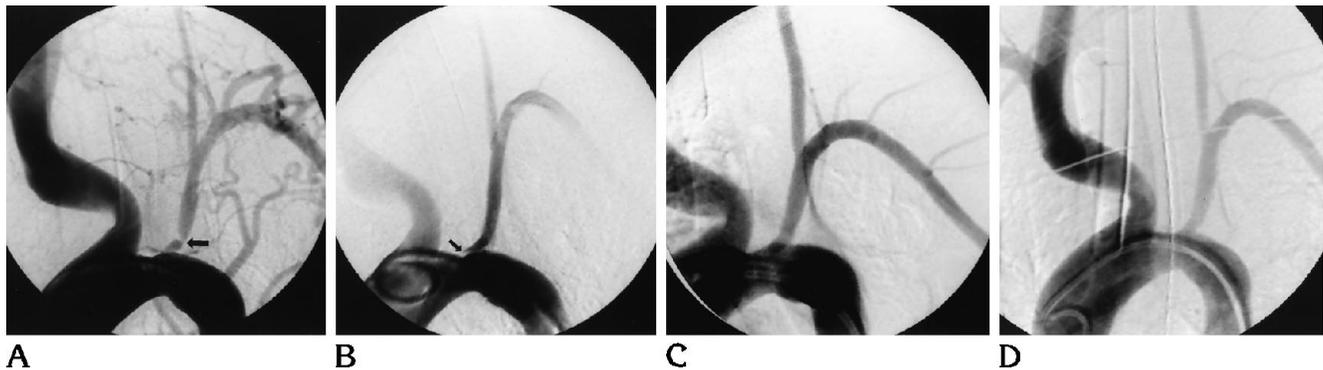


Fig 1. A, Preangioplasty aortogram. The left common carotid artery is occluded at its origin. There is a stenosis of the proximal left subclavian artery (*arrow*), with some filling of the left vertebral artery. This flow, however, reverses after injection and is in a retrograde pattern. Note also the somewhat small, presumably diseased aorta.

B, Postangioplasty aortogram. The angioplasty site is widely patent, but there is an intimal flap (*arrow*). Flow in the left vertebral artery is antegrade and as rapid as flow in the subclavian artery.

C, Follow-up aortogram 3 months after angioplasty. Intimal flap has healed and the artery is widely patent with excellent antegrade vertebral flow.

D, Follow-up aortogram 8 months after angioplasty. The left subclavian artery origin continues to be widely patent with excellent flow.

was based on the known left common carotid artery occlusion, the underlying cause, and the potential problems that could develop particularly in light of the patient's young age.

When treatment is considered necessary, the form of such treatment is also somewhat controversial. Cerebral vascular involvement in neurofibromatosis often manifests itself as stenoses, and the sites may not be amenable to easy or successful surgical repair. Angioplasty offers a viable treatment option in this subset of patients. PTA of great vessels is a relatively new technique, but is showing great promise particularly when applied to the subclavian artery (7, 8). Angioplasty of neurofibromatosis however has had mixed results, but limited reports are available in the literature (2, 3, 9). The pathogenesis of arterial lesions in neurofibromatosis is still unknown, and of course pathological data on the lesion are not available in this case. The arterial lesions in neurofibromatosis were initially thought to represent a proliferation of Schwann cells, with secondary degenerative changes resulting in lesions with different appearances (10). More recently, it has been postulated that the arterial changes may be related to a primary defect in the smooth muscle cells of the vessel much like in fibromuscular hyperplasia (11). Therefore, the fact that PTA was successful in this case should not be surprising, because it is well known that fibromuscular hyperplasia responds well to angioplasty (12).

The subclavian stenosis has responded well to angioplasty over the 10-month follow-up interval. Follow-up angiography with pressure measurements at both 3 and 8 months was probably unnecessary, but does provide definitive proof of vessel patency. The improved appearance at follow-up angiography is not surprising given the nature of angioplasty sites to remodel. To that end, it is quite common for lesions with even large dissections after PTA to heal completely and in a widely patent state, especially when dealing with causes other than atherosclerosis (13). In addition, delayed response of neurofibromatosis to renal PTA has also been reported by Gardiner et al (3).

In conclusion, angioplasty appears to be a definite alternative to surgery when treatment for subclavian stenosis is indicated; this treatment was safely applied to neurofibromatosis in a single case.

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