Bilateral Subclavian Steal: A Review of an Unusual Twist in a Common Disorder

Kenneth A. Giles and Virginia C. Poirier

Summary: The authors present two cases of bilateral subclavian steal syndrome, a rare condition that does not commonly cause neurovascular symptoms. Lateralizing hemispheric events occur usually with carotid lesions. Vertebrobasilar insufficiency is three times more common in bilateral than in unilateral subclavian steal syndrome. Arm-exercise-induced brain-stem dysfunction is rare, and is seen only in bilateral subclavian steal syndrome.

Index terms: Arteries, subclavian; Atherosclerosis; Arteries, magnetic resonance

Subclavian steal syndrome (SSS), first described by Contorni in 1960 (1), is a syndrome secondary to occlusive disease in the proximal subclavian artery. Blood supplied to the arm is sustained via reversal of flow in the ipsilateral vertebral artery. The retrograde flow in the vertebral artery is supplied by steal from the contralateral vertebral and/or basilar artery. Bilateral cases of SSS are very unusual (2-8), reported cases have been almost exclusively unilateral.

We present the clinical and angiographic findings in two cases of bilateral SSS, both of which were due to atherosclerotic disease.

Case Reports

Patient A

This 50-year-old white woman complained of bilateral shoulder pain with numbness and tingling of her arms, followed by dizziness, vertigo, and “blackouts.” Physical exam demonstrated a 3+/4 left carotid bruit. Pulses were diminished throughout the upper and lower extremities. Duplex carotid ultrasound demonstrated a 50%-75% stenosis of the left common carotid artery, occlusion of the right internal and external carotid arteries, an occluded left subclavian artery, and reversal of flow in both vertebral arteries. The aortic arch angiography confirmed complete occlusion at the origin of the innominate and left subclavian arteries. At the origin was 50% stenosis of the left common carotid artery, which was the only source of intracerebral circulation (see Fig. 1). Retrograde flow was seen in both vertebral arteries, perfusing the upper extremities (see Fig. 2). Collaterals to the right arteries were seen via costocervical and thyrocervical trunks fed by intercostal arteries off the arch. Collaterals to the left subclavian artery were noted from the thyrocervical trunk fed by external carotid artery and vertebral artery muscular branches. A surgical repair was performed with a bifurcated interposition dacron bypass graft from the right lateral ascending aorta to the innominate bifurcation with another anastomosis to the left subclavian artery. At 1 month follow-up, the patient complained of one transient ischemic attack manifested as left upper extremity weakness. She was no longer experiencing the bilateral upper-extremity symptoms or blackout spells that had been present preoperatively.

Patient B

This 67-year-old white woman had a past medical history of severe atherosclerotic disease, presenting clinically as episodic vertigo necessitating a right carotid endarterectomy in 1984. She had also had a left to right subclavian artery bypass graft for decreased flow to the right arm in 1984, which was revised in 1989 and ultimately failed. Upon admission, physical exam revealed no palpable pulses in the upper extremities and diminished pulses in the lower extremities. She was unresponsive to verbal and visual stimuli and had a left-sided hemiplegia. Her computed tomography scan of the brain showed generalized atrophy and a subacute right cortical infarction. Angiography was performed and demonstrated occlusion of the innominate and left subclavian arteries with a 50% stenosis of the left common carotid artery all at their origins (see Fig. 3). Bilateral subclavian steal via retrograde flow in both vertebral arteries was noted. Collateral vessels to the right subclavian artery were seen off the aortic arch with collaterals from the external carotids to the thyrocervical trunk (Fig. 4). Surgical repair of these vascular occlusions was not performed because of her severe neurologic deficits. The patient is currently in a demented state, at a nursing facility.
Discussion

An atherosclerotic lesion at the orifice of a subclavian artery is the most common etiology of SSS. These stenoses usually fibrose without any tendency to ulcerate or form thrombi (9). Stenotic or preocclusive disease of the proximal vertebral artery can also cause steal-type symptoms if there is a reversal of flow. Patients with SSS also tend to develop atherosclerotic disease in the coronary and cerebral arteries. Other etiologies of SSS include metastatic carcinoma, arteritis (especially Takayasu disease) (10), vascular thrombosis due to emboli, and following surgical bypass shunts (2). Congenital subclavian steal is usually associated with major cardiac anomalies such as atrial septal defect, ventricular septal defect, patent ductus arteriosus, tetralogy of Fallot, and aortic coarctation (2).

Symptoms of SSS include pain, numbness, or fatigue in the involved arm during use of the upper extremity. A blood pressure difference of greater than 20 mm Hg systolic between the upper extremities is seen in unilateral SSS (11). However, this discrepancy may not be present if there is bilateral disease, as in the two cases presented here. The occurrence of neurologic events depends on additional stenosis of the ca-
rotid arteries, patenty of the circle of Willis, the patient’s general cardiovascular condition, and the functional demand of the affected arms. Neurologic signs and symptoms of vertebral-basilar insufficiency such as vertigo, diplopia, and ataxia may be provoked by prolonged muscular effort with the affected arms.

In one study of 324 patients with SSS (9), 64% of patients had no neurologic symptoms. Lateralizing hemispheric cerebral vascular events occurred most frequently in patients with coincidental occlusive carotid lesions, suggesting that inadequate collateral circulation is likely to be the determining factor for neurologic defects. Bilateral SSS produces nonhemispheric central nervous system events from vertebral-basilar insufficiency more often than unilateral SSS by a factor of 3 to 1 (9). Arm exercise-induced brain stem dysfunction is extremely rare and is seen only in patients with bilateral SSS (9). Of 16 cases of bilateral SSS in the same series, 56% were neurologically asymptomatic, 31% had lateralizing hemispheric events, and only 13% had nonhemispheric events. In one review of over 100 cases of SSS, there was not a single case of brain stem infarction (12). Similarly, in the study by Hennerici, none of the 324 patients suffered a brain stem infarction. Transient ischemic attacks in the carotid distribution are far more common in these patients, which was explained by basilar-carotid artery steal via the circle of Willis with poor collateralization.

Diagnosis of SSS can be suggested by retrograde flow in the vertebral arteries on duplex ultrasound scanning (13, 15). Magnetic resonance angiography may become an accurate means of noninvasively demonstrating this entity; it can demonstrate the diminished or reversed flow in the vertebral artery (Fig. 5). Superior and inferior saturation pulses will confirm the reversal of flow (Figs. 6 and 7). Superior saturation pulses will cancel signal in the caudal flowing vessels, leaving signal visualized in the cephalad flowing vessels. Inferior saturation pulses will do the opposite, leaving only signal in the caudal flow. Care must be taken to perform both saturation pulses. For example, loss of vertebral artery signal with a superior pulse only may lead to the false conclusion of total occlusion or extremely slow flow. Also, in some spin-echo sequences, thrombus may or may not have signal. Currently, the diagnosis of SSS is generally made by contrast angiography and is still desired preoperatively for anatomical mapping.

Therapy of SSS has generally been surgical vascular bypass grafting; however, more recently, percutaneous transluminal angioplasty (PTA) has been attempted. In a series of 45 patients with SSS treated by PTA of the subclavian artery, two thirds benefitted by the treatment. Fifteen of these patients had bilateral occlusive lesions of the extracranial vertebral arteries and received PTA of the proximal vertebral arteries. Eight of these patients had a marked improvement of both subjective and objective clinical symptoms following vertebral PTA. Post-PTA occlusion was observed only in two of the 15 cases over a 2- to 25-month observation (14).
Conclusion

We have presented two cases of bilateral SSS. The diagnosis may be suspected on the basis of duplex ultrasound examination, which documents retrograde flow in the vertebral arteries. It can also be demonstrated with magnetic resonance angiography. Contrast angiography is still the gold standard and is used for a definition of anatomy.

Reversal of blood flow in the vertebral arteries is usually a benign vascular disorder that only occasionally produces cerebral vascular events. The latter is more commonly due to coexisting severe carotid obstruction or is the result of insufficient collateralization via the circle of Willis. Arm exercise-induced brain stem dysfunction is generally seen only in patients with bilateral SSS and is relatively rare.

References

10. Agee OF. Two unusual cases of subclavian steal syndrome. AJR 1966;2:447–457