Pearls & Oysters: Symptomatic innominate artery disease

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PEARLS

1. Innominate artery (IA) disease is an uncommon cause of stroke. A comprehensive clinical evaluation can promptly provide important diagnostic information and help tailor a straightforward diagnostic strategy.
2. The tetrad of right arm, eye, and hemispheric and posterior circulation ischemia is highly suggestive of an IA lesion.
3. Revascularization may be safely performed using an endovascular approach.

OY-STER

1. Unlike subclavian steal syndrome (SSS), IA disease is more frequently associated with symptoms of cerebral and retinal ischemia, and revascularization should be strongly considered in symptomatic patients.

CASE REPORT A 64-year-old man presented to the emergency department with left-sided weakness and speech impairment upon awakening. He had a history of multiple myocardial infarctions and had undergone coronary angioplasty and coronary artery bypass graft surgery 1 year previously. He was a heavy smoker and had arterial hypertension. He was taking aspirin 100 mg QD, clopidogrel 75 mg QD, simvastatin 40 mg QD, and enalapril 5 mg BID.

The neurologic examination revealed left-sided hemiparesis, dysarthria, and neglect. The NIH Stroke Scale score was 11. Simultaneous palpation of pulses in the 2 arms revealed an absent right radial pulse. Blood pressure was 120/80 mm Hg when measured on the left arm and 80/40 mm Hg on the right arm (arm blood pressure differential = 40 mm Hg). An MRI showed infarctions in the middle cerebral artery territory, consistent with borderzone and embolic lesions (figure 1A).

Echocardiogram revealed an ejection fraction of 0.25, pulmonary hypertension (estimated pulmonary artery systolic pressure, 81 mm Hg), and apical akinesis. Duplex ultrasound showed retrograde flow in the right vertebral artery (VA) and a parvus et tardus

pattern in the right common carotid artery (CCA). Reverse flow in the vertebral artery and reduced flow in the CCA were also observed in 2D time-of-flight MRI (figure 1B). Collateral supply through the anterior communicating artery was detected by transcranial Doppler.

A diagnosis of innominate artery occlusive disease was suspected and confirmed by contrast-enhanced CT angiography and magnetic resonance angiography (MRA) (figure 1C). Digital subtraction angiography showed subocclusion of the brachiocephalic trunk (IA) (figure 1E) and a mild stenosis in the left internal carotid artery. There was retrograde flow in the right vertebral artery; both right CCA and subclavian arteries were partly supplied by the ipsilateral vertebral artery (figure 1D).

Because of neurologic stability after the initial event, the patient was maintained on a conservative treatment strategy with mandatory bed rest in flat bed, suspension of antihypertensive medications, high-dose atorvastatin, and dual antiplatelet therapy with aspirin and clopidogrel.

On the subsequent week he had several spells of dizziness and amaurosis fugax of the right eye, not related to clinical deterioration or hypotension. Dizziness could be elicited by exercising the right arm. The persistence of these symptoms of eye and posterior circulation ischemia suggested either ongoing embolization or hemodynamic compromise in the IA artery territory. Angioplasty and stent placement in IA were then performed (figure 1F). The patient was discharged without transient or permanent new neurologic symptoms 5 days after the procedure.

DISCUSSION This patient had innominate artery stenosis, an entity that resembles subclavian steal phenomenon (SSP) in some aspects. His upper limb pulses and blood pressures were asymmetric, and there was retrograde flow in the right VA in the neck. SSP is generally associated with subclavian artery stenosis, and is usually caused by atherosclerosis, though it has also been associated with Takayasu arteritis.¹ Although SSP is somewhat common, most patients are

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Figure 1 MRI, magnetic resonance angiography (MRA), and digital subtraction angiography (DSA) imaging before and after angioplasty

(A) MRI diffusion-weighted imaging shows diffusion restriction in the right frontal operculum and centrum semiovale (arrowheads). (B) 2D time-of-flight (2D TOF) MRI: this flow-direction-sensitive acquisition shows absence of signal in the right vertebral artery (VA) which, in association with a patent vertebral artery on contrast-enhanced MRA (C), suggests reverse flow. The lower signal intensity in 2D TOF in the right carotid artery is suggestive of reduced flow. The innominate artery (IA) stenosis is shown in MRA (arrow in C). DSA: (D) Sequential imaging of the extracranial vessels following dye injection in the left subclavian artery demonstrates anterograde flow in the left subclavian artery (arrow 1) and VA (arrow 2) followed by retrograde flow in the right VA (arrow 3) and late perfusion of the right subclavian (arrow 4) and common carotid (arrow 5) arteries. Subocclusion of the IA is observed after dye injection at its origin (arrow in E). (F) Postprocedural angiography shows adequate stent placement (arrowhead) and anterograde flow in the IA and the right VA (arrows).

essentially asymptomatic,² and the phenomenon may be considered a marker of generalized atherosclerosis.³ Symptomatic disease (SSS) has been associated with higher arm blood pressure differentials.² In symptomatic disease, clinical presentation may involve upper limb claudication and transient symptoms related to the posterior circulation (ataxia, vertigo, diplopia, syncope). Neurologic symptoms may occur at rest or be elicited by ipsilateral arm exertion. The presence of anterior circulation symptoms is unexpected and largely associated with comorbid carotid artery disease.³ IA disease should be included in the differential diagnosis of SSP.

Subclavian and IA stenosis have some similarities (table). In both entities, large-vessel high-degree stenosis proximal to the VAs leads to the clinical picture of diminished arterial pulses in the ipsilateral arm, high arm blood pressure differential (usually greater than 20 mm Hg), reverse flow in the ipsilateral VA, and, occasionally, posterior circulation–related symptomatology.

IA stenosis is nevertheless unique, since the obstruction is proximal to both the right VA and CCA (figure 2). Presentation may contemplate all the findings usually imputed to SSS, with the addition of anterior circulation ischemia (table). An “array of ipsilateral arm and eye ischemia, accompanied by anterior and posterior circulation ischemia (or both)”⁴ is highly localizing to the brachiocephalic trunk. The case we presented is a clear example of this clinical constellation. Although definite diagnosis required vascular imaging (duplex ultrasound, MRA, and angiography), IA stenosis was suspected in the first evaluation.

The exact incidence of IA occlusive disease is unknown, but seems to be lower than that of SSP. Old angiographic studies suggest that it may account for 2.5% of occlusive lesions of extracranial and intracranial arteries.⁵ Brunhölzl and von Reutern identified only 20 cases of double steal phenomenon among 30,000 (<0.1%) patients referred for duplex ultrasound.⁵ IA occlusive disease seems to be highly associated with

Table	Clinical features of great aortic arch branch occlusive lesions			
	Carotid artery	Vertebral artery	Subclavian artery	Innominate artery
Eye ischemia	+	–	–	+ ^a
Hemispheric stroke/TIA	+	–	–	+ ^a
Upper limb ischemia	–	–	+	+ ^a
Posterior circulation symptoms	–	+	+	+

^a Right-sided.

smoking and hypertension, and not so much with diabetes or abnormal blood lipid levels.⁶ The average age in published case series was relatively young (around 50–60 years).^{6,7}

The prevalence of neurologic symptoms in IA disease may be higher than in SSP. In the series of Brunhölzl and von Reutern, 13/20 patients with ultrasound findings suggestive of IA stenosis had neurologic

symptoms. Three patients had right hemispheric TIAs and one a right hemispheric stroke. The remaining patients had symptoms of vertebrobasilar insufficiency.⁵ Grant et al.⁷ described neurologic symptoms in 8/12 patients in a similar series. Two had spells of amaurosis fugax involving the right side, and 2 had right hemispheric strokes. The remaining 4 patients had syncope or cerebellar infarcts. Among 37 patients referred for surgery for occlusive IA disease in the series of Brewster et al.,⁶ 30 had neurologic symptoms. Twelve had amaurosis fugax and 10 had symptoms of vertebrobasilar insufficiency. The prevalence of hemispheric stroke or TIA was relatively low.

Although a significant proportion of patients with IA disease are asymptomatic, the presence of neurologic symptoms may pose a complex clinical scenario. Large-vessel atherosclerosis is a systemic disease and patients may have severe comorbidities that affect clinical decision-making. Most studies about interventional treatment with either percutaneous angioplasty or surgery were designed to address intervention outcomes (primary success rates, late patency rates) rather than stroke recurrence or disability. Asymptomatic patients or patients with a wide array of symptoms such as arm ischemia, stroke, or myocardial ischemia were included.^{7–9} Neurologic outcomes were not studied in detail.

Before the advent of percutaneous techniques, open surgery was the standard therapy for IA lesions, with a fairly acceptable surgical risk. Among 37 patients with IA occlusive disease who underwent surgical repair with different techniques, 1 (3.4%) died and 2 (6.9%) had minor strokes.⁷

Angioplasty and stenting are interesting alternatives to high-risk open surgery in IA disease. Periprocedural stroke rates may be as low as 0%⁸ or 1.1%,⁹ and endovascular treatment seems to be an overall safe treatment for IA stenosis.

This apparent superiority of percutaneous treatment is in agreement with the findings of nonsystematic reviews of patients with miscellaneous supra-aortic lesions. Hadjipetrou et al.¹⁰ reviewed previous series of patients with either subclavian or IA lesions that were submitted to percutaneous stenting or surgical treatment. The risk of stroke was estimated to be 3% in surgical and 0% in percutaneous stenting series. Furthermore, mortality was found to be 2% after surgery but 0% after stenting.¹⁰

Beside evaluation with careful assessment of symptoms and signs of posterior circulation, anterior circulation, and eye and arm ischemia is key to diagnosis of IA disease, a treatable cause of stroke.

AUTHOR CONTRIBUTIONS

Dr. Guedes: study concept, analysis, literature review, and initial draft. Dr. Valeriano: acquisition of data, critical revision of manuscript for intellectual content. Dr. Puglia Jr: critical revision of manuscript for intellectual content. Dr. Arantes: acquisition of data, analysis, critical revision of

Figure 2 Innominate artery (IA) occlusive disease

Blood from the aorta flows cranially in the left vertebral artery (VA), then caudally in the right VA (arrows), leading to posterior circulation symptomatology. Perfusion of the right subclavian and internal carotid artery (ICA) is partly provided by the right VA, which explains low blood pressure in the right arm and ICA. Embolization from the plaque in the innominate artery to the anterior circulation is responsible for strokes and episodes of amaurosis fugax. BA = basilar artery; MCA = middle cerebral artery; OA = ophthalmic artery; PCA = posterior cerebral artery.

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