Stroke Caused by Arterial Thoracic Outlet Syndrome in an Adolescent

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Abstract
Arterial thoracic outlet syndrome is a rare condition characterized by compression of the subclavian artery, often with post-stenotic aneurysm formation. Artery-to-artery embolic strokes related to thoracic outlet syndrome have been reported in the posterior circulation and in the ipsilateral anterior circulation. We present a case in which a thrombus secondary to thoracic outlet syndrome caused a contralateral anterior circulation stroke in an adolescent and postulate mechanisms of this rare occurrence. This case demonstrates that a subclavian thrombus due to thoracic outlet syndrome can take a circuitous path and cause an anterior circulation stroke contralateral to the diseased subclavian artery. In addition, this case illustrates the importance of a high index of suspicion for thoracic outlet syndrome in patients with stroke and associated arm pain or discoloration.

Keywords
Brain, children, neuroimaging, pediatric, stroke

Introduction
Thoracic outlet syndrome is a rare condition caused by compression of the neurovascular structures in the thoracic outlet. The signs and symptoms that occur when the subclavian artery is compressed are referred to as arterial thoracic outlet syndrome (aTOS), which most commonly occurs secondary to a cervical rib.1 Thrombosis in the setting of aTOS may present with clinical manifestations of antegrade embolization, including loss of peripheral pulses, finger discoloration, and arm claudication. More rarely, aTOS can cause retrograde thromboembolism from the subclavian artery, resulting in stroke. Ischemic stroke related to aTOS has been reported in both adults and children in the posterior circulation and in the ipsilateral anterior circulation,1–5 consistent with the postulated mechanism of retrograde thrombus propagation to the vertebral or ipsilateral carotid artery, which may be exacerbated by arm abduction. Here, we present a case of TOS-related stroke in the contralateral anterior circulation in an adolescent that raises mechanistic considerations of embolus path to the cerebral vasculature and highlights the importance of a high index of suspicion for this rare cause of stroke in the young.

Case Report
A 16-year-old young woman presented to our facility with acute right hemiparesis and hemianesthesia. She had been healthy until 6 months prior to the presentation, when she developed intermittent right forearm pain and discoloration of fingers on the right hand resulting in diagnoses of tendonitis and unilateral Raynaud’s phenomenon. Three weeks prior to the current presentation, she developed worsening right upper extremity pain and arm discoloration. On presentation, she had acute right hemiparesis with an NIHSS score of 5. Carotid Doppler showed hypechoic lesions of the left internal carotid artery. MRI of the brain revealed a recent infarct in the left MCA region with extension into the left ACA territory. Ultrasound of the upper limbs and lower extremities showed a right subclavian artery thrombus with retrograde propagation to the right vertebral artery and left common carotid artery. Angiography confirmed a thrombus in the right subclavian artery that traveled through the right vertebral artery into the left common carotid artery, causing a left MCA and ACA territory ischemic stroke. The patient was started on antiplatelet therapy and referred for surgical resection of the cervical rib. She had a good recovery with minimal residual right hemiparesis. The patient was eventually discharged with recommendations for physical therapy and ongoing antiplatelet therapy.

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presentation, she presented to an outside hospital with left arm numbness. Head computed tomography (CT) was normal at that time and she was diagnosed with a functional neurological disorder. During the current presentation, the general examination revealed diminished right radial pulse. Neurologic examination was significant for moderate receptive and expressive aphasia, dysarthria, right-sided numbness, and right hemiparesis affecting the arm and face more than the leg (with no movement of the right arm and minimal effort against gravity of the right leg). Initial NIH Stroke Scale was 12. Emergent brain magnetic resonance imaging showed diffusion restriction within the left basal ganglia, a terminal internal carotid artery (ICA)/M1 occlusion, a large perfusion deficit in the left middle cerebral artery territory, and a chronic right thalamic infarct (Figure 1A–C). She underwent emergent thrombectomy of the left ICA terminus thrombus with TICI3 recanalization.

After thrombectomy, right upper extremity Doppler ultrasound was obtained due to diminished radial pulse, revealing diminished Doppler arterial waveforms consistent with thrombus in the axillary artery with occlusion of the brachial, radial and ulnar arteries. Doppler ultrasound and exam of left upper extremity was normal. Thrombectomy of the right arm was performed, with findings consistent with chronic thrombosis, and she was started on a heparin infusion. Negative antiphospholipid antibody testing excluded acquired severe thrombophilia syndrome, increasing suspicion for mechanical source of thrombosis. Chest CT angiogram demonstrated bilateral C7 cervical ribs with associated high-grade stenoses of the bilateral subclavian arteries, right greater than left (Figure 1D). She was diagnosed with thoracic outlet syndrome and underwent right cervical rib removal. Anticoagulation was continued with rivaroxaban as an outpatient, and 3 months later, the patient underwent removal of the left cervical rib. Several weeks postoperatively, antithrombotic therapy was transitioned from rivaroxaban to aspirin. Five months after the stroke, the patient exhibited significant recovery, with no cognitive, language, or sensory deficits. She has mild tone elevation of the right arm, nearly full strength of her right side, with only mild weakness of finger extension and forearm supination on the right, and mild right arm hyperreflexia with a positive Hoffman’s sign. Pediatric stroke outcome measure (PSOM) is 1 (right sensorimotor) at 5 months.

Discussion
Arterial TOS is a rare condition characterized by compression of the subclavian artery, often with post-stenotic aneurysm formation, frequently due to a cervical rib. Artery-to-artery embolic strokes related to aTOS have been reported in the ipsilateral and contralateral posterior circulation and in the ipsilateral anterior circulation, fitting with the postulated mechanisms of either retrograde thrombus propagation to the vertebral or ipsilateral carotid artery or transient retrograde flow within the subclavian artery exacerbated by arm abduction. Thrombus dislodgement can occur from a combination of pulsatile high pressure forward flow, as well as direct compression and mechanical disruption by the adjacent cervical ribs. One could also theorize a water hammer effect, whereby the momentum of the blood within the artery against a fixed obstruction results in marked flow derangements, including transient retrograde flow due to pressure reversals. Retrograde blood flow in the subclavian artery has been previously demonstrated in cases of aTOS using Doppler ultrasound. In our patient, we suspect that repetitive arm abduction in a base position during cheerleading may have provoked subclavian artery occlusion and retropropulsion of clot.

The present case is the first report in which a thrombus secondary to aTOS caused a stroke of the contralateral anterior circulation. We propose that the clot traveled from the right subclavian artery to the right vertebral and basilar arteries, then to the left PCA, and across the left posterior communicating artery, ultimately lodging at the left carotid terminus (Figure 2). Though the patient had bilateral cervical ribs, there was no evidence of thrombosis in the left arm (including normal Doppler ultrasound and physical examination) and there was no post-stenotic aneurysm on the left, making it unlikely that the left subclavian stenosis was contributory to the presentation. Furthermore, our patient’s symptoms three weeks prior to presentation, which were diagnosed as a functional neurological disorder at an outside hospital,

Figure 1. Panels A (axial DWI), B (axial ADC) and C (axial T2 FLAIR) are brain magnetic resonance imaging sequences illustrating restricted diffusion compatible with an acute infarct involving the left globus pallidus, corona radiata, and the body of the left caudate nucleus. Panel D is a 3D reconstruction image of CT angiography of the chest illustrating high-grade stenosis of the right subclavian artery with post-stenotic aneurysmal dilatation.
were in retrospect due to a thalamic stroke identified on neuroimaging, further supporting our proposed clot path through the posterior circulation. Prior to this report, aTOS-related anterior circulation stroke has only been reported ipsilateral to the diseased subclavian artery, suggesting that the thrombus passes through the ipsilateral carotid artery, instead of through the posterior circulation, in which case strokes would occur in equal frequency ipsilaterally and contralaterally, as they do in the posterior circulation. In this case, we propose that aTOS-associated emboli may traverse the posterior circulation and cause both ipsilateral and contralateral anterior circulation strokes.
In addition to highlighting that a subclavian thrombus can take a circuitous path and cause an anterior circulation stroke contralateral to the diseased subclavian artery, this case illustrates the importance of a high index of suspicion for aTOS in patients with stroke and associated arm pain or discoloration. In our patient and others, the signs of subclavian artery stenosis and antegrade embolization, including loss of peripheral pulse, finger discoloration, and claudication, went unrecognized. Early recognition of the peripheral signs of aTOS may result in prompt initiation of antithrombotic therapy and surgical management, which could prevent ischemic stroke in high-risk patients.

**Conclusion**

We report a case of an anterior circulation stroke caused by a thrombus that likely originated in the contralateral subclavian artery in an adolescent patient with aTOS. This case illustrates the importance of a high index of suspicion for thoracic outlet syndrome in patients with stroke and associated arm pain or discoloration.

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