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Cerebral Embolic Stroke and Arm Ischemia in a Teenager With Arterial Thoracic Outlet Syndrome: A Case Report

Thomas S. Lee and George L. Hines, MD

A rare presentation of arterial thoracic outlet syndrome (TOS) is described in a young woman. Arterial TOS caused by a cervical rib produced acute upper extremity ischemia due to subclavian artery aneurysm formation. Clinical presentation also included left hemiparesis caused by right subclavian artery thrombosis and retrograde embolization of thrombus via the common carotid artery to the right middle cerebral artery distribution. Surgical repair of the subclavian artery was performed, but permanent neurologic deficit remained. Acute thrombosis of the right subclavian artery can produce cerebrovascular complication. The assessment of such risk in patients with arterial TOS is warranted and the arterial lesion corrected surgically.

Keywords: thoracic outlet syndrome; arterial thoracic outlet syndrome; subclavian artery

Thoracic outlet syndrome (TOS) is a condition caused by the compression of brachial plexus elements or subclavian vessels in the cervicoaxillary region. It typically involves 3 regions, which are the interscalene triangle, costoclavicular triangle, and subcoracoid space.[1] Unfortunately, the diagnostic criteria especially for the neurologic form of TOS, as well as the treatment, still remain to be topics of much debate. As a result, even the incidence of TOS is debatable, ranging from 3 to 80 cases per 1000 people.[2,3]

This case report describes an unusual cerebral vascular complication secondary to TOS in a patient with bilateral cervical ribs. In the process, we hope to highlight the importance of assessing the risk of cerebrovascular complication when right subclavian artery involvement is suspected.

From the Division of Vascular Surgery, Winthrop University Hospital, Mineola, NY (GLH), and the School of Medicine, State University of New York Stony Brook, Stony Brook, NY (TSL).

Address correspondence to: George L. Hines, MD, Winthrop University Hospital, 259 First Street, Mineola, NY 11501; e-mail: mepstein@winthrop.org.

Case Report

A 15-year-old female with past medical history of TOS diagnosed 3 months prior at an outside institution came to the hospital emergency room. The patient reported a sudden onset of left facial drooling and left-sided weakness associated with sudden blurry vision, nausea, vomiting, and severe headache located behind the right eye that began 5 hours prior to arrival. She stated that she woke up from a nap with the left-sided weakness. There was no history of trauma, seizure disorder, or loss of consciousness. The patient stated that 1 week earlier her right arm was temporarily locked with a bent elbow. She also described a 2- to 3-month history of intermittent right index finger tingling, numbness, and pinprick sensation. Previous MRI and venogram revealed the diagnosis of TOS. She has been under the care of orthopedists and was treated conservatively with osteopathic manipulations, including increasing range of motion in shoulder, cervical neck massage, and spinal relaxations.

The examination on admission revealed an alert and oriented patient with significant left facial droop with mildly slurred speech. Extraocular movements were intact with equal and reactive pupils

bilaterally. The examination of extremities revealed pale and cool right upper extremity with nonpalpable radial and brachial pulses, but bounding left upper extremity radial and brachial pulses. Left upper and lower extremities displayed 3/5 strength, whereas right upper and lower extremities exhibited 5/5 strength. Deep tendon reflexes were significantly decreased on the left side with positive Babinski's sign. Lastly, sharp and dull sensory tests were intact bilaterally.

The admission chemistry, complete blood count, and head CT were all normal. The patient was admitted to the pediatric ICU with the working diagnosis of left hemiplegia. Within the next 12 hours, left-sided weakness worsened progressively from 3/5 to 0/5 in strength but with unaffected sensation. Subsequent MRI and MRA revealed the occlusion of the right middle cerebral artery (MCA) and right MCA infarct. Digital subtraction angiography showed the occlusion of right subclavian artery at the level of the mid-clavicle (Figure 1) and at the distal end of the right internal carotid artery. Although the right common carotid artery and common carotid bifurcation were normal, there was absence of flow to the right anterior or middle cerebral arteries due to an abnormal right internal carotid artery (Figure 2). Right posterior cerebral artery flow was preserved. Upper arterial duplex scan showed total occlusion of the right subclavian and brachial arteries, whereas cerebrovascular duplex scan demonstrated abnormal Doppler waveform in the right common carotid artery without obvious signs of stenosis or occlusion. Cervical x-ray revealed bilateral cervical ribs. Negative echocardiogram ruled out cardiac source of embolus. Lastly, hypercoagulation tests revealed factor V Leiden heterozygous status and a positive antinuclear antibody of 80. However, the tests for anti-double strand DNA antibody, anti-smooth muscle antibody, anticardiolipin, C3, C4, anti-thrombin III, protein C, protein S, and homocystinemia were all negative.

Upon diagnosis of arterial thrombus, heparin was administered with partial thromboplastin time in the range of 55 to 70. Requested by neurology, a combination of carbidopa and levodopa (Sinemet, Merck & Co, Inc, Whitehouse Station, NJ) was also started. On hospital day 8, the patient underwent a surgical intervention, including right subclavian, axillary, and brachial thrombectomy and resection of right cervical rib, using the supraclavicular approach. A segment of the right subclavian artery was resected

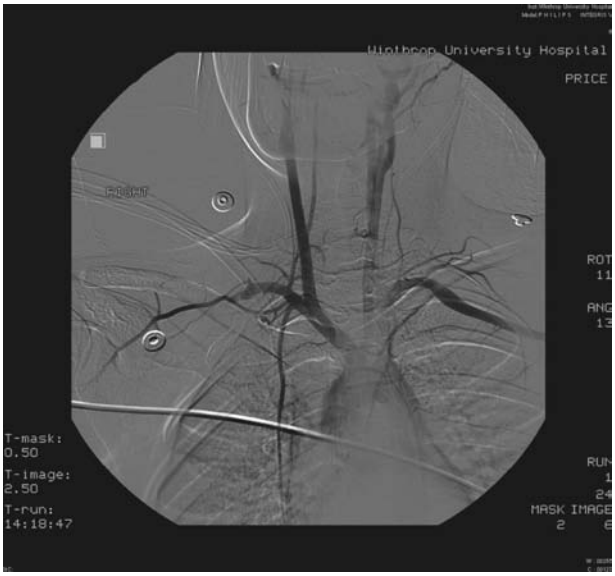

Figure 1. Digital subtraction angiography depicts the obstruction of flow in the right subclavian artery, whereas right common carotid artery flow is maintained. The left common carotid and subclavian arteries are shown to be patent.

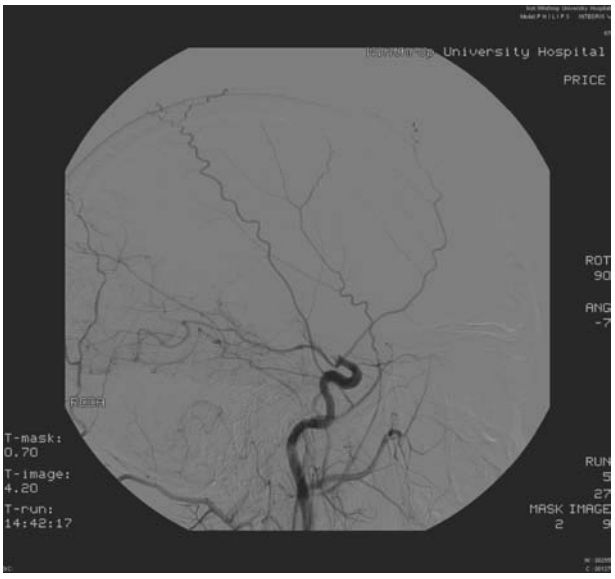

Figure 2. Sagittal view of the patient's head reveals occlusion of distal right internal carotid artery and the absences flow to right middle cerebral artery and right anterior cerebral artery.

owing to significant intimal damage, aneurysm formation, and arterial sclerosis. Right subclavian to right axillary artery bypass was performed using right great saphenous vein graft. The procedure was well-tolerated by the patient. Postoperatively, the patient's right upper extremity became warm and thin radial pulses were present. Since then, no cyanotic events of her right upper extremity have been reported. The

patient was discharged with an intense physical therapy regimen. At 3 months follow-up, the patient was able to ambulate without assistance but had residual left upper extremity weakness.

Discussion

TOS can involve the brachial plexus, subclavian vein, or subclavian artery. Reportedly, most TOS cases involve the brachial plexus (94%-96%) and result in neurological symptoms as initial complaints.[4,5] Pain, paresthesia, or weakness in ulnar distribution, involving C8 and T1, are the most frequent initial neurologic symptoms.[1] Rarer forms of TOS involve the subclavian vein (4%-6%) or subclavian artery (1%).[4,5] The signs of subclavian vein involvement include thrombosis, edema, and cyanosis of the involved upper extremity as well as the distension of superficial veins of the shoulder and chest.[6] Contrastingly, subclavian artery compression may present with pallor, pulselessness, coolness of the involved upper extremity, as well as antegrade or retrograde propagation of a thrombus or embolus.[6]

The subclavian vessel's involvement in TOS is often due to neck trauma or anatomic abnormalities such as cervical ribs, abnormal first rib, enlarged transverse process of seventh vertebra, or congenital fibromuscular bands.[7] In our patient's case, the most likely cause of TOS in the absence of neck trauma history was the presence of cervical ribs. Cervical ribs induce pathologic conditions by narrowing the passageway through which the brachial plexus and the subclavian artery pass in the interscalene triangle, which is defined by the anterior scalene muscle anteriorly, middle scalene muscle posteriorly, and medial surface of the first rib inferiorly.[1,8] The reported incidence of cervical ribs is about 1% in the general population.[8,9] Cervical ribs are found twice as frequently in females (68%) than in males (32%) and are bilateral in greater than 50% of involved cases.[10] Whereas cervical ribs are asymptomatic in 90% of cases, symptomatic cervical ribs typically result in neurogenic TOS and, less commonly, arterial TOS.[8] The pattern of involvement differs between incomplete and complete cervical ribs. Complete cervical ribs are associated with neurogenic or arterial TOS. However, only neurogenic TOS is typically observed with incomplete cervical ribs.[8,11] Furthermore, the onset of TOS in the absence of neck trauma was higher with complete cervical ribs (50%) than with incomplete cervical ribs (15%).[8] Better results from surgical intervention were observed with complete cervical ribs than with incomplete cervical ribs.[8]

Although subclavian artery involvement is the rarest form of TOS, such involvement can carry significant prognostic risks. Three major pathologic changes in arterial TOS include partial or complete thrombosis of the subclavian artery, poststenotic dilatation of the subclavian artery, and poststenotic subclavian artery aneurysm.[7] All 3 entities can result in distal embolization of the involved upper extremity as well as acute hand ischemia, claudication, vasomotor phenomena, and digital gangrene. In addition, as in our patient, the more uncommon complication of retrograde thrombus propagation from subclavian artery to vertebral or carotid arteries can result in cerebrovascular embolism.

The diagnosis of vascular TOS should begin with careful history and physical examination. Arterial TOS occurs typically in young adults with vigorous arm activity. Patients with arterial TOS may present initially with mild symptoms due to extensive arterial collateralization of upper extremities. As a result, patients may not seek medical attention until ischemic events have occurred, such as ulceration of digits, gangrene, absent pulses, or Raynaud's phenomenon.[12] Physical examination may reveal signs of arterial compromise such as pallor, pulselessness, and coolness of the involved limb. One should auscultate for bruits. In addition, blood pressure measurements of both arms should be evaluated. If the blood pressure in the affected limb is markedly reduced and such difference in the blood pressures is greater than 20 mm Hg from the contralateral limb, arterial involvement is likely.[1] However, provocative maneuver tests, such as Adson's and Halsted, are unreliable.[10]

Further imaging studies can be useful in the establishment of diagnosis and understanding the extent of pathology. Chest radiographs and cervical spine x-rays are useful tools in detecting abnormal bony structures, such as cervical rib and anomalous first rib. However, soft tissue abnormality causing TOS may require MRI or MRA. Color flow duplex and plethysmography are helpful noninvasive studies. Arteriography, venography, MRI, MRA/magnetic resonance venography (MRV), or cytotoxic assay can be used to confirm the diagnosis. Minimally, 4-vessel view arch aortogram should be conducted if a patient is suspicious for vascular TOS.

Surgical intervention is indicated in vascular TOS as it is typically resistant to conservative management.[1] The surgical treatment of arterial TOS includes the excision of the cervical and/or first rib, followed by reconstructive vascular procedure, such as resection and anastomosis or replacement with vein or prosthetic grafts.[7] Two common approaches to TOS are the supraclavicular and transaxillary approaches. The advantage of the supraclavicular approach is that it allows good exposure of the brachial plexus, medial two thirds of the first rib, and the cervical rib, if present.[1,12] As a result, first rib resection or anterior and middle scalenectomies can be done with this approach. The transaxillary approach provides access to the first rib through a hidden incision. However, there is limited exposure of neural structures and the cervical rib with the transaxillary approach.[1] The success rates of the supraclavicular and transaxillary approaches defined as complete to partial resolution of symptoms are 87.5% to 89% and 81% to 93%, respectively.[8,9,13,14] Nonetheless, Sanders reported comparable success rates for anterior and middle scalenectomy, transaxillary first rib resection, and combined supraclavicular scalenectomy and first rib resection.[15] Potential complications from TOS decompression surgeries include major neurovascular injuries, Horner's syndrome, phrenic nerve, long thoracic nerve, supraclavicular nerve, and intercostal brachial nerve injuries.[10,12]

There are limited reported cases of cerebral complication as a result of retrograde embolic event from subclavian artery involvement in TOS.[16-18] However, all those reports involved the right subclavian artery with the propagation of thrombus to the right brachiocephalic trunk and had a latent period of 3 months to 3 years from the onset of upper limb symptoms and major cerebral ischemic event.[16] Although an embolic event leading to cerebrovascular accident is a rare occurrence, vigorous diagnostic workup appears to be warranted to prevent such a devastating complication. Furthermore, when such risk is found, surgical treatment should be carefully considered, instead of conservative treatment.

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