PEARL Thoracic outlet syndrome causing distal subclavian artery disease is a rare cause of artery-to-artery embolic stroke.

Brainstem ischemic stroke is a result of compromise to the posterior circulation. This is often due to antegrade embolism from the heart or proximal vessels. Retrograde blood flow has been described in the subclavian artery, thus making the distal subclavian artery a source of possible retrograde embolism.

CASE SUMMARY A 21-year-old man was admitted with a history of acute dysarthria, dysphagia, and left hemiparesis for 1 day. He also had right arm pain suggestive of claudication for 1 month. He had transiently lost consciousness a month before, but did not have any permanent neurologic deficit subsequently, and he had not sought medical advice. Neurologic examination revealed a Glasgow Coma Score of 15, pinpoint pupils, conjugate deviation of the eyes to the left, spastic left hemiparesis, and palatal palsy. He had a pulseless and cold right arm. The rest of the examination was unremarkable.

Investigations revealed bilateral pontine infarctions (right more than left) on brain MRI, and a right cervical rib on cervical spine X-ray. An arch aortogram detected a stenosis of the right subclavian artery approximately 1 cm distal to the origin of the right vertebral artery (figure). There was poststenotic dilation of the artery with the distal axillary artery occluded and a number of collaterals present. Right vertebral angiography found total occlusion of the basilar artery with an embolus in situ. Retrograde blood flow was demonstrated in systole and diastole in both subclavian arteries (right greater than left) using Doppler ultrasound. Transthoracic echocardiography, thrombophilia screen, and C-reactive protein were normal. A screen for arrhythmias was negative.

DISCUSSION The abrupt onset of gaze paresis associated with hemiparesis and pinpoint pupils in this patient suggested ischemic stroke involving the pons, which was confirmed by MRI. The commonest cause of ischemic stroke in a young patient is embolism, originating from the heart. However, clinical evaluation, normal electrocardiographic assessment, and a negative transthoracic echocardiogram made the heart an unlikely source of embolism in this patient.

Less commonly, artery-to-artery embolism, especially from the proximal circulation and often secondary to dissection, is an important cause of stroke in the young. Free flow of contrast in the proximal vessels, including the cervical vertebral artery and innominate artery, made embolism from the proximal vessels unlikely.

Arch aortogram and cervical X-ray suggested a right cervical rib causing thoracic outlet syndrome (TOS) and distal subclavian artery disease. There are 2 possible mechanisms of cerebral embolism in this case: retrograde propagation of a thrombus and retrograde cerebral embolism.

The subclavian artery may be subject to trauma by a cervical rib, resulting in progressive stenosis and occlusion. Typically, there is poststenotic dilation (as seen in this patient) and aneurysm formation. A mural thrombus may form at the site of the compression or in the aneurysmal segment. A thrombus thus formed has an opportunity of extending proximally and embolizing via the right vertebral artery to occlude the basilar artery. Retrograde propagation of a thrombus has been described.

Subclavian artery flow patterns measured using Doppler ultrasound indicated both antegrade and retrograde flow occurring across both subclavian lumina in cardiac systole with small amounts of retrograde flow in diastole. Retrograde flow in diastole was greater in the occluded subclavian artery compared to the left. It was noted that retrograde flow increased on extreme rotation of the neck. Retrograde flow could carry an embolus toward the origin of the vertebral artery, and the next cardiac contraction would propel the embolus into the vertebral circulation. Previous case reports have demonstrated
bidirectional flow in normal as well as occluded subclavian arteries. These too have demonstrated greater retrograde flow in the occluded vessels suggesting greater reverse flow due to increased peripheral resistance.

TOS refers to compression of the neurovascular structures in the area just above the first rib and behind the clavicle. The subclavian artery is affected in 1% of cases of TOS. Cervical ribs and the first anomalous rib are rare conditions, present in approximately 1% of the population and in 4.5% of patients with TOS. The type of cervical rib is of significance in arterial complications. It has been established that short and incomplete ribs preferentially produce neurologic complications by nerve compression, while long or complete ribs as seen in this patient have arterial complications. Thromboembolism to the forearm and digits is the most typical clinical presentation of patients with distal subclavian artery disease due to TOS. Due to collateral formation, ischemic symptoms may be mild. This may explain the late presentation of our patient to the hospital. The presence of collaterals also suggests longstanding thrombosis of the distal subclavian artery.

Previous case reports have described distal subclavian artery disease secondary to TOS causing cerebral embolism. The majority of these cases described right TOS leading to cerebral embolism, involving commonly the right middle cerebral artery. This phenomenon is possibly due to the anatomic characteristics of the right carotid artery branching from the brachiocephalic artery and the differences in caliber of the common carotid and vertebral arteries with reduced resistance seen in the larger common carotid artery.

Surgical treatment may be considered in a patient with occlusive distal subclavian artery disease secondary to a cervical rib in order to prevent recurrent embolism. Surgery in this patient was deferred due to poor prognosis.

REFERENCES

Pearls & Oy-sters: Distal subclavian artery: A source of cerebral embolism
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