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An Interesting Case of Young Stroke

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Abstract: Cervical rib itself occurs in < 1% of the population. Of these only a small percent are symptomatic and when symptomatic it is mostly due to involvement of brachial very rarely due to subclavian artery compression. We report a case of stroke in a young female due to right sided cervical rib causing stenosis and post stenotic dilation of subclavian artery. Magnetic resonance angiogram showed evidence of thromboembolic episode in the right middle cerebral artery. Though what is expected is antegrade embolisation from the subclavian artery, retrograde embolisation into the carotid arteries occurs rarely leading to stroke.

Key words: Young Stroke · Cervical Rib · Subclavian Artery Stenosis · Retrograde Thromboembolisation

INTRODUCTION

Case History: A twenty seven year old female presented to our medicine department with weakness of the left upper and lower limb associated with deviation of angle of mouth to the right for four hours, which developed suddenly after waking up from sleep. She had no other co morbidities. The patient was previously admitted a week back in a local hospital with complaints of pain in the right upper limb which aggravated on elevating the right arm. Chest X ray (Fig.1) showed a right cervical rib.

CT thoracic angiogram and Right upper limb angiogram (Fig.2) was done and diagnosed to have Right cervical rib causing significant stenosis and thrombotic occlusion of distal subclavian artery.

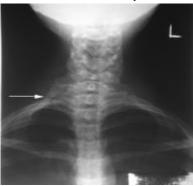


Fig 1: Showing a cervical rib on the right side

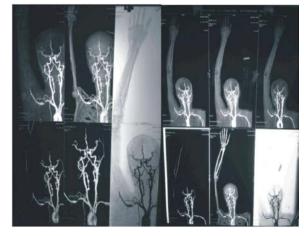


Fig 2: Showing significant stenosis of the right subclavian artery with post stenotic dilatation and decreased flow in the right brachial and distal vessels

On general examination the patient was conscious and oriented. Pulse rate was 82 beats/min, regular on the left radial artery. Right radial and brachial artery was not palpable. A soft systolic bruit was heard over the right subclavian artery. Blood Pressure was 110/70 mmhg on the left upper limb and was not measurable on the right upper limb. Examination of the central nervous system showed left hemiplegia and left UMN facial palsy.

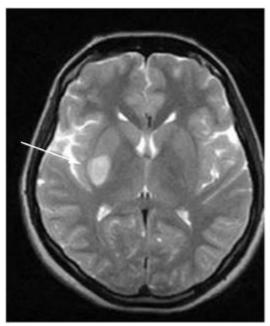


Fig 3: MRI brain showing acute non hemorrhagic stroke (bold arrow) in the right Gangliocapsular region



Fig 4: MRA Brain showing absent signal with abrupt cut off (bold arrow) seen beyond the proximal M1 segment of right MCA suggesting an Embolic Occlusion

Babinski's sign was extensor on the left side and was normal on the right. Examination of cardiovascular system, respiratory system and abdomen were unremarkable. Biochemical and hematological investigations were within normal limits.

CT Brain was normal. MRI Brain (Fig.3) with MR Angiogram (Fig.4) showed acute non hemorrhagic stroke in the right Gangliocapsular region, absent signal with abrupt cut off seen beyond the proximal M1 segment of right MCA suggesting an Embolic Occlusion.

Carotid and Vertebral artery duplex ultrasonography showed normal flow. 2-D echocardiography did not show any abnormality.

Patient was diagnosed as cerebrovascular accident right hemiplegia with right UMN facial palsy thromboembolic in origin, retrograde thrombosis from right subclavian artery thrombus. Patient was started on antiplatlet agents and heparin and referred to a vascular surgeon for further management.

DISCUSSION

Cervical rib is due to anomalous development of a rib, mostly from seventh cervical vertebrae. It can be associated with spinal anomalies elsewhere. Cervical ribs can be complete or incomplete. Only complete cervical ribs are known to cause vascular symptoms [1]. The incidence of cervical ribs in the general population is 0.76%. Seventy percent of cervical ribs are found in women. The majority of cervical rib cases are asymptomatic. When they are symptomatic, they present as Thoracic Outlet Syndrome (TOS)[2].

TOS can be classified into Neurogenic (Brachial Plexus 94-97%), Venous (Subclavian Vein -4-6%), Arterial (Subclavian Artery <1%). Neurogenic TOS is due to compression of brachial plexus trunks or cords (C5 to T1) and presents with symptoms of nerve irritation. Venous TOS occurs due to obstruction of subclavian vein secondary to a thrombus formed because of the intrinsic narrowing of the subclavian vein by compression and scar tissue.

Arterial TOS, the least common of the three produce symptoms secondary to subclavian artery stenosis or development of an aneurysm leading to thrombus formation with distal emboli. It presents usually with pallor, loss of peripheral pulse, decreased temperature in the hand, ischemia of the digits, claudication, paraesthesias and pain aggravated on abduction of arm. It is rare to have symptoms in the shoulder and neck. All these are due to antegrade embolisation.

Retrograde embolisation from a subclavian artery thrombus producing stroke is a rare phenomenon. It has been reported as early as 1884 by Gould *et al* [3]. But Symonds in 1927 was the first to explain that the thrombus in right subclavian artery extended proximally

to the junction of right common carotid artery and portion of this thrombus embolised to the cerebral circulation. Lazer et al. [4] found only one case of TIA among thirty cases with vascular TOS and Davis et al. [5] found five cases of cervical rib with stroke. Recently in 2008 Yamaguchi et al. [6] reported ten cases with cerebral embolism from a subclavian artery thrombus. In cases without a thrombus in common carotid extending from subclavian artery, cerebral embolism could be due to transient reversal of flow in the thrombosed subclavian artery. De Villiers [7] suggested the possibility of certain positions of arm that occluded the subclavian artery, thereby marked turbulence occurred in the proximal part of the vessel. And such positions are tolerated only during sleep. Few cases where patients woke up with cerebral dysfunction have been reported.

Patients with cervical rib with stroke more commonly have upper limb symptoms before having cerebral events. Right carotid artery stroke is more common than vertebrobasilar stroke probably because of the arterial anatomy [8]. But cerebellar infarction by an embolus via vertebral artery has been reported if the subclavian thrombus extended into the vertebral artery [9]. Right carotid artery stroke associated with right sided cervical rib while the vertebrobasilar stroke associated with left sided cervical rib [10].

Clinically Adson's test must reproduce the symptoms of the patient and there must be a decrease in the patient's radial pulse. Plain radiographs of the chest with upper thoracic and cervical spine studies, can easily detect congenital or acquired bony abnormalities. Duplex ultrasonograpy done at neutral and varying degrees of abduction show an increase in peak systolic velocity or obliteration of flow. Magnetic resonance (MR) imaging with angiography and computed tomography (CT) imaging with angiography of the thoracic inlet not only can find out the degree of stenosis, post stenotic dilatation and any thrombosis of the subclavian artery but also important details of the adjacent structures and can rule out other causes of thoracic outlet syndrome such as neoplasms and bony abnormalities. The most specific diagnostic examination for thoracic outlet syndrome is arteriography. It is indicated in a patient with ischemic upper extremity symptoms. The entire arterial circulation of the upper extremity from the aortic arch to the distal arteries of the fingers is visualised. It not only gives valuable information about subclavian artery, but delayed imaging after injection can show antegrade collateral circulation, which is helpful in planning surgery.

The syndrome of cervical rib with subclavian artery thrombosis must be treated immediately. The aim must be to prevent further embolism and restore function by surgical decompression of the thoracic outlet and resection of the embolic source. The cervical rib must be excised. Access to neurovascular structures can be further improved by resecting the clavicle or the first rib. Removing the latter may also prevent the development of neurological symptoms [11, 12]. Whatever the vascular pathology stenosis or aneurysm, the abnormal segment of subclavian artery should also be excised [13]. Primary end to end anastamosis is done if only a short segment of a vessel is involved. If a graft is required for arterial reconstruction, a large autogenous saphenous vein or expanded polytetrafluoroethylene or Dacron fabric grafts may be used [14-16].

Our patient had obvious upper limb symptoms before the cerebral event and the cerebral event occurred in the morning. She had a subclavian artery compression by a cervical rib with post stenotic dilatation and a thrombus in the subclavian artery. She had a normal carotid artery Doppler suggesting emboli from the subclavian thrombus to have retrogradely migrated to the cerebral circulation producing the cerebral event.

CONCLUSION

Thrombo-embolic Stroke has occurred in patients, only in the presence of obvious thrombus formation in the stenosed subclavian artery by a cervical rib. It may or may not be associated with a thrombus extending into the common carotid. In patients with cervical rib with vascular TOS, both retrograde and antegrade thromboemolism can occur. Immediate intervention and corrective surgery must be done when the subclavian artery is involved due to a cervical rib.

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