Thromboembolic stroke associated with arterial thoracic outlet syndrome

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Abstract/Summary

Background and Purpose: Thoracic outlet syndrome occurs due to compression of the neurovascular structures as they exit the thorax. Subclavian arterial compression is usually due to a cervical rib, and is rarely associated with thromboembolic stroke. The mechanism of cerebral embolisation associated with the thoracic outlet syndrome is poorly understood, but may be due to retrograde propagation of thrombus or transient retrograde flow within the subclavian artery exacerbated by arm abduction. We report an illustrative patient and review the clinical features, imaging findings and management of stroke associated with thoracic outlet syndrome.

Introduction

Thoracic outlet syndrome (TOS) occurs due to compression of the neurovascular structures as they exit the thorax between the scalene muscles, the clavicle and the first rib, and can present with neurological, arterial or venous occlusive symptoms [1,2]. Subclavian arterial compression is commonly attributed to a cervical rib. This abnormality is present in around 1% of the population, is bilateral in 50% of patients, and is twice as common in women [1]. Subclavian arterial compression leads to claudication, weak pulse and reduced blood pressure in the affected arm, all of which become more pronounced with arm abduction [1,2]. A subclavian bruit or a pulsatile supraclavicular mass may be present if there is a stenosis and associated post-stenotic aneurysm. If thrombus forms within the aneurysm, this may embolise to the axillary or brachial arteries leading to acute arterial occlusion. Thromboembolic stroke associated with TOS is a rare event first described by Symonds in 1927 [3], although a patient with obliterative arteritis described by Gould in 1884 and updated in 1887 may also represent the association [4,5]. We report a case of right middle cerebral artery (MCA) territory ischaemic stroke in a teenager with a concurrent diagnosis of cervical rib and subclavian arterial compression.

Case

A 16-year-old girl presented with left hemiparesis. Past history included several months of intermittent digital cyanosis, pain and numbness of the right hand only. She was otherwise healthy with no regular medications, had modest alcohol intake, smoked tobacco occasionally, and denied illicit drug use.

Ten days before admission, the patient had an episode of left-sided weakness, which lasted several hours but completely resolved. On the day of presentation the patient awoke with left-sided weakness and mild right-sided headache. Right brachial blood pressure was unrecordable and brachial, radial and ulnar pulses were absent. Her preferred sleeping posture was right lateral decubitus with extreme right shoulder abduction for head support.

MRI of the brain revealed acute right MCA territory infarction (Fig. 1). A plain radiograph demonstrated bilateral cervical ribs (Fig. 2). Digital subtraction angiography of the head, thoracic outlet, and right upper extremity revealed occlusion of the M1 segment of the right MCA, segmental occlusion of the right proximal brachial artery, and functional occlusion of the right distal subclavian artery (SCA) during shoulder abduction with post-stenotic dilatation (Fig. 3). No thrombus was seen in the dilated portion of the SCA.

Blood cell counts with film and inflammatory markers were normal. Thrombophilia and vasculitis tests were negative. Electrocardiogram showed sinus rhythm with no arrhythmia during 48 hours of telemetry. A trans-thoracic echocardiogram with agitated saline injection and delayed phase imaging did not reveal an interatrial shunt, and trans-oesophageal echocardiogram was normal.

The patient was commenced on warfarin and underwent rehabilitation. She subsequently underwent surgical resection of the right cervical rib via a supraclavicular approach. There was compression of the SCA between a complete cervical rib (which articulated with the first rib) and scalenus anterior (Fig. 4). Anterior scalenotomy was performed, and the first and cervical ribs were resected; SCA reconstruction was not required. The patient recovered without complication.

Discussion

The mechanism by which TOS-associated cerebral embolisation occurs is poorly understood. Subclavian arterial compression leading to stasis, intimal trauma and thrombus formation is likely the initial event. Retrograde propagation of thrombus to the origin of the vertebral or common carotid arteries may occur next [3]. In some patients with TOS and associated stroke, thrombus extending into the innominate artery has been found on vascular imaging [6,7], and during surgery [8]. An alternative explanation is transient retrograde flow within the SCA. This has been identified using ultrasonography in some patients with TOS associated with stroke [8,9], and experimental studies have demonstrated that retrograde SCA flow can be readily induced [10,11]. In this patient it is proposed that prolonged occlusion of the SCA during sleep with extreme shoulder abduction could have led to stasis and thrombus formation at the SCA origin. The episode of weakness a week prior to presentation is suggestive of a separate episode of embolisation from this source; extensive investigation did not reveal a cardiac abnormality to account for these events.

Including the patient reported here, 33 patients with stroke or transient ischaemic attack associated with SCA disease have been reported in detail, as well as an additional three patients within case series without detailed clinical information [12–14]. Twenty-six of these reported patients were associated with cervical rib [3–9,15–29]. Other reported causes of SCA disease associated with stroke include left first rib anomaly [30], a non-united right clavicular fracture [31], repetitive sporting injury [32], atheroma [8,15], dissection [33], and presumed congenital saccular aneurysm of the axillary artery [34].

Stroke associated with cervical rib is an entity seen in young people; among the 26 reported patients with stroke associated with cervical rib, the median age was 21 years (range, 14–49 years). Thirteen (50%) were male. Twenty-one (81%) patients had preceding symptoms suggestive of TOS for between 3 weeks to 12 years prior to stroke, however TOS had only

been diagnosed in a minority of patients. Symptoms of TOS ranged from claudication on vigorous activity and intermittent cyanosis to severe constant pain and ischaemia with gangrene. In some cases of stroke associated with cervical rib the onset of stroke symptoms was reported to occur at a time of arm abduction such as during sleep [23], whilst fast-bowling [22], or whilst hammering [6]. All patients with cervical rib had absent peripheral pulses in the affected limb; this examination finding in a young person with stroke should prompt investigation for TOS.

Of the 20 patients with cervical rib that had vascular imaging, all had SCA stenosis with post-stenotic dilatation. SCA occlusion was seen in six patients; in two thrombus was found to extend as far back as the innominate artery. Occlusion of the axillary or brachial arteries alone was seen in eight patients, reflecting clot disruption with distal embolisation. Twenty (76%) patients had anterior circulation emboli, four patients had posterior circulation infarcts, and two patients had anterior and posterior circulation territory infarcts; the reason for the predominance of anterior circulation infarcts is unclear. Right-sided SCA disease was present in all patients except one, reflecting that the innominate artery gives rise to the subclavian and common carotid arteries on the right side. One case of posterior circulation infarction was associated with left SCA occlusion [8].

Seventeen (65%) patients underwent thoracic outlet decompression via rib resection, usually via a supraclavicular approach. Eleven (42%) patients had thrombectomy or resection of the diseased portion of SCA; the decision to proceed with this intervention depended on the degree of arterial dilatation and the amount of associated mural thrombus. Eight (31%) were treated with anticoagulation peri-operatively. Long-term outcomes following surgical intervention and the role of anticoagulation are unclear, although one study of 55 operations for arterial TOS suggested that surgical decompression with or without arterial resection is safe and effective at relieving symptoms from arm ischaemia [2].

Stroke due to SCA compression associated with TOS is rare, and to our knowledge has not previously been systematically studied. In a minority of reported patients imaging or operative findings have suggested either retrograde propagation of thrombus or retrograde flow within the SCA, which may be exacerbated by abduction of the arm and complete occlusion of the SCA. It is mainly seen in young people. There is no evidence base to guide the management of this condition, however surgical intervention to decompress the SCA with or without resection of the diseased section of SCA is thought to be the most effective strategy to minimise the risk of stroke recurrence.

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Table 1. Cases of thromboembolic stroke with associated subclavian arterial disease.

Reference	Age	Sex	Aetiology of TOS	Location of infarct	Treatment
Gould AP, 1884	19	M	Cervical rib	Presented with left hemiparesis, no brain imaging reported	-
Symonds CP, 1927	29	F	Right cervical rib	Presented with left hemiparesis, no brain imaging reported	-
Symonds CP, 1927	20	F	Right cervical rib	Presented with left hemiparesis & aphasia, no brain imaging reported	-
Yates AG & Guest D, 1928	41	F	Non-united fracture of right clavicle	Autopsy: basilar artery thrombus	Died from stroke
Smith GW, 1941	22	M	Autopsy: saccular aneurysm of axillary artery	Autopsy: occlusion of right internal carotid artery	-
Hoobler SW, 1942	38	M	Bilateral cervical ribs	Presented with left hemiparesis, no brain imaging reported	-
Samiy E, 1955	23	F	Cervical rib	Presented with left hemiparesis, no brain imaging reported	Cervical exploration
Rabaté M, 1959	28	F	Cervical rib	Presented with left hemiparesis, no brain imaging reported	Excision of cervical rib, transection of scalenus anterior muscle
Shucksmith HS et al, 1963	17	F	Bilateral cervical ribs	Presented with left hemiparesis, no brain imaging reported	Cervical rib and 1 st rib partially excised
Davis JM and Golinger D, 1966	19	M	Right cervical rib	Presented with left hemiparesis, no brain imaging reported	Cervical rib partially excised, SCA ^a aneurysm excised & closed with saphenous vein patch, axillary and brachial artery thrombectomy Complicated by brachial plexopathy
De Villiers JC, 1966	15	F	Bilateral cervical ribs	Presented with left hemiparesis, right MCA ^b occlusion on arteriogram	Cervical rib partially excised, SCA ^a aneurysm excised, cervical sympathectomy
Eriksson I & Hierton T,	43	M	Right cervical rib	Presented with left sided weakness, no brain	Division of scalene muscle, right SCA ^a explored,

1968				imaging reported	thrombectomy
Blank RH & Connor RG, 1974	18	F	Bilateral cervical ribs	Right medulla	Surgical intervention refused by patient
Prior AL et al, 1979	35	M	Right cervical rib	RightMCA ^a territory	Excision of cervical rib, SCA ^a found to be thrombosed to its origin, anticoagulated
Prior AL et al, 1979	21	F	Bilateral cervical ribs	Presented with transient left hemiparesis & aphasia(5 episodes), no brain imaging reported	Scalenus anterior and fibrous band compressing SCA ^a divided, cervical rib excised
Prior AL et al, 1979	38	M	Bilateral cervical ribs	Presented with vertigo, ataxia & diplopia, no brain imaging reported	Left cervical rib and fibromuscular band excised, anticoagulated
Prior AL et al, 1979	50	M	Atheroma	Autopsy: Extensive right hemispheric infarct, saddle embolus at right common carotid bifurcation	No intervention, died from stroke
Fields WS et al, 1985	30	M	Repetitive injury in a baseball pitcher	Right basal ganglia, occipital lobe, cerebellum	Clot removal from innominate, right subclavian and proximal right common carotid arteries Subsequent subclavian-brachial bypass, using external iliac autograft, right first rib resection, scalenectomy, upper thoracic sympathectomy
Al-Hassan HK et al, 1988	28	M	Right cervical rib	RightMCA ^b territory	Medial right clavicle and cervical rib excised, resection of stenosed and dilated portions of SCA ^a , reconstruction with synthetic graft
Al-Hassan HK et al, 1988	36	M	Right cervical rib	RightMCA ^b territory	Partial excision of clavicle and cervical rib, resection of SCA ^a , reconstruction with autologous vein graft
Bearn P et al, 1992	41	M	Right cervical rib	RightMCA ^b territory	Cervical rib excised, SCA ^a aneurysm resected and synthetic graft placed
Nishibe T et al, 2000	48	F	R cervical rib	R MCA territory	Cervical rib excised, anterior and middle scalenectomy
Matsen SL et al, 2003	19	F	Right cervical rib	Right lateral medulla	Scalenectomy, cervical rib & SCA ^a aneurysm excised & synthetic graft placed, anticoagulation
Chen W-H et	90	F	Possible	RightMCA ^b territory	Thrombectomy

al, 2005			axillary arterial dissection		
Naz I & Sophie Z, 2006	18	M	Right cervical rib	Right basal ganglia	Excision of cervical rib, saphenous vein conduit between rightsubclavian& brachial arteries
Naz I & Sophie Z, 2006	52	F	Atheroma	Bilateral cerebellum	Resection of SCA ^a stenosis (found to be atheroma) & synthetic graft placed, anticoagulated
Lee TS & Hines GL, 2007	15	F	Bilateral cervical ribs	RightMCA ^b territory	Right cervical rib excised, rightsubclavian, axillary &brachial thrombectomy, resection of SCA ^a & saphenous vein graft placed, anticoagulated
Yamaguchi R et al, 2008	22	M	Left first rib anomaly	Left cerebellum	First rib partially excised, anticoagulated
Gooneratne IK et al, 2009	21	M	Right cervical rib	Bilateral pons	Surgery not done due to ‘poor prognosis’
Sharma S et al, 2010	18	M	Right cervical rib	Right basal ganglia	Cervical rib excised
Kataria R et al, 2012	14	F	Right cervical rib	Right brainstem & cerebellum	Cervical rib excised, anticoagulated
Jusufovic M et al, 2012	49	M	Bilateral cervical ribs	Right cerebellum & right superior parietofrontal lobe	Cervical rib excised, anticoagulated

^aSubclavian artery

^bMiddle cerebral artery

Figure 1. DWI (1a) and ADC (1b) MRI images demonstrating acute right MCA infarction (arrows).

Figure 2. Plain radiograph demonstrating bilateral cervical ribs (arrows).

Figure 3. Digital subtraction angiography demonstrating right M1 segment of MCA occlusion (arrow).

Figure 4. Angiography demonstrating occlusion of the axillary and brachial arteries (arrows).

Figure 5. Angiography demonstrating fusiform dilatation of the distal SCA (dashed arrow, 5a) and subclavian arterial occlusion on abduction of the arm (solid arrow, 5b).

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