Thoracic outlet syndrome

Tim Beckitt

Abstract

Thoracic outlet syndrome (TOS) consists of a group of three syndromes resulting from compression of the subclavian artery (arterial – ATOS), brachial plexus (neurogenic – NTOS) or subclavian vein (venous – VTOS) as they pass through the thoracic outlet. The thoracic outlet is not one, but three distinct anatomical spaces and compression at differing sites results in differing combinations of symptoms. NTOS is the most common, usually caused by compression of the brachial plexus in the scalene triangle or pectoralis minor space. In making the diagnosis of NTOS, neurogenic compression in the cervical spine, carpal and cubital tunnels should be excluded. Management of NTOS is usually conservative, with physiotherapy and postural exercise, but pain or muscle wasting may be indications for surgery. VTOS is caused by compression of the subclavian vein at the costoclavicular junction. It may present as acute venous thrombosis (Paget–Schroetter syndrome) or positional swelling of the upper limb without thrombosis (McCleery's syndrome). In acute thrombosis, clot lysis, rib excision and venoplasty may be indicated. ATOS occurs due to compression of the subclavian artery, often in association with an anomalous structure, such as a cervical rib. Post-stenotic aneurysmal dilatation of the artery can result in thrombosis and distal embolization. Acute upper limb ischaemia necessitates urgent rib excision and arterial reconstruction.

Keywords Cervical rib; McCleery's syndrome; neurogenic compression; Paget–Schroetter syndrome; scalenectomy; thoracic outlet syndrome

Introduction

The thoracic outlet is defined as the space in the lower neck between the thorax and axilla through which the subclavian vein, subclavian artery and brachial plexus travel.

It consists in fact of three distinct spaces through which these structures pass (Figure 1):

- Scalene triangle
- Costoclavicular space
- Pectoralis minor space

The thoracic outlet syndrome (TOS) is divided into three main types depending on the anatomical structure affected by the compression or impingement:

- Subclavian artery – arterial thoracic outlet (ATOS)
- Brachial plexus – neurogenic thoracic outlet (NTOS)
- Subclavian vein – venous thoracic outlet (VTOS)

There is significant overlap between these syndromes of ATOS, NTOS and VTOS, which often coexist as a result of the site of anatomical compression.

Tim Beckitt *MD FRCS is a Consultant Vascular Surgeon in Bristol, UK.*
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Epidemiology

TOS usually presents in young patients, but the diagnosis can be difficult to make and, as a result, delays in diagnosis are not uncommon. More than 80% of symptomatic patients present with NTOS, with an estimated incidence of 3 cases per 100,000 per year.[1] A delay in diagnosis in this group can lead to permanent nerve damage. Venous thoracic is the second most frequent variant, accounting for 16%.

Pure arterial thoracic outlet syndrome is comparatively rare, but missed diagnosis can result in acute limb ischaemia with severe consequences for the patient. Whilst the *Reporting Standards of the Society for Vascular Surgery (SVS) for Thoracic Outlet Syndrome*[2] have standardized the diagnosis and investigation of TOS, it should be noted that dynamic compression of the subclavian artery is a frequent finding in asymptomatic patients with normal anatomy and as such the SVS stipulate that diagnosis of ATOS requires 'clinical symptoms due to documented symptomatic ischemia or objective arterial damage.'

Neurological TOS

NTOS may be associated with normal anatomy if the problem is due to traction of the lowest trunk of the brachial plexus as it passes over the neck of the first rib. More severe cases are seen in conjunction with anatomical abnormalities such as a cervical rib or band and increasing congenital abnormalities of the brachial plexus and scalene muscles are being recognized as contributing factors[3] alongside hypermobility.

Clinical features

Patients with NTOS often have a long history of symptoms with numerous previous negative investigations. Whilst most patients are young, it is important to note the symptoms of NTOS can be provoked or unmasked by trauma. Patients complain of positional pain and/or paraesthesia arising in the neck or supraclavicular fossa. Pain radiates over the deltoid into the medial forearm in an ulnar nerve distribution toward the little finger and medial half of the ring finger. A characteristic unilateral occipitofrontal headache with facial pain is common. In advanced cases, muscle wasting of the intrinsic muscles of the hand (Figure 2) with clawing of the little finger may be seen. Once motor units have been lost, they will never recover. More commonly patients complain of a lack of strength or unreliability of grip, and this may be accompanied by tremor. The proximity of the subclavian artery to the lowest trunk of the brachial plexus may result in mixed symptoms and patients often experience symptoms attributable to arterial compression such as coldness, discoloration and fatigue, which is often positional. The SVS[2] diagnostic criteria for NTOS recommend that diagnosis requires three of four of the criteria (Table 1). Provocation tests such as the Extended Arm Stretch Test (EAST) and the Upper Limb Tension Test (ULTT) may support the diagnosis but are of little value determining the site of impingement. Careful examination with palpation and percussion in the scalene triangle and over pectoralis minor is a crucial part of the examination. NTOS may coexist with other forms of peripheral nerve compression such as carpal tunnel syndrome or cervical radiculopathy, and this coexistence is known as double-crush syndrome. Disuse atrophy of the rotator cuff may lead to subluxation of the glenohumeral

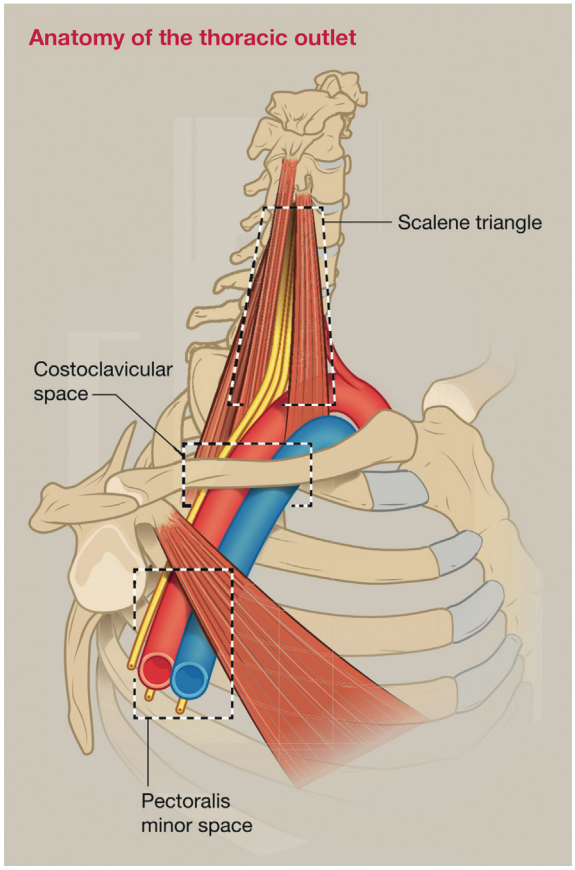

Figure 1

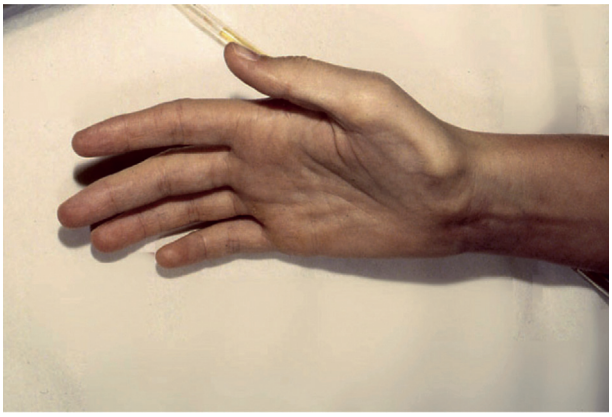

Figure 2 Hypothenar and intrinsic muscle wasting in NTOS.

Clinical features of neurologic thoracic outlet syndrome

- Positive local findings (irritation in the scalene triangle in NTOS or pectoralis minor space in NPMS).
- Positive peripheral findings (hand or arm symptoms consistent with central nerve compression).
- Absence of other potential diagnoses (cervical spondylosis or disc disease; carpal tunnel syndrome; cubital tunnel syndrome; brachial neuritis, etc.).
- Positive response to scalene or pectoralis minor local anaesthetic muscle blocks, with relief of symptoms due to muscle relaxation.

Table 1

joint with resulting shoulder pain and restriction of movement as a result of adhesive capsulitis (frozen shoulder).

Investigations

X-ray may reveal a cervical rib, but cervical bands are frequently not seen on plain radiography. Duplex scanning may demonstrate dynamic arterial compression (Figure 3), a good surrogate for lower root brachial plexus compression. Cervical spine MRI to exclude cervical radiculopathy and brachial plexus MRI may reveal deviation of the plexus or oedema. Neurophysiology studies are useful in excluding other neurological compression syndromes such as carpal tunnel which can be treated more easily but are often normal and cannot exclude the double crush phenomenon.

The electromyography (EMG) features of NTOS include a decrease in the sensory nerve potential in the medial antebrachial cutaneous nerve (C8—T1) and abnormalities of F-wave conduction.

Where a structural abnormality has not been identified by X-ray or MRI, ultrasound of the scalene triangle by an experienced practitioner may also detect aberrant scalene muscle anatomy and/or bands and subsequent diagnostic local anaesthetic block to the scalene muscles and/or to pectoralis minor is of value in identifying the site of impingement and may predict a favourable response to surgery. The local anaesthetic blockade of intramuscular nerve branches causes muscle relaxation, which may provide short-lived relief of symptoms and improved performance in provocation testing (EAST and ULTT).

Treatment

In early cases, NTOS can be caused by traction on the lowest trunk of the brachial plexus, which is exacerbated by poor posture or occupational factors. These cases may respond well to physiotherapy targeting posture correction and strengthening. In patients without an identified structural abnormality there may be a role for ultrasound guided scalene block with botulinum toxin (BTX) in providing medium term relief of symptoms and facilitating effective physiotherapy. The one randomized trial[4] recruited only 38 patients, with improvements seen in pain scores at 6 weeks not achieving statistical significance. Where there is evidence of muscle wasting along with pain and paraesthesia, intervention is justified to prevent progressive functional deterioration. As previously discussed, lost motor units do not recover and in patients with severe muscle wasting and a painless hand there may be no benefit in intervention.

Surgical decompression in NTOS is described by either the transaxillary or supraclavicular approaches. The supraclavicular approach gives a better view of the entire brachial plexus and allows both anterior and middle scalenectomy with a good view of the cervical rib or band, which may be the precipitating factor. After mobilizing the scalene fat pad, anterior scalenectomy is performed, taking care to identify and preserve the phrenic nerve. The subclavian artery is slooped, allowing retraction to access the brachial plexus. The plexus is explored, taking care to protect the long thoracic nerve at the lateral edge of scalenus medius muscle. The cervical rib or extended transverse process of C7 is then divided and any cervical band excised. There remain advocates of rib sparing scalenectomy,[5] but as up to 30% of patients with NTOS suffer recurrent or ongoing symptoms, most surgeons advocate proceeding to first rib resection. It is important in NTOS

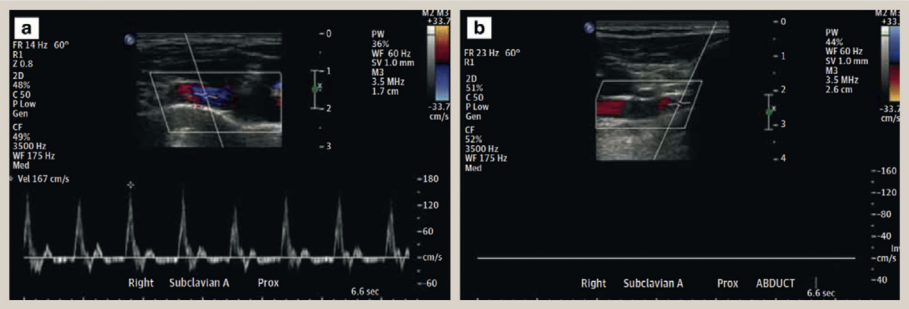

Figure 3 Duplex ultrasonography demonstrating cessation of arterial flow in stress positioning with arm abduction.

to ensure an adequate resection of the neck of the first rib posteriorly to adequately decompress the brachial plexus.

Pectoralis minor syndrome

Neurogenic pectoralis minor syndrome (NPMS) may present in isolation but more often occurs alongside other causes of NTOS. NPMS is an important cause of ongoing or recurrent symptoms after decompression. Symptoms result from compression of the neurovascular bundle as it passes below the pectoralis minor tendon and can be difficult to distinguish from compression at the scalene triangle, though may be suspected in patients with chest wall or axillary pain. The most useful clinical finding is tenderness over the pec minor tendon in the subcoracoid space. The diagnosis can be confirmed by local anaesthetic pectoralis minor muscle block. The role of BTX in this patient group is controversial. Surgical decompression can be achieved by pectoralis minor tenotomy. An incision is made in the deltopectoral groove, dissection along the lateral edge of pectoralis major leads to the tendon, which is divided below its insertion to the coracoid process.

Outcome

Where a definite abnormality is identified at operation, relief of symptoms can be expected in 80% of patients with NTOS. Residual or recurrent NTOS[2] is reported in up to 30%, and half of the recurrences occur within the first postoperative year. This may result from postoperative scar tissue growth, inadequate surgical decompression or a different pathology. NPMS should be considered if pectoralis minor tenotomy was not undertaken at the first procedure. Patients should be thoroughly counselled and, where surgery is not mandated by evidence of progressive muscle wasting, a period of conservative management should be undertaken. The consent process should not only discuss the risks of surgery, but also of the possibility of both the failure to improve symptoms and of recurrence.

Venous TOS

VTOS occurs more commonly in the dominant limb and has male preponderance. The presentation is often acute with a subclavian vein thrombosis and the association of thrombosis with strenuous and repetitive activity of the upper limb is known as Paget–Schroetter syndrome (PSS).

Pathophysiology

The subclavian vein passes into the thorax through the costoclavicular space, passing over the medial end of the first rib below the clavicle (Figure 4). Hypertrophy of the subclavius muscle may contribute to extrinsic compression of the subclavian vein. This phenomenon is observed in athletes undertaking upper body activity, such as weightlifters and swimmers, but may be seen in other patients undertaking repetitive arm movements. Repetitive shearing motion leads to fibrosis and scarring in the vein wall and may result in venous stenosis. Thrombosis of the subclavian and axillary vein may result. Rarely, venous compression resulting in thrombosis may occur in the pectoralis minor space.

Clinical features

Patients may present with intermittent exercise-related arm swelling associated with aching, purplish discoloration and prominent collateral veins over the shoulder, but without evidence of thrombosis (McCleery's syndrome). Acute venous thrombosis presents with an aching, swollen arm with blueish-purple discoloration, venous distension and collateral veins over the shoulder and chest wall (Paget–Schroetter syndrome, PSS). Patients with PSS are usually young and healthy and often have well-developed shoulder musculature. Provocation testing (EAST) often results in a bursting feeling in the affected limb. A history of paraesthesia may be indicative of coexisting NTOS. In patients presenting with upper limb venous thrombus in later life, other causes such as malignancy should be considered.

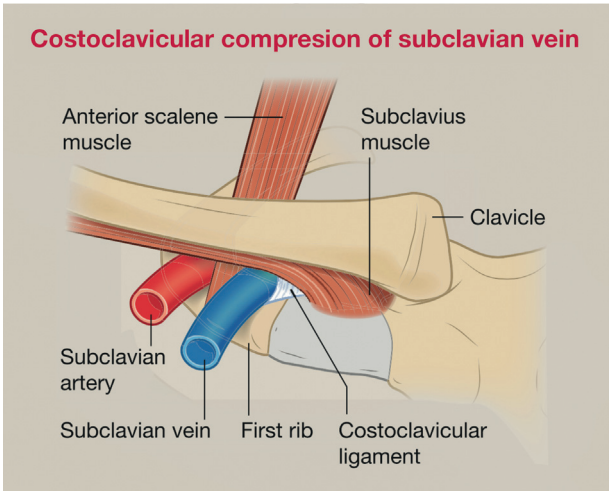

Figure 4

Natural history

Untreated, the majority of patients with PSS recanalize their subclavian vein with anticoagulation alone, but venous function does not return to normal and 28% will have significant residual symptoms of post-thrombotic syndrome (PTS), primarily with disabling upper limb swelling.[6]

Investigation

Plain X-ray is usually normal with no evidence of cervical rib. Duplex ultrasound scanning is the first line investigation. The vein behind the medial clavicle is often not seen directly but a lack of flow augmentation on inspiration and the presence of collateral veins are sufficient to diagnose the underlying thrombosis. MRI and CT venography are not mandatory but may establish the extent of thrombosis when planning intervention. Patients in whom treatment is being considered require formal contrast venography (Figure 5a) and this may be undertaken with a plan to proceed directly to catheter thrombolysis (Figure 5b). Thrombophilia screening in patients with upper limb venous thrombosis[7] may identify a thrombophilia in up to 22% and this is of value in identifying patients who may need lifelong anticoagulation.

Treatment

Medical management with oral anticoagulation is essential to prevent thrombus extension or pulmonary embolus. In older patients who are relatively asymptomatic and where the non-dominant limb is affected, anticoagulation alone will often suffice.

Figure 5 (**a**) Catheter venograms showing an occluded subclavian vein. (**b**) Catheter venogram after lysis, rib resection and venoplasty with restoration of flow.

Younger patients with a recent thrombosis who do not want to accept the 30% risk of PTS with a swollen limb should be offered the choice of intervention. The optimal management of PSS is a combined approach with percutaneous catheter-based thrombolysis, followed by early first rib resection and subsequent percutaneous balloon venoplasty. Stenting of the vein is avoided as it is associated with a high incidence of re-thrombosis if costoclavicular compression persists.

Thrombolysis in this young age group is safe with a <1% risk of bleeding. Where there is a large thrombus burden, mechanico-chemical lysis may be used first to reduce the thrombus burden before starting catheter lysis with an alteplase infusion for up to 48 hours. Check venography is performed and, once successful, therapeutic anticoagulation with either low molecular weight heparin or a direct oral anticoagulant (DOAC) is continued until surgery.

First rib resection is carried out on the next available theatre list. Resection of the first rib, subclavius tendon and costoclavicular ligament are undertaken. Surgical approaches include the transaxillary and infraclavicular approaches. Operative risks include neurovascular damage, recurrent venous stenosis and pneumothorax or haemothorax, which can necessitate pleural drainage.

The infraclavicular approach provides good access to the first rib, subclavius muscle and costochondral junction, allowing good visualization of the subclavian vein. Where patients have combined features of NTOS, this may be combined with a supraclavicular incision (a paraclavicular approach), which allows improved access to extend resection to the posterior part of the first rib. An infraclavicular approach can be used for surgical vein patch angioplasty of a fibrosed stenosis within the subclavian vein, but most authors favour an endovascular approach. Postoperatively, patients are discharged on a DOAC. Repeat venography is undertaken and if there is a residual stenosis, balloon venoplasty is performed. Advances in surgical techniques include the development of minimally invasive approaches with video-assisted thoracoscopic surgery (VATS) and robot-assisted approaches. These video-assisted approaches provide access with excellent visualization to both the anterior and posterior rib. The absence of dissection of the neurovascular structures may reduce the risk of injury, though chest drainage is routinely required. These techniques may not replace supraclavicular approaches in NTOS where dissection of the plexus with neurolysis is required, but published series have demonstrated their safety and they may lend themselves particularly to VTOS surgery.[8]

Outcome

Functional outcome according to objective measures such as the QuickDASH (disabilities of the arm, shoulder and hand) questionnaire is excellent following successful lysis and surgery, intermediate in those undergoing delayed decompression and poor in those treated conservatively. Recurrent venous thrombosis is rare, but when it occurs repeat lysis can be offered. CT venography is recommended to assess if there is ongoing bony impingement of the vein and venography can assess whether further balloon venoplasty is required of a fibrotic segment.

Arterial TOS

ATOS is the least common of three variants of TOS. Complicated ATOS, where there is arterial damage, is usually associated with an anatomical abnormality; either a cervical rib, ligamentous band or previous clavicle fracture. Uncomplicated ATOS with intermittent positional ischaemia is often seen alongside NTOS due to the proximity of the subclavian artery to the brachial plexus in the costoclavicular space.

Clinical features

Patients with uncomplicated ATOS present with loss of strength and aching of the forearm and hand whilst working. This is exacerbated with the arm elevated and common precipitating activities include driving, holding a telephone, computer use and hair drying. ATOS often coincides with paraesthesia in the C8/T1 nerve root distribution because the lowest trunk of the brachial plexus lies immediately behind the subclavian artery. Sympathetic dysfunction with unilateral Raynaud's phenomenon can occur and may result from irritation of the peri-arterial sympathetic plexus. Delayed presentation of ATOS can have serious consequences as post-stenotic aneurysmal dilatation of the subclavian artery can give rise to digital emboli or limb-threatening acute limb ischaemia secondary to thromboembolism from a post-stenotic subclavian aneurysm (Figure 6).

Diagnosis

Bony anomalies in association with ATOS are often visible on plain X-ray of the lower cervical spine. An elongated transverse process of C7 may been seen, which is associated with a scalene band often extending to the scalene tubercle of the first rib (Figure 7). Ultrasound duplex sonography may de diagnostic in uncomplicated ATOS with occlusion of the artery in abduction (Figure 3). When intervention is planned for complicated ATOS, CT angiography (CTA) is essential, providing 3D reconstruction of the vessels and corresponding bony anatomy (Figure 6).

Treatment

ATOS resulting from intermittent symptoms of brachial ischaemia with normal anatomy is often associated with poor posture, shoulder injury or repetitive movement. A combination of reassurance, physiotherapy focusing on postural improvement and workplace adaptations can be sufficient in many cases. As with uncomplicated NTOS, surgery in the form of scalenectomy or first rib resection should only be offered if a trial of multimodal treatment has failed. Diagnostic local anaesthetic block also has a place in determining if there is compression in the pectoralis minor space.

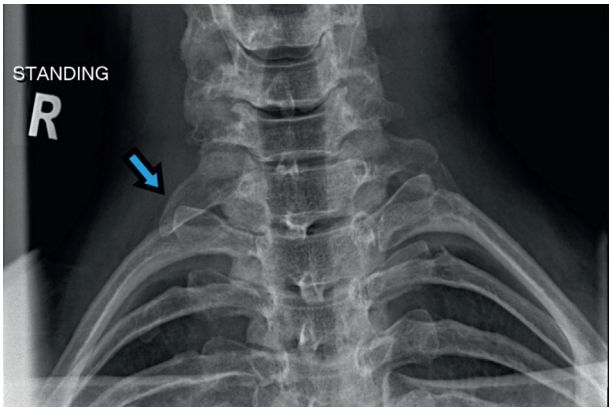

Figure 7 A plain X-ray of prominent C7 transverse process indicative of a fibrous band.

Complicated ATOS is a different matter as the threat of distal emboli must be considered, particularly in the presence of a subclavian aneurysm which contains thrombus on duplex scanning or where digital embolization has occurred. The patient should be started on an antiplatelet agent immediately. CTA is performed to identify the underlying bony abnormality, to assess run-off vessels in the event of distal embolization and to plan reconstruction (Figure 8). Surgery is best achieved from a supraclavicular approach. Anterior scalenectomy facilitates proximal control of the subclavian artery, which is controlled with slings after division of the transverse cervical artery. Retraction of the artery exposes the underlying cervical rib or band, which is carefully resected, including the scalene

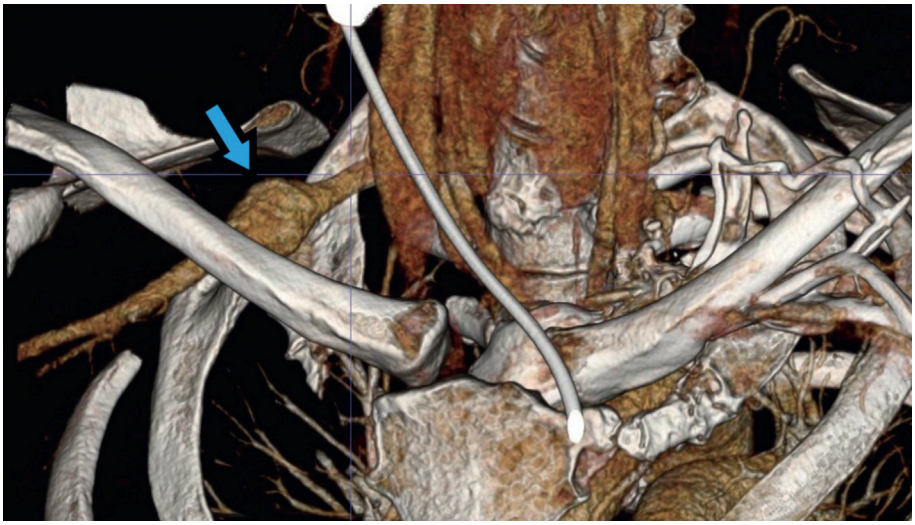

Figure 6 A CT angiogram with 3D reconstruction showing a cervical rib and its relationship to the broad and flat first rib below.

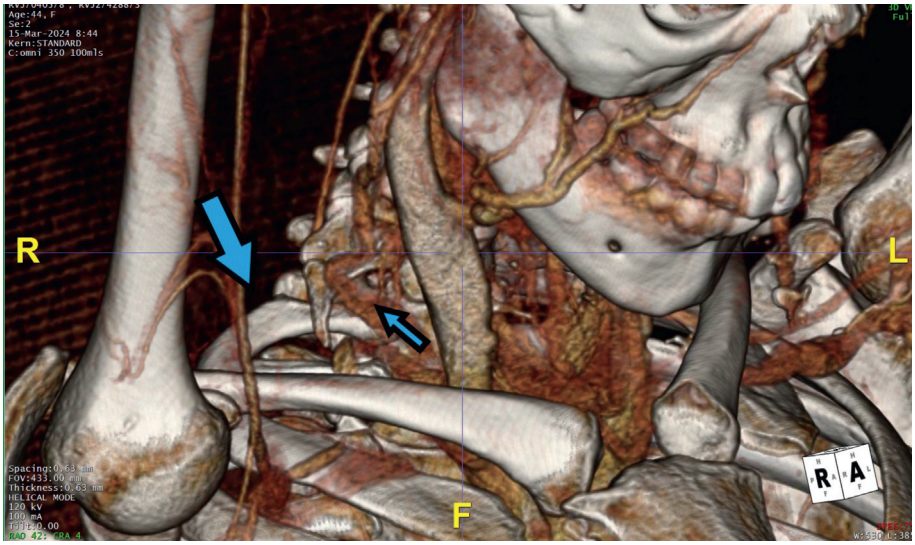

Figure 8 CT angiogram with 3D reconstruction showing a subclavian artery aneurysm secondary to a cervical band.

tubercle. Where there is a large pseudoarthrosis with the first rib, first rib resection may also be required for adequate decompression. In some cases, resection of the aneurysmal portion of the artery and direct end-to-end anastomosis is possible, but more often an interposition bypass is required. In acute limb ischaemia resulting from brachial embolization, brachial embolectomy and forearm fasciotomies may be required (Figure 9).

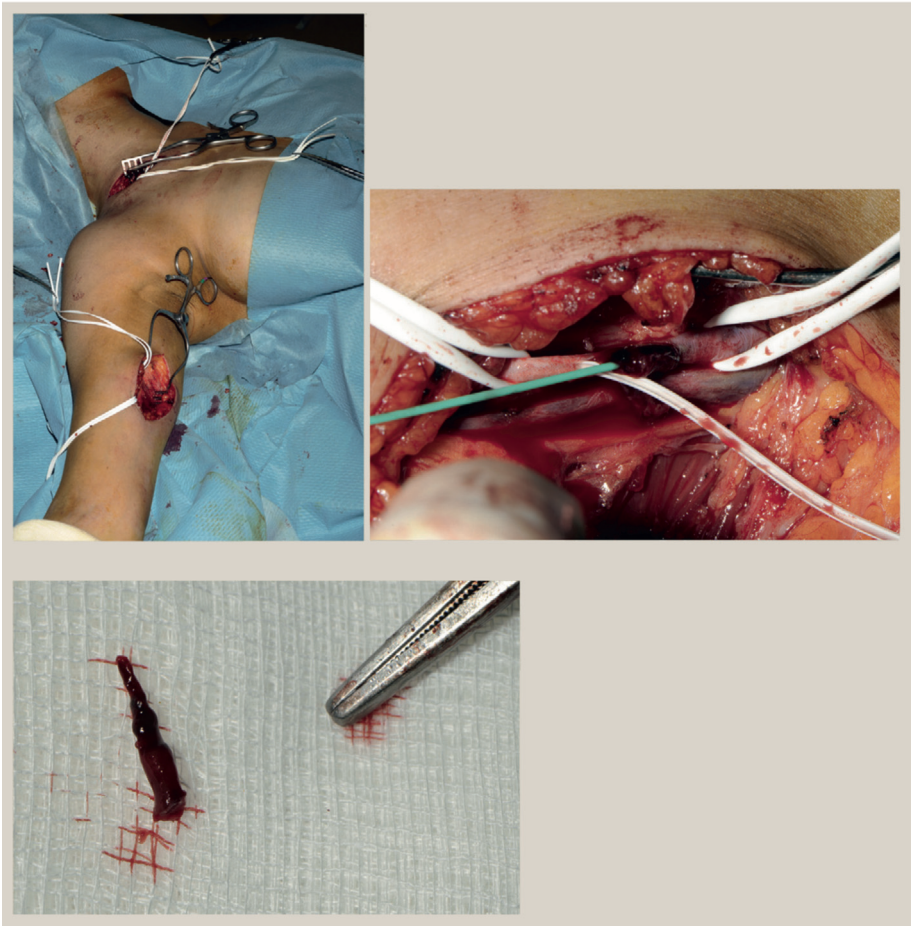

Figure 9 Right brachial embolectomy for arterial occlusion caused by thromboembolus from a subclavian aneurysm, secondary to compression by cervical rib, which was resected.

Summary

People with upper limb symptoms and suspected neurological, venous and/or arterial thoracic outlet compression need careful history taking and clinical examination. This is both to establish a probable diagnosis and exclude other causes of their symptoms. Multi-modality imaging is needed to confirm the diagnosis and the site of the compression. Conservative management may be appropriate for neurological, venous and uncomplicated arterial TOS. When surgical decompression is indicated this may involve resection of bone (i.e. cervical or first rib), muscle (i.e. scalenus anterior) and tendons (pectoralis minor) and necessitates different operative approaches dependent on the type and level of compression. ◆

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Practice points

- Thoracic outlet syndrome (TOS) should be considered as a cause of shoulder girdle or upper limb pain
- The presence of a cervical rib can be associated with TOS but compression may equally be from other structures
- Venous impingement may present as acute upper limb DVT. Treatment with anticoagulation and venous lysis with rib resection should be considered
- Arterial impingement may present with upper limb or digital ischaemia. Treatment may require both decompression and arterial repair
- Neurological compression is the most difficult to diagnose and treat. As with other TOS, its management should be under a specialist multidisciplinary team and conservative measures should be tried prior to surgery