Cervical Ribs and Thoracic Outlet Syndrome

Description

Cervical ribs are an anomaly that arise from the lowest cervical vertebrae but their relationship to thoracic outlet syndrome (TOS) is not so constant that the two conditions should be seen as synonymous. Perhaps no more than 10% of people who have cervical ribs develop TOS and the syndrome may well occur in the absence of ribs. Diagnosis of TOS may be difficult and depends upon a thorough history and examination together with supportive tests.

Two main types have been identified - vascular and neurogenic. The vascular type can be further categorised into arterial or venous. Neurogenic TOS can be subcategorised into true or disputed. The true form is accompanied by objective findings whereas the disputed form (which is in fact far more common) is not.

Epidemiology

Because of a lack of a gold standard for diagnosis it is difficult to obtain meaningful epidemiological figures. Differences between disciplines exist with reports that surgeons diagnose the condition 100 times more frequently than neurologists. Irrespective of the overall incidence, it is estimated that over 90% of all TOS cases are neurogenic, whereas 3-5% are venous. Less than 1% are arterial. The true neurological type probably affects no more than 1 person in 1 million. The overall incidence is given as between 3 and 80 per 1,000. Onset is from the 2nd to the 8th decade with a peak in the 4th decade. It is more common in women than in men, with an excess of between 3- and 9-fold.

Analysis of 1,352 chest radiographs in London suggested the prevalence was 0.74% with a higher rate in females compared with males.

Presentation

- Symptoms will depend on the type of TOS. In the neurogenic type, there may be painless wasting of the muscles of the hand, resulting in weakness (eg, difficulty in grasping a racket). Numbness or tingling of the upper limb may be reported. Symptoms are often vague and general and may affect the whole arm. There may also be a painful neck and headache.
- If there is compression of autonomic nerves, cold hands, swelling or blanching can occur. Stellate ganglion involvement may be possible.
- Vascular TOS is seen less frequently than the neurogenic type. If the subclavian vein is compressed, there may be swelling of the arm, distension of the veins or a diffuse pain in the arm or hand.
- If the subclavian artery is compressed, patients may notice colour changes, claudication or a vague pain in the arm or hand. Early symptoms may be ignored and patients may not seek medical advice until the condition becomes worse, with the development of ulceration or gangrene.
- Pure types are rare and patients often present with symptoms indicative of more than one type.
- There is often a history of trauma. This may be whiplash from a road traffic accident. It may occur in sportsmen, especially swimmers and throwers.
- The interval between trauma and symptoms may be hours to weeks.

Examination

A careful neurological and musculoskeletal examination is required. Neurological examination is discussed elsewhere - see separate Neurological Examination of the Upper Limbs article. Disease of the neck and arm must be excluded. Stress tests or provocative manoeuvres form the basis of examination. They have very low specificity, sensitivity and predictive value.
The most common tests are Adson's manoeuvres. The head is extended and bent to one side and the patient takes a deep breath and holds it, followed by rotation to stretch or tether the brachial plexus and/or artery between the anterior and middle scalenes. The position is held for 15-30 seconds while the examiner checks for the onset of symptoms and obliteration of the pulse.

Symptoms have been reported both to the side of bending and, more commonly, to the opposite side. If the symptoms occur on the side of bending, this is consistent with Spurling's sign for the diagnosis of cervical radiculopathy. This sign is described further in the separate Cervical Disc Protrusion and Lesions article. Some clinicians ask the patient to pull the head forward while maintaining the test position, causing the anterior scalene to contract against the plexus to enhance the stress effect.

Hyperabduction of the arm can also be used as a test to stress the outlet. This often causes symptoms and loss of pulse even in normal people and may be misleading.

Costoclavicular bracing reduces the space between the clavicle and first rib and may reproduce symptoms.

Focal stress tests involve direct application of pressure to the anterior scalene or upper segment of the pectoralis minor. A positive result is if symptoms are reproduced within 15-30 seconds. Some people use Tinel's sign, where percussing over the plexus reproduces symptoms.

The elevated arm stress test is sensitive. The upper extremity is held in the 'hands-up' position with the arms abducted and elbows flexed at 90° for three minutes, while the patient vigorously flexes and extends the fingers. A positive sign is if the patient cannot complete the full three minutes. This test is demanding even for those without neurovascular symptoms and so its practical application is limited.

The upper chest wall may be asymmetrical after a previous fracture of a clavicle. A non-tender, hard mass over the middle third of the clavicle is typical. A fracture that failed to unite or has excessive callus can cause direct compression of the plexus. Pressure on the clavicle can produce or aggravate symptoms, especially an ununited fracture. Motion can be felt between the fragments.

Aetiology

- Cervical ribs or fibrous bands are just one feature that predisposes to narrowing and compression at the outlet.
- Poor posture can produce mechanical problems. Sometimes people who are depressed or just have a bad habit with regard to posture let the head drop forward; shoulders droop and this allows the thoracic outlet to narrow and compress the neurovascular structures.
- Large breasts can pull the chest wall forward and cause symptoms. Reduction mammoplasty may have a beneficial effect. Sometimes the cause may be excessively large breasts that are not natural but have been surgically augmented.
- Trauma can move structures in the shoulder and chest wall. Fracture of the clavicle can cause compression by bone fragments, excessive callus, haematoma or pseudoaneurysm.
- Some of the typical precursors of myofascial pain can cause this syndrome too: sleep disorder, oestrogen or thyroid deficiency, inflammatory disease including rheumatoid arthritis, fibromyalgia and disorders of posture such as kyphosis and scoliosis.
- It is necessary to exclude thrombosis, embolism and nerve entrapment in other places. This includes Pancoast's syndrome, where lung cancer infiltrates the brachial plexus. Paget-Schrötter syndrome is thrombosis of the subclavian vein following heavy exercise of the upper limb.

Differential diagnosis

- Acromioclavicular joint injury.
- Brachial plexus injury.
- Cervical disc injuries.
- Cervical discogenic pain syndrome.
- Cervical radiculopathy.
- Clavicular injuries.
- Elbow and forearm overuse injuries.
- Shoulder impingement syndrome.
- Thoracic disc injuries.
- Thoracic discogenic pain syndrome.
Investigation

- The value of blood tests is to exclude other conditions.
- CXR with apical lordotic views and cervical spine should be obtained. An X-ray may show cervical ribs, which may be the cause, or have fibrous bands with them. It may also show elevated first ribs, caused by tight anterior or middle scalene muscles. Displaced fractures of clavicle, non-union and excessive callus may be apparent. Cervical spine degenerative changes may be causing neck or shoulder pain or impingement of the spinal nerve roots. Exclude a malignant lesion in the chest.
- MRI scan of the cervical spine and supraclavicular or brachial plexus area is useful to find other causes. A CT scan of the area of the brachial plexus and apex of lung may be indicated. MRI and CT can distinguish cervical root injury from degenerative spurs, herniated discs or other causes. Adjunctive tests such as CT angiography can be helpful in difficult cases.
- Doppler and plethysmography studies can show impediment of blood flow. A near-complete cut-off of flow during the stress manoeuvre with reproduction of the symptoms would be most impressive. Occlusion can occur in normal subjects but is unusual and is not related to age.
- Angiography and venography can show blockage of the vessels from thrombi or emboli. Angiography can demonstrate aneurysms that may be compressing the plexus and causing neurological features.
- Asymmetry of temperature suggests interference with blood flow from obstruction of autonomic dysfunction. It should improve with successful treatment. The technique is controversial as the sensitivity is high but the specificity low.
- An anterior scalene block is sometimes used as a diagnostic test but it is both difficult and dangerous, as the needle may damage the nerve. However, if it does give positive results then there is a good chance of successful surgical decompression.
- Musculoskeletal sonography may be useful in children.

Treatment

The diffuse and uncertain nature of this condition makes the application of randomised controlled trials (RCTs) very difficult. Because of this, the evidence base supporting the use of any treatment other than some specific surgical options is limited.

- Conservative management is generally considered first-line (but see early surgical intervention, below). In cases where postural deviations contribute substantially to compression of the thoracic outlet, rehabilitation helps to decompress the outlet. This includes graded restoration of scapular control at rest and through movement. Humeral head control, isolated strengthening of weak shoulder muscles, taping and other manual therapy techniques are other adjunctive approaches.
- Occupational therapy may help with back protection techniques and better working practices.
- Injection of trigger points and associated muscles may be necessary. An injection of deep muscular structures, as in a scalene block, is dangerously close to the brachial plexus.
- Non-steroidal anti-inflammatory drugs (NSAIDs) have a good analgesic as well as anti-inflammatory action and it may be the former that is rather more important.
- Some people recommend muscle relaxants such as methocarbamol but they cause sedation and the benefit is unclear.
- Calcium-channel blocking agents may possibly be of value where there is vascular instability.
- One study found that early surgical intervention leads to better functional outcome than late surgery. This is because early intervention prevents degeneration of the brachial plexus and increases the effectiveness of postoperative physiotherapy.
- Surgical intervention may help where there is an obvious physical lesion but, in many cases where this is not the case, it not only fails to improve matters but may lead to deterioration. Damage to the long thoracic nerve or the brachial plexus is a possibility. Patients therefore need careful appraisal before neurosurgery.
- Paget-Schrötter syndrome requires thrombolysis followed by surgical decompression of the subclavian vein. This gives rather better results than more conservative practices such as anticoagulation.
- Some surgeons resect the first rib through a transaxillary approach and others remove the scalene muscles too. Cervical ribs and fibrous bands should be removed if they are tethering the plexus.
- Another option is decompression with neurolysis of the involved regions of the brachial plexus, especially the C7, C8 and T1 nerve roots, through a supraclavicular approach (supraclavicular "neuroplasty").
- There is some evidence that transaxillary rib resection is more effective than supraclavicular neuroplasty in relieving pain but large RCTs are needed.
Complications and prognosis

Generally prognosis is good, unless the condition is severe enough to merit surgery and spontaneous recovery will occur. However, sometimes complications occur:

- Chronic pain.
- Loss of function.
- Depression.
- Neurological complications.
- Thrombosis, ischaemia or pseudoaneurysm.
- Post-thrombotic syndrome of the lower limb following deep vein thrombosis is well recognised but problems may also follow thrombosis of the upper limb, although they have been less well documented [21].

Further reading & references

- Where the clavicle has been fractured, the removal of excessive callus may be required. Fixation of a fracture that has failed to unite may be required.
- As mentioned earlier, reduction of very large breasts may be beneficial.
- Botulinum toxin may help with symptom relief but further research is needed [20].

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