Arterial thoracic outlet compression syndrome: a differential diagnosis of painful right supraclavicular swelling?

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DISCUSSION

The narrow triangle described by the first rib inferiorly, the scalenus medius posteriorly and the scalenus anterior anteriorly confines the neurovascular structures of the upper extremities as they exit from the mediastinum and the spinal canal. In patients with cervical ribs or abnormal congenital fibrous connections between the cervical transverse process and the first rib, the base of the triangle maybe raised to the height of the vertebrae resulting in the angulation or compression of the neurovascular structures. This compression and angulation results in the syndrome known as thoracic outlet compression syndrome (TOCS) (Mann et al, 1995).

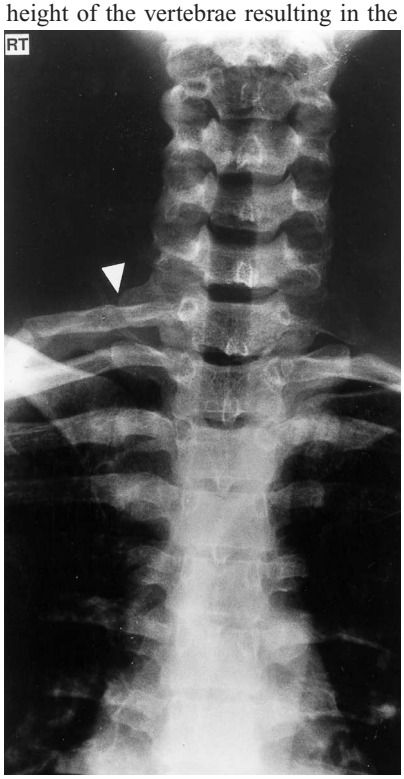

Figure 1. Thoracic inlet view showing a well developed right cervical rib.

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Arterial complications of TOCS are uncommon, accounting for only 5–10% of cases (Durham et al, 1995).

When subclavian artery compression is present in this condition, it is usually related to cervical ribs. Mayo (1831) and Murphy (1905) made the first reports of the association between a cervical rib and a subclavian artery aneurysm (Durham et al, 1995). Cervical ribs occur in about 1% of the population and can vary from a small structure to a complete rib articulating with the manubrium. In about 50% of cases, it is bilateral.

Between 5 and 10% of patients with cervical rib will have symptoms and, of these, more than 50% will have vascular lesions usually affecting the subclavian artery (Young and Hardy, 1983). Less common causes of vascular complications include first rib and clavicle fractures, clavicle pseudoarthrosis or hyperostosis, spasms of the pectoralis minor caused by costochondritis and enlargement of the seventh cervical transverse process (Baumgartner et al, 1989).

The diagnosis of arterial TOCS is often delayed and is usually made when there is structural damage of the artery resulting in thromboembolic complications (Casselman et al, 1997). The subclavian artery should be auscultated for the presence of a bruit. Adson's and Allen's manoeuvres are only rarely helpful because they are present in a large proportion of the normal population (Riddell and Smith, 1986).

Scher et al (1984) classified the three stages of arterial injury based on 12 cases:

CASE REPORT

A 16-year-old female presented to the emergency department complaining of a painful swelling above the right clavicle. She noticed this after running into a door 4 months before presentation.

The swelling was described as intermittently painful, however, the size remained constant. One month before her presentation, she developed pain and weakness down the right arm and hand. She also noticed a marked coolness of the right hand compared to the left side. These symptoms gradually improved over a period of hours. Four hours before presentation to the department, she had symptoms similar to those described above, which improved over a period of hours.

Clinical examination revealed a tender pulsatile mass over the right supraclavicular region. There was a well-defined thrill and a bruit. She had a full compliment of pulses in the upper limb and a demonstrable radial pulse even with the shoulder fully abducted. An impression of a subclavian aneurysm was entertained and a thoracic inlet and chest radiograph requested. This demonstrated a fully developed right cervical rib (*Figure 1*).

She was admitted under the vascular specialties. Further investigations were requested including an ultrasonogram (which failed to define the subclavian artery) and an intravenous arteriogram, which demonstrated a sharp angulation in the right subclavian artery, a post-stenotic dilatation but no thrombus and full flow (*Figure 2*).

Five days after her admission, she underwent surgery which demonstrated a large cervical rib with a fibrous band corresponding to the level of angulation of the subclavian artery, but no subclavian artery aneurysm. The cervical rib was excised along with the fibrous band. Postoperative recovery was uneventful.

Figure 2. Intravenous arteriogram showing angulation in the right subclavian artery with a post-stenotic dilatation.

1. Cervical rib associated with stenosis, often with minor post-stenotic dilatation
2. Intimal lesions, intraluminal thrombus or subclavian aneurysm
3. Arterial lesions with distal thromboembolic complications (Baumgartner et al, 1989).

Regardless of the cause of arterial TOCS, standard therapy requires decompression of the thoracic outlet, i.e. removal of the offending compressive element, whether the element is soft tissue or a bony structure, provided there is no suggestion of arterial damage. In some cases, repair of the subclavian artery to restore blood flow is necessary. This is applicable in cases with aneurysmal degeneration of the artery or intimal damage (with or without embolic sequelae). Resection of the damaged artery by arterial reconstruction with the use of a saphenous vein graft is recommended (Durham et al, 1995).

This patient presented with a palpable pulsatile mass believed to be a subclavian artery aneurysm. Following decompression of the thoracic outlet, by resection of the first rib and cervical rib, the subclavian artery was found to be of normal diameter. This is an interesting observation and appears to be an example of a 'haemodynamic aneurysm', where the dilatation occurs in a normal artery between the proximal stenosis and the distal peripheral resistance (Thompson and Webster, 1990). Decompression of the thoracic outlet appears to have removed this distal peripheral resistance and resulted in the subclavian artery returning to its normal diameter.

CONCLUSION

TOCS occurring in teenagers is rare. Unilateral symptomatic cervical rib is even rarer. Our patient initially presented with a painful clavicular swelling following trauma 4 months before presentation. The trauma may have precipitated the symptoms of transient arterial insufficiency as various papers have demonstrated a relationship between trauma and arterial TOCS. Neurogenic TOCS presents with pain but this is usually over the shoulder and the arm. There was no evidence that our patient had a concomitant neurogenic TOCS.

Finally, it is important that arterial TOCS is recognized and treated in the early stages to ensure preservation of limb and prevent the very real possibility of amputation. **HM**

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