Neurogenic thoracic outlet syndrome associated with cervical rib

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ABSTRACT

A true neurogenic thoracic outlet syndrome (TOS) associated with a cervical rib is considered extremely rare.

The authors present their experience with 5 cases of true neurogenic TOS associated with a cervical rib. All patients were female and had a cervical rib confirmed radiographically pre-operatively. Average age was 34,8 years. Although all patients had been treated with several combinations of diverse drugs and a rehabilitation program before referral to surgery, all described their pain as intense and debilitating before surgical treatment. All patients had pre-operative electromyographic abnormalities.

Patients were operated on via a supraclavicular approach and the cervical rib was resected. No intra-operative or postoperative complications were noted.

Two years postoperatively, all patients mentioned improvement. However, only 2 were symptomless, and on no medication. In one patient there was significant improvement, and in the remaining 2 patients some residual pain persisted that had to be dealt with pharmacologically. All patients were able to resume their daily life activities.

Recovery was poorer in the 2 patients that had been referred to surgery after a longer period of time since the beginning of symptoms.

Keywords: Thoracic outlet syndrome; Brachial plexus; Cervical rib; Nerve compression syndrome; Surgery.

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RESUMO

Os síndromes de desfiladeiro torácico neurogénicos puros associados a uma costela cervical são considerados muito raros.

Os autores apresentam a sua experiência no tratamento de 5 doentes nestas circunstâncias. Os doentes eram todos do sexo feminino e tinham uma costela cervical confirmada radiologicamente pré-operatoriamente, bem como alterações electromiográficas. A idade média era de 34,8 anos. Embora todas as pacientes tivessem sido tratadas com uma combinação de fármacos e com um programa de reabilitação antes de serem referenciadas para cirurgia, todas descreviam uma dor intensa, incapacitante e persistente. As doentes foram operadas por abordagem supraclavicular, tendo sido ressecada a costela cervical.

Não se registaram complicações intra ou pós operatórias. Dois anos após a cirurgia, todas as doentes apresentavam melhoria clínica. Contudo, apenas 2 estavam completamente assintomáticas, sem qualquer medicação. Numa doente registou-se melhoria significativa e nas duas restantes havia persistência de dor residual que tinha de ser controlada farmacologicamente. A recuperação foi pior nas duas doentes que tinham sido referenciadas para cirurgia ao fim de mais tempo desde o início da sintomatologia. No entanto, todas as pacientes retomaram as suas actividades quotidianas.

Palavras-chave: Síndrome do desfiladeiro torácico; Plexo braquial; Costela cervical; Síndrome compressivo nervoso; Cirurgia.

INTRODUCTION

Thoracic outlet syndrome (TOS) is defined as a group of symptoms that result from the entrapment of the brachial plexus and/or subclavian vessels in the thoracic outlet region, that is to say, between the neck and the axilla[1-4]. The name of the syndrome was coined in 1957 by

Peet[5-7]. However, it has also been referred to as scalenus anticus syndrome, cervical rib syndrome, costo-clavicular syndrome, and hyperabduction syndrome[7,8].

Most authors agree that the TOS is the most overlooked and misdiagnosed nerve entrapment syndrome in the upper limb, as well as one of the most debilitating, and certainly one of the most difficult to manage[8-11]. The complexity in the diagnosis is due to the lack of any specific clinical or ancillary confirmatory tests[1,7,11,12]. The reported incidence of TOS varies between 3 to 80 cases /1.000 inhabitants, making it a significant pathology for anyone who deals with upper limb pathology[1,13].

TOS has been the subject of various classifications[1], being the Wilbourn's classification the most widely used in clinical practice[7]. According to this classification, TOS can be of two main types: vascular and neurogenic[4]. The vascular type is subdivided into arterial and venous. The neurogenic type can be "true" neurogenic or "disputed" neurogenic, if nerve conduction studies show changes or not, respectively[7]. Vascular TOS corresponds to about 1% of cases, whereas the "disputed" TOS accounts for up to 97% of patients diagnosed with this syndrome[7,12]. True neurogenic TOS is rare, having a prevalence of around 1 in 1 million patients[7,11]. In these patients, there is usually an anatomical anomaly causing brachial plexus compression, namely a cervical rib, a prolonged transverse process of the seventh cervical vertebra, an anomalous first rib, first rib or clavicle fracture's, a scalenus minimus, subclavius tendon anomalies, anomalies of scalene muscle development or insertion, and thick fibrous bands in the thoracic outlet region[7,11,13,14]. However, a true neurogenic TOS associated with a cervical rib is considered extremely rare[7,11,13,14]. For example, among the more than 1000 operations performed for neurogenic TOS during a period of over 28 years in a tertiary referral centre, Sanders et al. found only 37 patients with cervical ribs[13].

In the present work, the authors present their experience with 5 cases of true neurogenic TOS associated with a cervical rib.

CLINICAL CASES

The authors retrospectively reviewed the charts, and clinical images of 5 patients referred to either the Brachial Plexus, Vertebro-medullary or Neurosurgery Outpatient Clinic at São José Hospital (Lisbon, Portugal). The basic demographic features, clinical picture and outcome one year after surgery of those 5 patients are described in Table I.

All patients were female and had a cervical rib confirmed radiographically pre-operatively (Fig. 1). Average age was 34.8 ±19.6 years, ranging from 18 to 65 years. All patients had been treated conservatively with several combinations of drugs (analgesics, anti-inflammatory drugs, muscle relaxants, and antidepressants), and a rehabilitation program before referral to the Clinic. The average time from the onset of symptoms to referral to our Clinic was 2.9 ± 2.4 years, varying from 1 to 7 years.

Three patients experienced neck and upper limb pain and paresthesias, whereas 2 patients had complaints only at the upper limb. All patients described the pain as intense, debilitating and persisting. Two patients were unable to work due to their symptoms, and one of them (patient 4) had been forced to give up gymnastics which she had been performing competitively for the 10 years before the onset of symptoms. Other patient (patient 3), although retired, complained that she could no longer do most of her daily life tasks. All patients had positive provocative tests (Adson's, hyperabduction, costo-clavicle, Halsted's, Roos's, Wright and upper limb tension tests). Diminished strength in the intrinsic hand muscles was noted in 2 patients: in the territory of the ulnar nerve in one patient, and in the territory of both the ulnar and median nerve in another patient. Hand muscle wasting was observed in 2 of these patients. All patients had electromyographic abnormalities, as depicted in Table I.

Patients were operated on via a supraclavicular approach, isolating the constituents of the brachial plexus, the subclavian artery and vein and the cervical rib (Figs. 2 and 3). This rib was resected, freeing the brachial plexus (Fig. 4). No intra-operative or postoperative complications were noted, being the patients discharged home a few days after the surgery (3 to 5 days).

Patients were followed up for a minimum of two years after surgery.

Two years postoperatively, all patients mentioned improvement. However, only 2 were symptomless, and on no medication. One had occasional hand pain that she controlled satisfactorily with analgesics. Two others, reported residual hand pain for which they continued to receive pain killers, with reasonable pain control. All patients resumed their prior daily life activities. Patients who had muscle wasting and diminished muscle strength recovered only partially from these defi-

TABLE I. SUMMARY OF THE DEMOGRAPHIC FEATURES, CLINICAL PICTURE AND OUTCOME TWO YEARS AFTER SURGERY OF 5 PATIENTS WITH NEUROGENIC THORACIC OUTLET SYNDROME ASSOCIATED WITH A CERVICAL RIB. F- FEMALE.

Patient	Age (years)	Sex	Occupation	Affected Side	Complaints of symptoms	Physical exam	Electrodiagnostic tests	Time from the development surgery to surgery (years)	Outcome two years after
1	29	F	Cruise ship stewardess	Left	Neck and upper limb pain and paresthesias; Unable to work	Positive provocative tests; Atrophy and diminished strength of intrinsic hand muscles	Chronic denervation in the territory of the inferior trunk of the brachial plexus	2	Occasional hand pain managed with pain killers; Partial recovery of strength and intrinsic hand muscles bulk; Resumed working
2	43	F	Office clerk	Left	Debilitating neck and upper limb pain and paresthesias; Unable to work	Positive provocative tests	Chronic denervation in the territory of the inferior trunk of the brachial plexus	3	Residual hand pain managed with pain killers; Resumed working
3	65	F	Retired	Right	Debilitating neck and upper limb pain and paresthesias; Allodynia in the ulnar nerve territory; Unable to perform most daily life activities	Positive provocative tests; diminished strength in the territory of the ulnar and median nerves; atrophy of intrinsic hand muscles	Chronic denervation in the territory of the middle and inferior trunks of the brachial plexus	7	Residual hand pain managed with pain killers; Partial recovery of strength and intrinsic hand muscles bulk; Resumed most daily life activities
4	19	F	Student	Right	Intense upper limb pain and paresthesias; Unable to exercise	Positive provocative tests; diminished strength in the territory of the ulnar nerve	Denervation in the territory of the inferior trunk of the brachial plexus	1.5	Symptomless
5	18	F	Student	Right	Intense upper limb pain and paresthesias	Positive provocative tests	Denervation in the territory of the inferior trunk of the brachial plexus	1	Symptomless

FIGURE 1. Radiograph showing a right cervical rib (asterisk).

FIGURE 2. Intraoperative view of the supraclavicular contents of the thoracic outlet in a patient with a cervical rib.

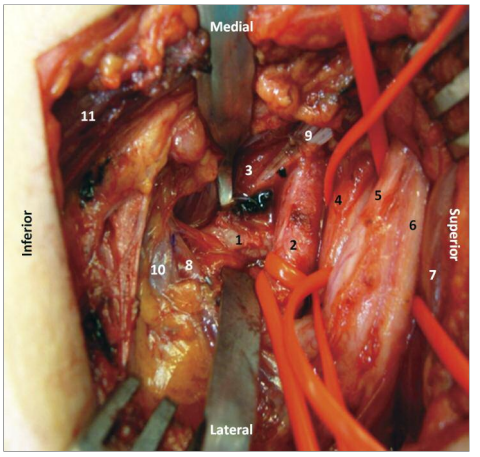

FIGURE 3. Photograph of the thoracic outlet in a close-up view showing the intimate relation between the cervical rib and the subclavian artery and the lower portion of the brachial plexus. 1- cervical rib; 2- subclavian artery; 3- subclavian vein; 4- inferior trunk of the brachial plexus; 5- middle trunk of the brachial plexus, 6- superior trunk of the brachial plexus; 7- trapezius muscle, 8- subclavius muscle; 9- anterior scalene muscle, 10- clavicle; 11- sternocleidomastoid muscle.

FIGURE 4. Photograph of the cervical rib after resection.

cits. All surgical wounds healed uneventfully (Fig. 5).

DISCUSSION

Cervical ribs are usually asymptomatic and thus found incidentally on routine image exams, being present in 0,01 to 0,5 % of the general population[1,13,15]. These supernumerary ribs are more common in women, being bilateral in more than half of cases[7,13]. However, the cervical rib is considered a risk factor for the development of TOS following cervical trauma, particularly after whiplash injuries[12,13]. In fact, it has been proposed that TOS is due to a combination of a constitutional tendency coupled to muscle dysfunction and repetitive trauma[4,5].

The existence of a higher placed bone piece, usually connected to the first thoracic rib directly or through a thick fibrous band, narrows the thoracic outlet. Hence, people with these anatomical variants are more prone to TOS, especially if they do sports or jobs that demand prolonged arm hyper-abduction, like swimming, gymnastics or weight throwing[1,7,16]. Supporting this way of thinking is the report by Boles et al. of 15 TOS patients in a single family, with a cervical rib or an apophysomegaly of the seventh cervical vertebra[17]. Interestingly, in our series one patient was an active

FIGURE 5. Photograph of the surgical incision 4 months after surgery.

gymnast before the development of symptoms.

In the cases we reviewed, only female patients were found. This is in accordance with the literature that usually states that TOS is almost twice as common in women than in men[7,8,18]. This sex predilection is attributed to the fact that usually women present comparatively weaker muscles which makes their scapulas to be placed lower, predisposing to the compression of the structures involved in TOS[1,7]. Authors usually agree that most TOS patients present between 20 to 50 years[7,8,18]. In our series this was also observed, as all patients were in this age interval except for one.

The differential diagnosis of TOS is extensive, including cervical disc disease; osteophytes; neoplasms (e.g. Pancoast tumor, nerve sheath tumors, spinal cord tumors) peripheral nerve entrapments (ulnar and/or median nerve entrapment); brachial plexitis; shoulder pathology (e.g. rotator cuff injuries), muscular spasms; fibromyalgia; multiple sclerosis; coronary artery disease; vasculitis (e.g. Takayusu's arteritis); Raynaud's phenomenon; complex regional pain syndrome; venous thrombosis, micro-embolism; hand–arm vibration syndrome; and myofascial syndrome[1,8,19].

The diagnosis of TOS is made by the patient history associated with physical examination[1,8]. The most common complaints are chronic pain of insidious onset involving the shoulder girdle, neck and upper back, combined with paresthesias and hypoesthesia in the medial arm and forearm, and in the territory of the ulnar nerve and/or the median nerve[1,8,19]. Provocative tests (e.g. Adson's, hyper-abduction, costo-clavicle, Halsted's, Roos', Wright and upper limb tension tests) are frequently positive in the normal population, and are neither sensitive nor specific for TOS[1,4,8,19]. Their value is therefore limited[1,4,8,19].

Regarding ancillary diagnostic tests, a chest radiography in an apical lordotic view or a cervical radiography may allow identification of bone abnormalities that may predispose to TOS, narrowing down the differential diagnosis[20]. Ultrasonography, computed tomography scans and magnetic resonance imaging, sometimes performed in association with postural maneuvers, are helpful in analyzing the dynamically induced compression, as well as places of neurological compression[7,19,20]. Electromyography and nerve conduction velocity tests are normal in the large majority of patients with clinical signs of TOS, which led some authors to argue that TOS is frequently underdiagnosed in the primary care setting and over-diagnosed in patients demanding compensation from insurance companies[7,13].

It is generally accepted that the first line of treatment of TOS should be conservative, including combinations of several drugs (analgesics, anti-inflammatories, muscle relaxants, antidepressants, and particularly the anticonvulsants gabapentin and pregabalin), avoiding activities and positions that aggravate symptoms, and rehabilitation with strengthening of pectoral musculature[7]. The improvement after conservative treatment varies from 50 to 90% and depends on its etiology, being less efficient in "true" neurogenic TOS[1-3]. Surgery should be considered after failure of appropriate conservative treatment of 6 months' duration[1-3,7]. Surgical treatment involves surgical decompression by cervical rib excision and/or first rib excision; resection of cervical muscles, brachial plexus neurolysis; and, when necessary, vascular reconstruction. The presence of a cervical rib is not in itself an indication for surgery, unless there is failure in conservative treatment or debilitating symptoms[1,7,11,19].

Surgical treatment is considered successful in approximately 80% of selected TOS cases, being deemed unsatisfactory in around 20% of patients in the best series. In contrast, Sanders et al., reviewing the largest series of cervical ribs associated with TOS (n=37), considered their long term results as good to excellent in only 59%, and fair and poor in 13% and 28%, respectively[13]. These results are not significantly different from the ones we observed in the present study, in which, two years postoperatively, all patients had registered some improvement. However, in only 2 of the patients (40%) was the recovery complete. In 1 patient

(20%) there was significant improvement, and in the remaining 2 patients (40%) some residual pain persisted that had to be dealt with pharmacologically. However, surgery allowed all patients to resume their previous daily activities and occupations. Recovery was poorer in the 2 patients that had been referred to surgery after a longer period of time since the beginning of symptoms (3 and 7 years), which is also according to the literature[7,11].

Therefore, to maximize the recovery of TOS patients associated with cervical ribs, it is reasonable to suggest that these patients should be managed by a multidisciplinary team involving a rheumatologist, a pain specialist, a surgeon, and a physiotherapist, with possible advice from a psychologist or a psychiatrist, in order to maximize recovery and facilitate a rapid return to work[5,2].

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