Introduction

Thoracic Outlet Syndrome (TOS) is an entrapment syndrome where major vessels and nerves of the upper extremity, while travelling from the thoraco-cervical region to the axilla, are compressed due to narrowing of the spaces in the thoracic outlet. A number of different anatomic or functional characteristics can be attributed to produce the symptoms during elevation of the upper limbs. Common etiologies include scaring of the scalenus anticus muscle because of previous trauma, costoclavicular, hyper abduction, cervical rib, fibrous bands and first thoracic rib. Patients with TOS usually present with neurogenic, venous, and/or arterial symptoms [1-3]. The incidence of cervical ribs is 0.2% to 1.0% which is most often picked up incidentally on routine chest radiographs [4-7]. Most of these ribs are asymptomatic and necessitate no further management [8]. Gruber in 1869 was the first to describe cervical ribs [9]. He classified them into 4 types: In Type I the rib extends up to the transverse process of C7 whereas in type II it extends beyond the transverse process with no connection to the first thoracic rib. In type III it extends beyond the transverse process with partial fusion to the first rib by fibrous bands or cartilage and is seen completely fused to the first rib by a bony pseudo articulation in type IV. Type III and IV cervical ribs commonly lead to vascular compromise, including arterial compression, thrombosis, and aneurysm formation. About 95% of the patients have symptoms pertaining to neurogenic patients with venous TOS often presents acutely after either thrombosis of the subclavian vein or due to intermittent compression of the vein. Arterial TOS is the least common variety often presenting with acute or chronic secondary to aneurysm, occlusion, or embolization of the subclavian artery. These patients are frequently found to have bony abnormalities, such as cervical ribs or rudimentary ribs [10].

Different provocative tests can reproduce the symptoms. These include East test or "Hands-up" test, Adson or scalene maneuver, costoclavicular maneuver and Allen test. Doppler ultrasonography aids in the initial evaluation of the vascular flow through the compromised territory and distally. Helical Computed Tomography (CT) angiography can be diagnostic, to ascertain the extent of vascular compromise and also helps in establishing the adequacy of flow during the follow up course.
The initial plan of management conventionally is to continue conservative care where as surgical intervention is reserved in patients for acute vascular insufficiency, neurological dysfunction and pain refractory to conservative measures [11]. The surgical maneuver involves the release or removal of the confounding structures such as cervical rib excision, scalene muscle release, first rib resection and resection of fibro muscular bands [12].

**Case Report**

Thoracic Outlet Syndrome (TOS) is caused by impingement of the neurovascular structures passing through the thoracic inlet. We report a case of a 27 years old female who was referred to the Vascular Surgery Department of King Fahd Military Medical Complex from a peripheral hospital where she presented with complain of progressive left upper limb pain and paresthesias that started 4 weeks prior to presentation after elevating an object at home. The pain was gradually increasing in intensity more so with repeated movement of left arm at the shoulder joint. This was associated with left hand claudication and numbness. Initial workup showed left brachial artery thrombus for which she underwent brachial artery embolectomy. With this procedure her pain started improving but it did not subside and the numbness persisted in the left hand. She was thereafter shifted to our hospital after 10 days of initial procedure for further management.

On initial general examination, the patient is an average built, right handed lady sitting comfortably on the couch. A scar of previous surgical intervention was visible at the distal aspect of the left arm. Her left hand palmar surface was pale and cold on palpation. The capillary refilling was delayed with absent radial and brachial pulses, however with weak ulnar pulse. Neurologically she had normal left hand grip as compared to the right with a power of 5/5 in all muscle groups of left forearm and hand. There was no muscular atrophy with normal sensations and deep tendon reflexes. There was no restriction of movement at any of the joints in the left upper limb. Tip of left cervical rib was palpable with no cervical spine tenderness.

Laboratory investigations were unremarkable. Hand Doppler had shown no signals in radial and weak signals in ulnar and brachial arteries. CT angiogram revealed a filling defect measuring 40 x 11.5 mm at the second part of subclavian artery suggestive of thrombus due to cervical rib compression with non-visualized proximal segment of brachial artery (clavicle to the mid humerus), with narrowed and attenuated left axillary, brachial and radial arteries (Figure 1). Ulnar artery had normal caliber with no filling defect. Magnetic Resonance Imaging (MRI) of cervical spine and brachial plexus showed the left cervical rib crossing the left scalene triangle with no abnormal thickening or enhancement of the left brachial plexus nerve roots.

Thereafter patient and family members were counseled regarding our plan of excision of cervical rib with thrombectomy and stenting of the subclavian artery, to which they agreed. She underwent the above mentioned surgical procedure though a supraclavicular approach by the Neurosurgery team. Per operatively, a curvilinear incision in the supraclavicular region was given, and platysma opened. The phrenic nerve was identified on the surface of the anterior scalene muscle as it proceeds distally and was retracted medially with the anterior scalene muscle. Dissection was done up to the cervical rib which was identified compressing the vasculature, and excised along with release of the fibrous band (Figure 2). After excision of the rib, pulsations were visible in the previously compressed subclavian artery. Vascular surgery team performed catheter thromb-emoembolectomy of the axillary and brachial artery by Fogarty’s balloon catheter, followed by subclavian artery angioplasty and stenting through the left brachial artery with covered stent (Viabahn/Gore 7 mm x
symptom free with warm hands, good capillary refilling and ulnar day for early pain free mobility. She was later discharged in a transverse process with fibrous bands attached to the first rib. The examination and radiological evidences, therefore we opted for vascular structures causing TOS, as elucidated by history, physical and asymptomatic in 90% of cases [18]. The fact that our patient due to a cervical rib can lead to subclavian artery thrombosis and aneurysms distal to the point of compression by the rib and its attachments [16]. In 1934, Lewis and Pickering stated that trauma with cervical ribs in which 125 had vascular symptoms and 27 had arteriovenous fistulas [15]. Halsted in 1916 had studied a large series of 716 case reports of the thoracocervical region to the axilla coursing between the anterior and middle scalene muscle, through the costoclavicular space and under the pectoralis minor tendon [13]. Its prevalence was found to be 0.74% with higher rate in females as compared in males [14].

Two major types of TOS have been identified-vascular and neurogenic. The vascular type has been further divided into arterial or venous. Neurogenic TOS can be sub categorized into true or disputed. The former is accompanied by objective findings while the disputed form, which is more common, is not. 95% of the patients have a neurogenic form, while 2% have venous symptoms and only 1% has arterial compression [15]. Often patients present with symptoms indicative of more than one type. Common etiological factors producing these variable symptoms include cervical rib, fibrous bands, first thoracic rib and scalenus anticus muscle. Although in many cases no cause is identified [15].

Halsted in 1916 had studied a large series of 716 case reports with cervical ribs in which 125 had vascular symptoms and 27 had aneurysms distal to the point of compression by the rib and its attachments [16]. In 1934, Lewis and Pickering stated that trauma due to a cervical rib can lead to subclavian artery thrombosis and embolization [17]. Cervical rib anomaly is twice as common in women as men. It is present bilaterally in more than 50% of cases and asymptomatic in 90% of cases [18]. The fact that our patient had only the cervical rib as a sole factor compressing the major vascular structures causing TOS, as elucidated by history, physical examination and radiological evidences, therefore we opted for the supraclavicular approach to alleviate the symptoms. Our patient had Gruber Type III cervical rib that extended beyond the transverse process with fibrous bands attached to the first rib. This along with Type IV commonly leads to vascular compromise, including arterial compression, thrombosis and aneurysm formation.

Complete cervical ribs have a 50% incidence of spontaneous onset of symptoms in neurogenic TOS as compared with 20% with incomplete ribs. On the contrary only complete ribs have been noted to produce arterial TOS.

A meticulous history and detailed neurovascular examination will guide to a probable diagnosis and also helps in distinguishing the type of TOS. Stress tests or provocative maneuvers form the basis of physical examination. The most common of these are the Adson’s or scalene maneuvers. Others include East test or "Hands-up" test, costoclavicular maneuver and Allen test. They have very low specificity, sensitivity and predictive value [19]. Direct application of pressure on to the anterior scalene or upper segment of the pectoralis minor elicits focal stress test. A positive response is said if symptoms are reproduced within 15 to 30 s of application of the pressure. Some people use Tinel’s sign, where percussing over the plexus reproduces symptoms.

X-ray cervical spine is done mostly as an initial investigation of choice that may demonstrates a cervical rib. Patients having arterial TOS were more likely to have cervical ribs on chest x-ray than patients with neurogenic or venous TOS [20]. Chest X-ray with apical lordotic views should be obtained to exclude a malignant lesion in the chest. Preoperative duplex ultrasound scanning demonstrates thrombosis or aneurysm of the vascular structure compromised with impediment of blood flow. It sometimes also detects compression missed by provocation test. This can serve as an important portable tool to evaluate the immediate post-operative status. Electromyography/nerve conduction velocity (EMG-Nerve) study is not a routine investigation and is usually considered when a diagnosis of carpal or cubital tunnel syndrome is suspected. MRI scans of the cervical spine and brachial plexus are useful to exclude other possible differentials. CT scan of the area of the brachial plexus and apex of lung may be indicated. Helical 3D CT angiography has an edge over conventional techniques for determining the mechanism and site of compression especially in difficult cases [21].

Conservative treatment with postural control and physiotherapy is an acceptable initial plan of management, unless there is failure of conservative means (for 3-12 months), progressive neurological deficits, refractory pain, and acute or severe initial vascular insufficiency [22]. Arterial TOS has no non-surgical treatment and physical therapy in such an instance does not help. More than a century later Brown and Charlesworth published their data over 18 patients with 23 cervical ribs mentioning that resection of cervical rib was effective in improving their symptoms [24]. Fibrous bands should be removed along with cervical rib especially if they are tethering the plexus.

Prolonged arterial compression by a cervical rib results in subclavian aneurysm formation and thrombosis which necessitates further intervention in addition to cervical rib resection. This can be dealt in the form of Polytetrafluoroethylene
(PTFE) or saphenous vein grafts [25]. On occasions thrombolysis or thrombectomy is considered obligatory in addition to a bypass in order to establish an effective arterial flow [26,27]. Instances where a subclavian artery stent placement is desired, it should be kept in mind that it must be placed after the cervical rib resection in order to secure the stent from being thrombosed from the rib compression [10].

The optimal surgical approach for the excision of rib removal has always been a topic of controversial debate. Popular ones are supraclavicular, transaxillary and infraclavicular but only the transaxillary and supraclavicular routes have coined their names. Earlier studies suggested that the transaxillary approach is appropriate only for first rib resection, and a supraclavicular approach is best when gaining access to the subclavian artery is desired along with cervical rib excision [28,29]. Now the transaxillary approach allows removal of both cervical and the first rib through a single incision site and a supraclavicular approach is indicated to allow access to the subclavian artery for aneurysm repair or to facilitate placement of a bypass graft [10]. The supraclavicular route has minimal risk to neurovascular structures during primary or secondary surgery. It allows the operating surgeon an excellent visualization of the entire brachial plexus, palpation of every structure, extensive micro neurolysis under the operating microscope, and room for appropriate nerve reconstruction, especially in cases of iatrogenic brachial plexus injury, after prior transaxillary first rib resection. A combination of supraclavicular and infraclavicular approach has also been suggested to allow for access to both the subclavian and axillary arteries [4,27]. Microsurgical neurovascular decompression of TOS without removal of the cervical or first rib using a supraclavicular approach has also proven its effectiveness for relief of neurological symptoms [30].

If neurological symptoms dominate than early surgical intervention in the course of the disease prevents degeneration of the brachial plexus and increases the effectiveness of postoperative physiotherapy [31]. Low cure rates have been attributed in certain cases due to longstanding intimal damage to the subclavian artery. This is secondary to a cervical rib fused to the first rib causing immense compression resulting in arterial injury. In these specific circumstances surgical removal of the cervical and first rib was ineffective.

Functional prognosis is the main factor in determining the outcome after surgical decompression. Post-operative management plays a pivotal role in establishing an early functional status of the extremity involved. Appropriate physical therapy for at least 3 months gives the desired practical response. Anticoagulation for a period of 3 to 6 months is to be continued depending upon the extent of vascular procedure [10].

In our case the use of thrombolysis is contraindicated due to recent surgery and the chronicity of the situation. The classic surgical repair was not ideal due to poor run off, and the decision for angioplasty and placing of a covered stent in order to cover the lesion of the subclavian artery and the chronic thrombus was chosen. We did not find any previous literature or case report describing a similar situation, and no trials of angioplasty for an organized arterial thrombosis; although it was extensively reported for cases of venous thrombosis and occlusion of IVC, and or iliac veins with or without the use of thrombolysis.

**Conclusion**

Although no single approach is novel but excision of the cervical rib through supraclavicular approach is not only simple but it also enables precise evaluation of the thoracic inlet and better exposure of the middle and upper trunks of the brachial plexus with release of confounding abnormal anatomical structures especially the cervical rib with minimal risk to the surrounding neurovascular structures. It also offers vascular surgeons the opportunity for conventional vascular repair. Since massive thrombo embolic shower may be amenable for the treatment, hence early diagnosis and management is crucial in TOS with an arterial component. Long term results are better with minimal complications and patients are able to function independently with their daily activities.

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