

Case Report

Cervical Rib Leading to Arterial Thoracic Outlet Syndrome Causing Gangrene and Necessitating Digital Amputation: A Case Report

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Abstract

Thoracic outlet syndrome (TOS) is a syndrome caused by compression of subclavian vessels with/without brachial plexus involvement as it travels through the thoracic outlet. Cervical ribs are congenital anomalies with an incidence of 0.05% to 3%. Usually, they aren't relevant clinically but it may cause TOS due to compression of subclavian vessels and/or brachial plexus by the rib itself or a fibrous band that connects the cervical rib to the first rib. Arterial compression is rare. Here we report a rare case of Thoracic outlet syndrome because of cervical rib leading to gangrenous changes of digits in a 40-year female.

Keywords: TOS, Cervical, rib

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Introduction

Thoracic outlet syndrome can be classified into three subtypes based on the structure involved – this includes

Neurogenic TOS (NTOS)- most common type of TOS

Venous TOS(VTOS)

Arterial TOS (ATOS)- only about 1% of cases are arterial[1-3].

ATOS results from obstruction of the subclavian artery usually caused by cervical or anomalous first rib, scalene muscle, fibromuscular bands or pectoralis minor.[4]with the majority of surgical cases (74%-100%) attributed to abnormal ribs[5,6]. chronic arterial compression may lead to claudication, thrombus formation and possible embolization[6].

According to Edington⁷ cervical ribs were first described by Galen. A well-documented case of bilateral cervical rib was described by Hunauld as an incidental finding in 1742. Since then many authors have described this anomaly manifesting as weakness, peripheral neuritis or vascular disturbances of the upper limb. There are very few cases in the literature where due to supernumerary rib the vascularity became so compromised that gangrene developed and subsequent amputation of the involved part of the limb was required. Because of the rarity of this phenomenon and its devastating consequences, we feel it is justified to add to the literature another

case observed at Post Graduate Institute of Medical Sciences, Rohtak, Haryana. We report a case of 40-year-old female with complain of pain in bilateral upper limb for the past 6 months with gangrenous changes of digits. Surgery could not be performed due to restrictions imposed because of the COVID-19 lockdown. The purpose of this case report was to show that disease symptoms and further progression can be managed conservatively with drugs. Reporting of this case and all investigations were conducted in conformity with the institution's ethical principles of research. Informed consent for participation in the study was obtained from the patient.

Case Report

A 40-year-old female patient, housewife by profession presented to OPD with severe progressive pain in bilateral upper limbs with gangrenous changes in left 2nd to 4th distal phalanges. The patient had a history of pain for the past 2 years for which she was taking analgesics, however, she developed discolouration of digits for the past 6 months for which she was taking medicine as prescribed by a local charlatan. When symptoms didn't subside and pain became unbearable patient presented to OPD. On physical examination here left-sided digits were cold to touch, blood pressure on left-side was non-recordable while on the right side it was 108/76. Radial pulse on the left side was not palpable. Her other peripheral pulses were palpable. Signs of claudication like exercise intolerance and pain at rest were present. All other physical and systemic examinations were normal. Laboratory investigations were also normal. However, chest x-ray of the patient revealed bilateral cervical rib. Bilateral upper limb angiography showed bilateral cervical ribs compressing the 2nd

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part of subclavian artery with mild luminal narrowing. Soft plaque with non-contrast opacification of left brachial artery suggestive of complete occlusion of left brachial artery. Proximal radial artery showed attenuated calibre with complete luminal obstruction of left mid and distal radial artery with reformation of distal most radial artery. left ulnar artery showed diffusely attenuated calibre with non-contrast opacification of distal ulnar artery. The patient was initially started on conservative management for 2 weeks on Cilostazole,

aspirin, ramipril, clopidogrel, atorvastatin, folic acid, levocarnitine and analgesics. Amputation of gangrenous digits and excision of the cervical rib was planned however due to lockdown restriction due to COVID-19 pandemic patient was lost to follow up. However patient showed up 6 months later, with relief in pain and the gangrenous changes didn't progress beyond distal phalanges. Amputation of gangrenous digits was done however since the patient had relief of symptoms; she didn't give consent for cervical rib excision.



Fig 1:Gangrenous changes on dorsal side of distal phalanges of left upper limb
Fig 2:Showing gangrenous changes on palmer side of distal phalanges of left upper limb

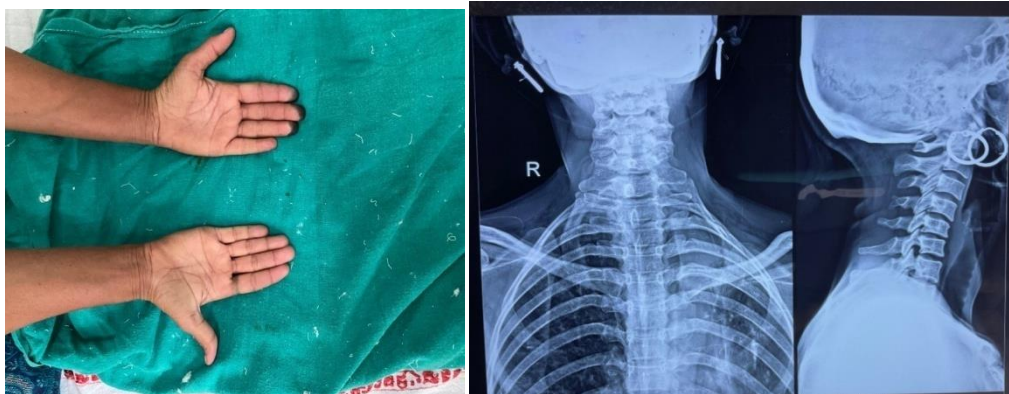


Fig 3:Comparing right and left upper limb with gangrenous changes seen on left digits
Fig 4:Radiograph showing bilateral cervical rib leading to thoracic outlet obstruction

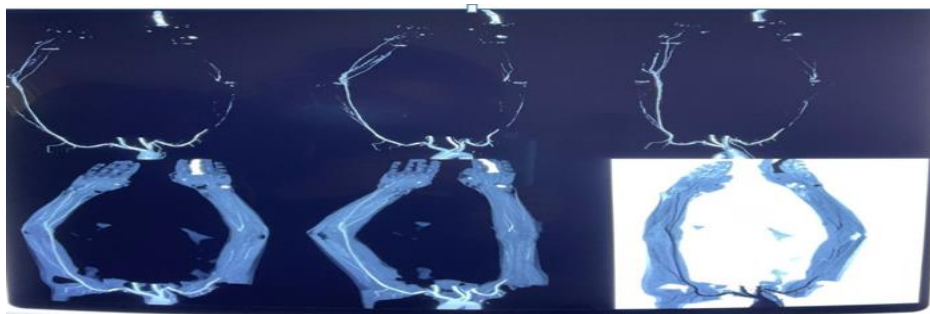


Fig 5:Angiography showing bilateral cervical ribs compressing the 2nd part of subclavian artery with mild luminal narrowing. Soft plaque with non-contrast opacification of left brachial artery suggestive of complete occlusion of left brachial artery. Proximal radial artery showed attenuated calibre with complete luminal obstruction of left mid and distal radial artery with reformation of distal most radial artery. left ulnar artery showed diffusely attenuated calibre with non-contrast opacification of distal ulnar artery

Discussion

A cervical rib is a rib that arises from the 7th cervical vertebrae and is abnormally located above the normal first rib. cervical rib occurs in 1-2% of population and is commonly bilateral, though often asymmetrical[8]. Durham et al. found that 16 (73%) of 22 patients with subclavian artery compression had cervical ribs, and in five (31%) of these, bilateral cervical ribs were present[9]. In addition, soft tissue anomalies, such as scar tissue after neck-shoulder trauma and clavicle trauma, may also predispose an individual to subclavian artery compression[10]. Cervical ribs are recognized as complete or incomplete types[11]. Only complete cervical ribs have been reported to produce vascular symptoms[12,13]. Pain in arm and hand is the most common symptom present in arterial compression which is seen in young otherwise healthy patients. Common findings on physical examination consist of a pulseless, pale and cold distal upper limb. In order not to delay the diagnosis and correct treatment of arterial compression, the clinician must differentiate such arm ischemia from Raynaud syndrome, vasospastic disorders, distal small-artery obstructive disease, or proximally large-artery occlusions which may result in this symptom complex[14]. This disability on rare occasions can have very dire significance this includes a recent case of cervical rib associated with subclavian artery thrombosis and cerebellar and cerebral infarcts. In our case, the patient did have some vascular sequelae and was initially treated with combined anti-coagulant and anti-platelet therapy and followed post amputation for 12 months. The patient responded well to pharmacotherapy with reduced pain and return to activity of daily living. As in our case, pharmacotherapy can be tried initially however when it becomes evident that a serious complication from this anomaly is incipient, then immediate and adequate surgical intervention is necessary.

References

1. Brewin J, Hill M, Ellis H. The prevalence of cervical ribs in a London population. *Clin Anat.* 2009;22:331–6.
2. Viertel VG, Intrapromkul J, Maluf F. Cervical ribs: a common variant overlooked in CT imaging. *AJNR Am J Neuroradiol.* 2012;33:2191–4.
3. Kuhn JE, Lebus VGF, Bible JE. Thoracic outlet syndrome. *J Am Acad Orthop Surg.* 2015;23:222–32.
4. Schon N, Netzsch C, Kroger K. Subclavian vein thrombosis and backpacking. *Clin Res Cardiol.* 2007;96:42–4.
5. Sanders RJ, Hammond SL, Rao NM. Diagnosis of thoracic outlet syndrome. *J Vasc Surg.* 2007;46:601–4.
6. Reeser JC. Diagnosis and management of vascular injuries in the shoulder girdle of the overhead athlete. *Curr Sports Med Rep.* 2007;6:322–7.
7. Edington GH. Cervical Ribs. *Glasgow M J.* 1932;118:298-313.
8. Verschakelen JA. The chest wall, pleura and diaphragm. In: Grainger RG, Allison D, Adam A, Dixon AK eds. *Grainger & Allison's Diagnostic Radiology: A Textbook of Medical Imaging.* 4th edition. London: Churchill Livingstone, 2001, 320p.
9. Durham JR, Yao JS, Pearce WH, Nuber GM, McCarthy III WJ. Arterial injuries in the thoracic outlet syndrome. *J Vasc Surg.* 1995;21:57-70.
10. Davidovic LB, Kostic DM, Jakovljevic NS, Kuzmanovic IL, Simic TM. Vascular thoracic outlet syndrome. *World J Surg.* 2003;27:545-50.
11. Adson AW, Coffey JR. Cervical rib: a method of anterior approach for relief of symptoms by division of the scalenus anterior. *Ann Surg.* 1927;85:839.
12. Sanders RJ, Hammond SL. Management of cervical ribs and anomalous first ribs causing neurogenic thoracic outlet syndrome. *J Vasc Surg.* 2002;36:51-6.
13. Short DW. The subclavian artery in 16 patients with complete cervical ribs. *The J Cardiovas Surg.* 1975;16:135-41.
14. Jusufovic M, Sandset EC, Popperud TH, Solberg S, Ringstad G, Kerty E. An unusual case of the syndrome of cervical rib with subclavian artery thrombosis and cerebellar and cerebral infarctions. *BMC Neurol.* 2012;12:48.

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