Thoracic Outlet Syndrome Caused by Synostosis of the First and Second Thoracic Ribs: 2 Case Reports and Review of the Literature

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We present 2 cases of combined arterial and neurogenic thoracic outlet syndrome triggered by trauma in patients with congenital synostoses of the first and second ribs. These patients were successfully treated with supraclavicular resection of the first and second ribs and scalenectomy. We review these cases and the associated literature on thoracic outlet syndrome and rib synostosis. *(J Hand Surg Am. 2014;39(12):2444–2447. Copyright © 2014 by the American Society for Surgery of the Hand. All rights reserved.)*

Key words Brachial plexus, rib fusion, rib synostosis, subclavian, thoracic outlet syndrome.

THORACIC OUTLET SYNDROME (TOS) is a heterogeneous group of disorders in which neurovascular structures are compressed within the subclavicular space. It is estimated to occur in 0.3% to 2% of the general population.[1] The most common type of TOS is the disputed neurogenic type, in which patients present with upper extremity numbness, pain, paresthesias, and positional exacerbation but demonstrate no objective neurological or vascular findings (up to 97% of patients).[1,2] True neurogenic TOS presents with similar symptoms and objective evidence of brachial plexus compression such as hypothenar atrophy or positive electrodiagnostic testing (fewer than 1% of patients).[1,2] In vascular TOS, compression of the subclavian vessels will present with venous symptoms such as arm swelling and cyanosis (2% to 3%) or arterial symptoms such as ischemic claudication and exertional fatigue (1% to 2%).[1,3] In a distinct minority of cases, bony anomalies are the primary cause of TOS.[4] We present 2 cases in which traumatic events in previously asymptomatic individuals with congenital synostoses of the first and second ribs led to combined arterial and neurogenic TOS.

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Received for publication April 23, 2014; accepted in revised form August 28, 2014.

No benefits in any form have been received or will be received related directly or indirectly to the subject of this article.

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0363-5023/14/3912-0014$36.00/0
http://dx.doi.org/10.1016/j.jhsa.2014.08.034

CASE 1

A 22-year-old dance student presented with numbness and tingling in the left upper extremity for 2.5 years. The symptoms began after a low-speed motor vehicle accident in which she sustained a left shoulder contusion. She initially underwent therapy and shoulder arthroscopy for shoulder instability without symptomatic improvement over one year. At presentation, she had incapacitating numbness and color changes in the hand elicited by placing the shoulder in an abduction–external rotation position.

On examination, there was full strength in all muscles and intact sensation in the C5–T1 dermatomes. Adson and Roos tests were positive, with loss of radial pulse and a sensation of numbness in the entire hand.

Apical-lordotic chest radiographs identified no cervical ribs but the left second rib was marginally elevated (Fig. 1). Magnetic resonance imaging (MRI) of the brachial plexus indicated synostosis of the first and second ribs (Fig. 2). During shoulder abduction and external rotation, the MRI revealed reduced costoclavicular space with neurovascular compression and

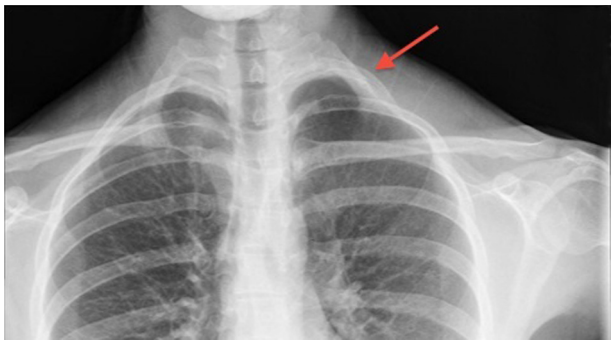

FIGURE 1: Apical-lordotic chest radiograph indicating asymmetry of the left first and second thoracic ribs and elevation of the second rib (arrow).

signal hyperintensity of the brachial plexus divisions and cords (Fig. 3).[5] Contrast-enhanced magnetic resonance angiography with the shoulder abducted and externally rotated demonstrated a segment of subclavian artery occlusion with complete reconstitution distally. Electrodiagnostic testing performed one year after symptom onset was normal.

Thoracic outlet decompression was performed via a supraclavicular approach. The first to second rib synostosis was identified (Fig. 4) with the first rib noted to be abnormally prominent anterosuperiorly. The subclavian artery was draped across the first rib, forming an acute angle as it entered the subpectoral space. The costoclavicular space was almost completely obliterated when the shoulder was abducted. The synostosis was removed via first and second rib osteotomies. The patient reported marked resolution of symptoms by the second postoperative week and returned to dance training without restriction.

CASE 2

A 17-year-old avid athlete presented with progressively increasing numbness in the right arm and hand over a 6-month period after a shoulder injury while snowboarding. He failed to improve with therapy for 2 months and the progressive symptoms precluded a return to sports. On examination, the intrinsic hand muscles were weak (Medical Research Council grade 4) but there was no wasting or clawing. Adson test was positive, resulting in a loss of radial pulse and a sensation of numbness in the shoulder, upper arm, and hand.

Radiographs indicated a subtle asymmetry in the upper thoracic ribs with first rib elevation. Magnetic resonance imaging demonstrated a rudimentary first rib fused to the second rib, resulting in narrowing of the costoclavicular space. There was subclavian vessel impingement and signal hyperintensity of the cords and divisions of the brachial plexus. A computed

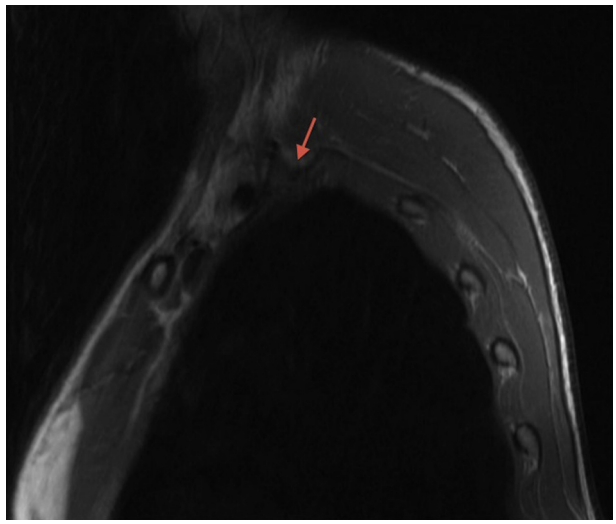

FIGURE 2: Sagittal MRI section indicating synostosis between the first and second thoracic ribs (arrow).

tomography scan with 3-dimensional reconstruction better delineated the patho-anatomy (Fig. 5). Electromyography indicated a chronic, incomplete denervation pattern of the intrinsic muscles, consistent with lower trunk (C8–T1) involvement.

At the time of decompression through a supraclavicular approach, an abnormal fascial sling elevated and kinked the subclavian artery. This was decompressed and the synostosis was removed. The patient reported complete resolution of symptoms within 2 weeks. He recovered without complication and returned to athletic and other recreational pursuits without restriction.

DISCUSSION

Thoracic outlet syndrome is commonly classified as venous TOS, arterial TOS, and disputed or true neurogenic TOS; true neurogenic TOS accounts for fewer than 1% of cases.[1,2] Compression can take place in the interscalene, costoclavicular, or subpectoral minor spaces.[1] Causes of compression include congenital fibrous bands, posttraumatic muscular hypertrophy or scarring, posture problems, tumors, and bony anomalies. Although the prevalence of anomalous first ribs approximates that of cervical ribs (0.2% to 0.5%), anomalous first ribs are less commonly associated with TOS.[6,7] Weber and Criado[4] recently reported that about one-third of 400 TOS cases involved a bony anomaly, with 20% involving a cervical rib and 3% involving an isolated first rib anomaly. However, they did not identify the specific first rib anomalies observed.

Sporadic cases of TOS caused by synostosis of the first and second ribs have been reported.[8–14] The

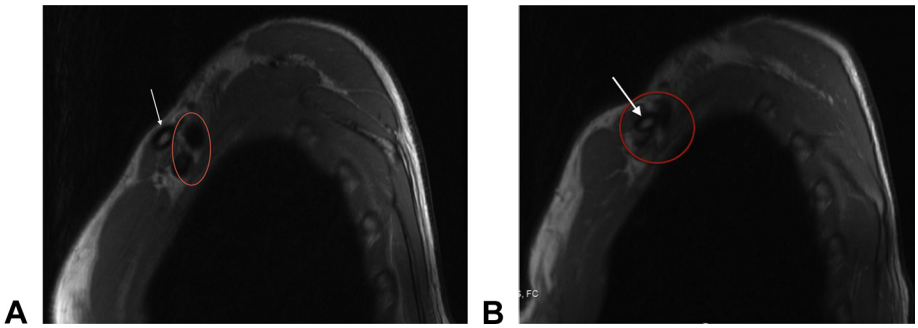

FIGURE 3: A Magnetic resonance image with shoulder in neutral position indicates adequate space for the subclavian vessels (oval) behind the clavicle (arrow). **B** When the arm is abducted, the costoclavicular space (circle) is significantly diminished with evident compression of the vessels.

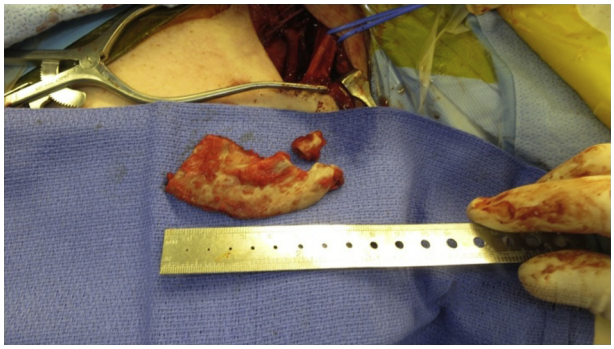

FIGURE 4: Resection specimen demonstrating an abnormal synostosis between the first and second ribs. The smaller bone fragment was the part of the first rib that was initially removed using a bone rongeur.

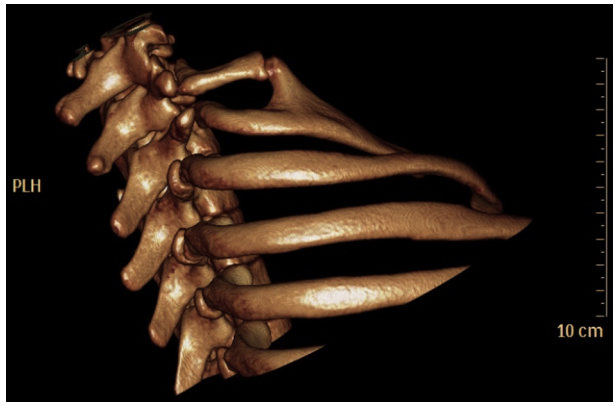

FIGURE 5: Computed tomography scan with 3-dimensional reconstruction showing the synostosis between the first and second ribs.

structural pathology was a hypoplastic first rib positioned slightly higher than usual and inserting into the anterior margin of the second rib. The scalenus anticus occasionally inserted into the fusion site, resulting in a bony exostosis that compressed adjacent neurovascular structures.[11,14] Whereas previous reports describe distinctly arterial or neurogenic findings, the 2 cases presented here demonstrated combined arterial and neurogenic compression caused by the fused ribs.

Nonspecific upper extremity numbness and pain after trauma to the shoulder girdle should raise the possibility of TOS in a previously asymptomatic individual. It is likely that neurovascular compression is exacerbated in patients with a structural predisposition, possibly owing to posttraumatic fibrosis, disuse-induced muscular imbalance, or altered joint kinematics. This phenomenon was observed in a series of 7 neurogenic TOS cases involving first rib anomalies, in which 5 patients became symptomatic after automobile accidents.[14]

It is essential to rule out additional causes of TOS even when an obvious source is identified. In our second case, there was a fascial sling kinking the subclavian artery in addition to the thoracic rib synostosis. Furthermore, the subclavian vessels can be compressed at multiple sites.[8,15] First thoracic rib anomalies are bilateral in about 5% of cases, and contralateral symptoms should be ruled out.[11,12] Concomitant sites of compression can be thoroughly evaluated with a combination of computed tomography (with 3-dimensional reconstruction), MRI, and angiography. Magnetic resonance imaging has a 79% sensitivity and 88% specificity for determining brachial plexus compression and may be useful in identifying hypertrophied muscles and fibrous bands.[16] Dynamic MRI with the arm in various provocative positions is useful; symptomatic patients demonstrate narrowing of the minimum costoclavicular distances (about 10 mm) compared with neutral position (about 25 mm).[17]

Upper extremity surgeons should be aware that arterial compression poses an increased risk of aneurysm, thrombus, and embolus, which can result in occlusion of the cerebral or upper extremity circulation.[6,9,10] In addition to distal occlusions in the

upper limb, compression of the subclavian artery by anomalous first ribs and cervical ribs may cause thromboembolic occlusions of the vertebrobasilar and carotid arteries via retrograde arterial propagation.[9] Therefore, such cases should be thoroughly evaluated to rule out evolving arterial compromise, preferably in conjunction with a vascular surgeon and advanced vascular imaging studies.[14] The presence of vascular involvement directly affects management strategy. Neurogenic TOS without electrodiagnostic findings is usually treated conservatively, with a recommended 3 to 12 months of therapy directed at stretching and posture correction. If this fails and symptoms are disabling, surgery may be indicated.[14] In contrast, arterial TOS might require earlier intervention because of the risks of embolization, occlusion, and aneurysm. Surgical indications may include dilatation of the subclavian artery, intimal injury, or mural thrombus.[14,18] In the current cases of combined arterial and neurogenic TOS, earlier operative intervention was indicated for the arterial TOS, which treated the neurogenic component as well.

We elected to use the supraclavicular approach to obtain optimal exposure of the scalene triangle and brachial plexus. The transaxillary approach allows good visualization of the first rib and is ideal for treating venous TOS (owing to good exposure of the subclavius muscle) and may also provide a superior aesthetic result.[3] It does not, however, permit ideal visualization of the subclavian artery or the plexus.[3] In contrast, the supraclavicular approach allows for excellent exposure of the subclavian artery and brachial plexus and permits a more thorough anterior and middle scalenectomy.[1,3,14] Complications have been reported from either approach, including hematoma requiring evacuation, pneumothorax, subclavian artery laceration, and transient phrenic nerve injuries.[4,11]

REFERENCES

1. Meyer R, Jones KJ. Thoracic outlet compression syndrome. In: Wolfe SW, Hotchkiss RN, Pederson WC, Kozin SH, eds. *Green's Operative Hand Surgery*. 6th ed. Philadelphia, PA: Churchill Livingstone; 2011:1015–1034.
2. Sanders RJ, Hammond SL, Rao NM. Diagnosis of thoracic outlet syndrome. *J Vasc Surg*. 2007;46(3):601–604.
3. Fugate MW, Rotellini-Coltvet L, Freischlag JA. Current management of thoracic outlet syndrome. *Curr Treat Options Cardiovasc Med*. 2009;11(2):176–183.
4. Weber AE, Criado E. Relevance of bone anomalies in patients with thoracic outlet syndrome. *Ann Vasc Surg*. 2014. http://dx.doi.org/10.1016/j.avsg.2013.08.014.
5. Demondion X, Herbinet P, Van Sint Jan S, Boutry N, Chantelot C, Cotten A. Imaging assessment of thoracic outlet syndrome. *Radiographics*. 2006;26(6):1735–1750.
6. Haug CE, Sanders RJ, Arterial TOS. In: Sanders RJ, ed. *Thoracic Outlet Syndrome: A Common Sequela of Neck Injuries*. Philadelphia, PA: JB Lippincott Co; 1991:211–231.
7. Etter L. Osseous abnormalities in the thoracic cage seen in forty thousand consecutive chest photoroentgenograms. *Am J Roentgenol Radium Ther*. 1944;51:359–363.
8. Khashram M, Dharmaraj RB, Ramanathan A, Buckenham T. Medical image: unusual case of thoracic outlet syndrome. *N Z Med J*. 2011;124(1330):78–80.
9. Yamaguchi R, Kohga H, Kurosaki M, et al. Acute basilar artery occlusion in a patient with left subclavian artery occlusion due to first rib anomaly: case report. *Neurol Med Chir (Tokyo)*. 2008;48(8):355–358.
10. McSweeney SE, Al-Kelani R, Harte P, Brady A. Thoracic outlet syndrome secondary to first rib anomaly: the value of multi-slice CT in diagnosis and surgical planning. *Ir Med J*. 2005;98(8):246–247.
11. Nguyen T, Baumgartner F, Nelems B. Bilateral rudimentary first ribs as a cause of thoracic outlet syndrome. *J Natl Med Assoc*. 1997;89(1):69–73.
12. Baumgartner F, Nelson RJ, Robertson JM. The rudimentary first rib: a cause of thoracic outlet syndrome with arterial compromise. *Arch Surg*. 1989;124(9):1090–1092.
13. Siegel RS, Steichen FM. Cervicothoracic outlet syndrome: vascular compression caused by congenital abnormality of thoracic ribs. A case report. *J Bone Joint Surg Am*. 1967;49(6):1187–1192.
14. Sanders RJ, Hammond SL. Management of cervical ribs and anomalous first ribs causing neurogenic thoracic outlet syndrome. *J Vasc Surg*. 2002;36(1):51–56.
15. Edwards PR, Moody AP, Harris PL. First rib abnormalities in association with cervical ribs: a cause for postoperative failure in the thoracic outlet syndrome. *Eur J Vasc Surg*. 1992;6(6):677–681.
16. Panegyres PK, Moore N, Gibson R, Rushworth G, Donaghy M. Thoracic outlet syndromes and magnetic resonance imaging. *Brain*. 1993;116(pt 4):823–841.
17. Demirbag D, Unlu E, Ozdemir F, et al. The relationship between magnetic resonance imaging findings and postural maneuver and physical examination tests in patients with thoracic outlet syndrome: results of a double-blind, controlled study. *Arch Phys Med Rehabil*. 2007;88(7):844–851.
18. Sanders RJ, Haug C. Review of arterial thoracic outlet syndrome with a report of five new instances. *Surg Gynecol Obstet*. 1991;173(5):415–425.