
Neuroimaging Highlight

Editors: Richard Farb, David Pelz

Clavicle Pseudarthrosis: A Rare Cause of Thoracic Outlet Syndrome

Submitted by: Kathleen Joy Khu, Rajiv Midha

Can. J. Neurol. Sci. 2010; 37: 863-865

CASE REPORT AND DISCUSSION

A 22-year-old man presented with a one year history of right-sided shoulder pain, hand weakness and tingling, and purplish discoloration of the upper extremity upon abduction. He had congenital pseudarthrosis of the right clavicle since childhood. Aside from an obvious deformity characterized by asymmetry of the shoulders and a palpable bony depression over the right clavicle, the patient had been previously asymptomatic. Physical examination revealed the musculoskeletal deformities as described, as well as prominent veins over his right shoulder, arm, and chest. With arm elevation, his right arm became dusky and his radial pulse diminished. Neurologically, the patient had no deficits except for mild weakness (Grade 4+/5) of the ulnar-innervated intrinsic muscles of the right hand. The clinical findings were consistent with a combined neurogenic and vascular form of thoracic outlet syndrome.

Electrodiagnostic studies revealed a marked decrease in the right medial antebrachial cutaneous response, indicative of medial cord involvement. On electromyogram testing, there were signs of chronic neurogenic change in the T1-innervated abductor pollicis brevis muscle on the right.

X-ray of the right clavicle showed a deformity in the midshaft with evidence of pseudoarticulation at the ends of the medial and lateral segments. The lateral segment was subluxed superiorly and demonstrated a serpentine course (Figure 1A). Computed tomogram of the thorax demonstrated that the right clavicle was divided into three discrete fragments articulating via a pseudarthrosis at each end. These formed an abnormal U-shaped structure directed inferiorly and posteriorly (Figure 1B), exerting mass effect on the underlying neurovascular structures and narrowing of the subclavian vein (Figure 1C). Dynamic magnetic resonance venography revealed delayed filling and narrowing of the right subclavian vein (Figure 1D) with the patient's arms elevated, while magnetic resonance angiography demonstrated a focal hourglass-shaped narrowing of the right subclavian artery (Figure 1E).

The patient underwent brachial plexus decompression and resection of his clavicle pseudarthrosis via an anterior supra-clavicular approach. The brachial plexus elements and subclavian artery were found to be compressed by the U-shaped lateral segment of the pseudarthrosis that was projecting posteriorly (Figure 2A). The neurovascular structures were dissected and protected prior to resection of the bony abnormality (Figure 2B). No clavicular reconstruction was performed. Immediately post-operatively, the patient experienced improvement in his symptoms, and his shoulder range of movement remained full. At his two-month follow-up visit, the veins in his arm and chest had become less prominent, and his radial pulse remained full on abduction. The paresthesia in his fingers had improved significantly, but the mild weakness in his hand intrinsics remained unchanged.

Congenital pseudarthrosis of the clavicle is rare, with around 200 cases reported in the literature.¹ It occurs on the right side in 90% of patients and bilaterally in 10%.² It frequently presents as a painless mass or swelling over the clavicle and is usually associated with normal function and full range of motion of the shoulder girdle and upper limb.¹ Etiology is unknown, but it is presumed to be due to failure of fusion of ossification centers or pressure of the subclavian artery on the developing clavicle.³

There have been a few reports of thoracic outlet syndrome secondary to pseudarthrosis of the clavicle.³⁻⁹ Most of them^{4-6,8,9} presented with symptoms of vascular thoracic outlet syndrome. Young et al³ described a similar case, with the patient exhibiting both neurologic and vascular symptoms. In our patient, the presence of the pseudarthrosis resulted in dynamic symptomatic compression of the brachial plexus, subclavian artery, and subclavian vein.

Treatment involves surgical resection of the pseudarthrosis and decompression of the neurovascular structures. Some authors reconstruct the clavicular defect with bone grafts,³ while others advocate a simple resection without reconstruction since this would not result in any functional handicap.^{4,8,9}

From the Division of Neurosurgery, Department of Clinical Neurosciences and Hotchkiss Brain Institute, University of Calgary, Calgary, Alberta, Canada.

RECEIVED MARCH 22, 2010. FINAL REVISIONS SUBMITTED MAY 3, 2010.

Correspondence to: Rajiv Midha, Division of Neurosurgery, Department of Clinical Neurosciences, University of Calgary, Room 1195, 1403 - 29 Street NW, Calgary, Alberta, T2N 2T9, Canada.

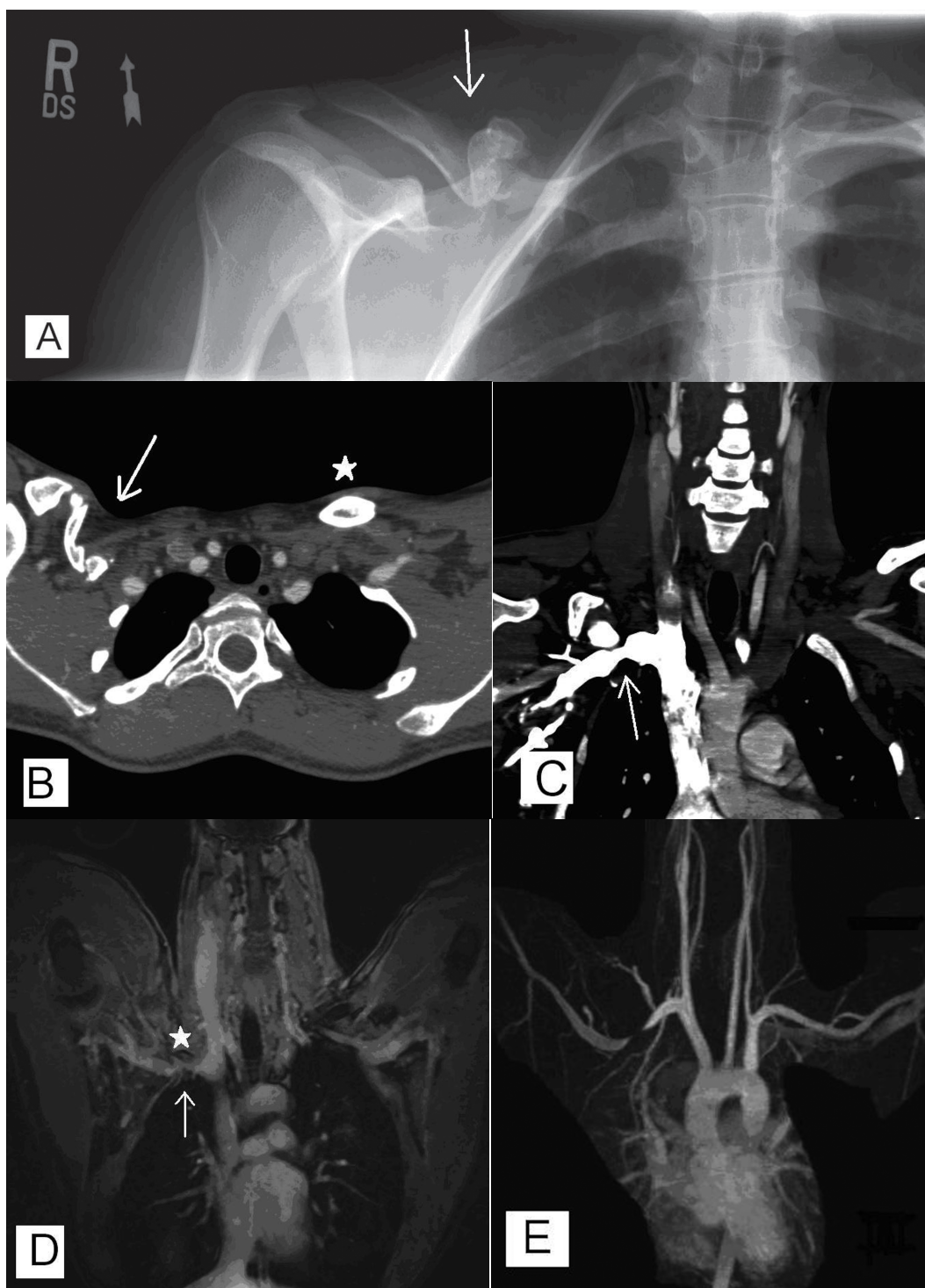


Figure 1: (A) X-ray of the right clavicle shows a deformity in the midshaft (arrow) with evidence of pseudoarticulation at each end. The lateral segment is subluxed superiorly and demonstrates a serpentine course. (B) Axial CT with contrast of the thorax shows the U-shaped pseudoarthrosis of the right clavicle (arrow) displaced posteriorly and compressing the underlying structures. The usual anterior position of the contralateral normal clavicle (star) is visualized for comparison. (C) Coronal sequences of CT angiography of the neck and thorax shows narrowing of the right subclavian vein (arrow). Superior to the vein are two fragments of the pseudoarthrosis showing the pseudoarticulation between the two. (D) Coronal sequences of magnetic resonance venography of the neck and thorax with patient's arms elevated demonstrate the pseudoarthrosis (star) causing compression and narrowing of the right subclavian vein. (E) Coronal sequences of magnetic resonance angiography of the neck and thorax with patient's arms elevated show an hourglass-shaped narrowing of the right subclavian artery.

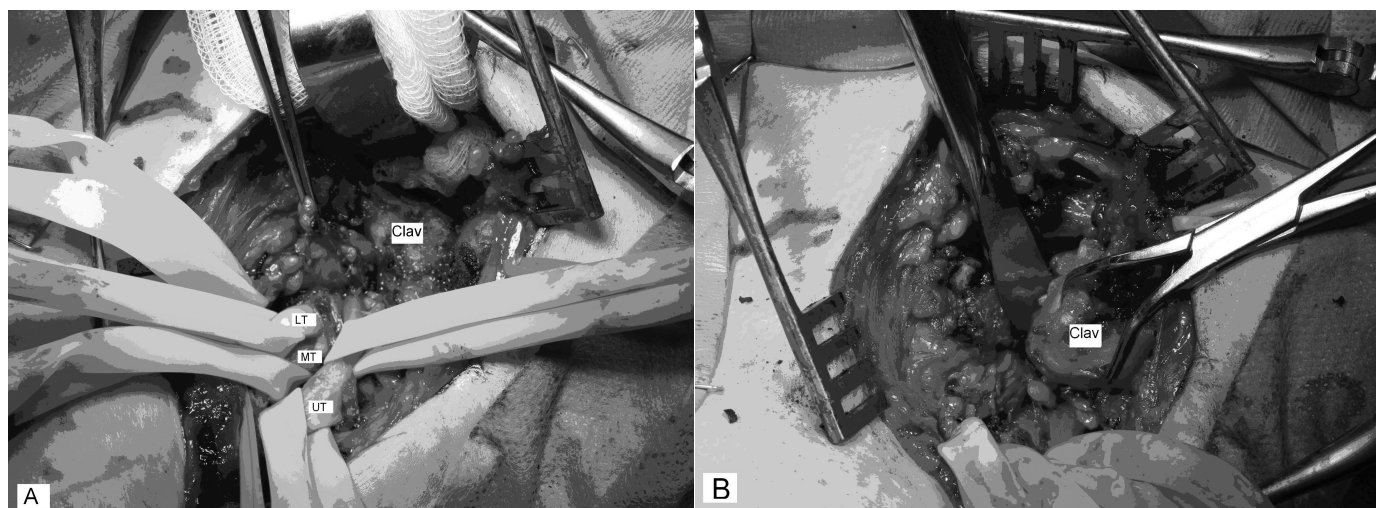


Figure 2: Intraoperative photographs of the patient's right supraclavicular area. The upper part of the image is the caudal end while the bottom is the cranial end of the patient. (A) The brachial plexus elements have been dissected and isolated using Penrose drains prior to resection of the pseudarthrosis. The trunks are visualized just cranial to the pseudarthrosis (Clav). Legend: UT = upper trunk, MT = middle trunk, LT = lower trunk. (B) The U-shaped segment of the pseudarthrosis (Clav) is being gently dissected off the underlying fibrous tissue. At this point, the osseocartilaginous structures connecting it to the medial and lateral ends of the clavicle have already been resected to decompress the neurovascular structures.

REFERENCES

1. Cadilhac C, Fenoll B, Peretti A, et al. Congenital pseudarthrosis of the clavicle: 25 childhood cases. *Rev Chir Orthop Reparatrice Appar Mot.* 2000;86(6):575-80.
2. Sloan A, Paton R. Congenital pseudarthrosis of the clavicle: the role of CT-scanning. *Acta Orthop Belg.* 2006;72(3):356-8.
3. Young MC, Richards RR, Hudson AR. Thoracic outlet syndrome with congenital pseudarthrosis of the clavicle: treatment by brachial plexus decompression, plate fixation and bone grafting. *Can J Surg.* 1988;31(2):131-3.
4. Bargar WL, Marcus RE, Ittleman FP. Late thoracic outlet syndrome secondary to pseudarthrosis of the clavicle. *J Trauma.* 1984;24(9):857-9.
5. Hahn K, Shah R, Shalev Y, et al. Congenital clavicular pseudarthrosis associated with vascular thoracic outlet syndrome: case presentation and review of the literature. *Cathet Cardiovasc Diagn.* 1995;35(4):321-7.
6. Lozano P, Doaz M, Riera R, et al. Venous thoracic outlet syndrome secondary to congenital pseudarthrosis of the clavicle. Presentation in the fourth decade of life. *Eur J Vasc Endovasc Surg.* 2003;25(6):592-3.
7. Sales de Gauzy J, Baunin C, Puget C, et al. Congenital pseudarthrosis of the clavicle and thoracic outlet syndrome in adolescence. *J Pediatr Orthop B.* 1999;8(4):299-301.
8. Valette H. Congenital pseudarthrosis of the clavicle and thoracic outlet syndrome. Report of a case and review of the literature. *J Mal Vasc.* 1995;20(1):51-2.
9. Garnier D, Chevalier J, Ducasse E. Arterial complications of thoracic outlet syndrome and pseudarthrosis of the clavicle: three patients. *J Mal Vasc.* 2003;28(2):79-84.