

THE CHONDROEPITROCHLEARIS MUSCLE: A RARE CAUSE OF AXILLARY VEIN THROMBOSIS AND LYMPHEDEMA

C. Thomet, J-P. Belgrado, S. Vankerckove, S. Grijseels,
S. Heijmans, S. De Smet, L. Vandermeeren

Departments of Plastic, Reconstructive and Aesthetic Surgery (CT,SDS,LV), Radiology & Echography (SG), Université Libre de Bruxelles; Lymphology Clinic of Brussels, (J-PB,SV,LV) and Lymphology Research Unit (J-PB,SV,LV), Academic Medical Center, St. Pierre, Belgium.

ABSTRACT

The chondroepitrochlearis muscle is a rare anomaly of the pectoralis major crossing over the neurovascular bundle in the axilla. Often associated with other supernumerary muscles like the arch of Langer, it has been reported in the past as a cause of restriction of arm abduction, cosmetic defects and compression of the ulnary nerve.

This case report describes the first known vascular complication due to a chondroepitrochlearis muscle, causing intermittent compression of the axillary vein and thrombosis, associated with pain, upper limb lymphedema, and impaired movements. The diagnosis was suspected from the medical history and confirmed by palpation and dynamic ultrasonography. Surgery was performed to divide the muscle slip with the help of lymphofluoroscopy to prevent damage to the lymphatic structures. Pain and impairment of movement disappeared within a few days after surgery and lymphedema decreased significantly.

Keywords: chondroepitrochlearis muscle, lymphedema, axillary vein, thrombosis, lymphofluoroscopy

The most common anatomical variation

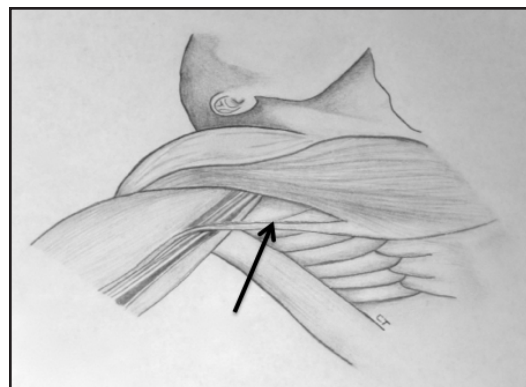


Fig. 1. The chondroepitrochlearis muscle (arrow) arises from the pectoralis major, crosses the axilla, and inserts on the humerus.

found in the axilla is called the arch of Langer, a tendinous slip arising from the anterior border of the latissimus dorsi and joining the tendon of the pectoralis muscle. Its incidence is 7%. On the other hand, the chondroepitrochlearis muscle is much rarer, it arises from the pectoralis major, crosses the axilla, and inserts on the humerus (Fig. 1). Clinical expression of the potential presence of the chondroepitrochlearis muscle has been described as shoulder contracture, ulnar nerve entrapment, and cosmetic issues.

In this paper, we describe what is likely the first known vascular complication caused



Fig. 2. Early family picture of the subject as a child showed the presence of right forearm lymphedema and telangiectasia (arrow).

by a chondroepitrochlearis muscle expressed through thrombosis and intermittent compression of the neurovascular bundle including the axillary vein, lymphatic pedicle, and nerves. For the patient, it resulted in pain, upper limb lymphedema, and an impaired range of motion of the shoulder.

CASE REPORT

A 41 year old woman, right-handed hairdresser, consulted at the Lymphology Clinic of Brussels with chronic swelling of the right upper limb associated with coldness of the hand, itching, pain on exertion, and shoulder abduction restriction. The patient was diagnosed at birth with a venous malformation of the upper limb called hemangioma. She reports that she was born with a swollen and bluish limb and that she was treated with cryotherapy during infancy. Old family pictures, showed the early presence of the right forearm lymphedema and telangiectasia (*Fig. 2*).

Medical history describes epilepsy since the age of 27 and secondary hypothyroidism after thyroidectomy for cold nodules. Later

in adulthood, pain exacerbated during movements like writing, painting, ironing, exercising, and also at night disturbing sleep. She also suffered from a cold hand, itching, and paresthesia. As a hairdresser, the working position – arm in abduction – was painful, forced her to stop frequently and resulted in a swelling of her hand and forearm at the end of the day. However, during rest periods, all symptoms decreased.

At age 38, symptoms suddenly worsened after lifting a barbell at the gym, and she consulted another hospital. On physical examination, she presented bluish telangiectasia of the right upper limb and lymphedema which was more evident at the forearm with a 1.5 centimeter difference of wrist circumferences. Echo-Doppler of the upper limb showed no expansive process in the subcutaneous tissues. Deep veins of the forearm and arm presented no abnormalities; however, the axillary vein was almost invisible and the presence of a superficial network was noted. The radiologist suggested an obstacle in the axillary region evoking a chronic thrombosis with partial secondary fibrosis of the principal axis and development of a collateral network. The patient was referred to a vascular malformations center for further advice and investigation. There, based on her medical history and physical findings, the diagnosis of venous malformation was suspected again but after performing an MRI, it was ruled out. This time, an abnormal thickening of the subcutaneous fat was documented. Blood tests were ordered to rule out a localized intravascular coagulation and thrombophilia, and all returned negative. No definitive diagnosis was made. The pain was attributed to a post-thrombotic syndrome and a grade I compression garment was prescribed. The patient was also encouraged to restart working out.

Because of persistence of symptoms, the patient was referred to an internal medicine department where the hypothesis of a Klippel-Trenaunay syndrome was investigated. After an angio-MRI, no vascular



Fig. 3. The diagnosis was confirmed using bilateral dynamic ultrasound and duplex which demonstrated compression of the right axillary vein during voluntary muscle contraction of the shoulder. In the examination, combined movements of adduction and protraction are shown.

malformation was found: however, the absence of the right axillary vein portion (although present in the left side) was highlighted. Chronic thrombosis of the right axillary vein was diagnosed again and the physician started a low molecular weight heparin treatment in order to develop collaterals and decrease the venous insufficiency. The symptoms decreased slightly but not significantly. Therefore, an arterial echo-Doppler was ordered to rule out a thoracic outlet syndrome. It showed good patency of the arterial axis with no stenosis during Adson's maneuver. A phlebography of the right upper limb was performed showing no residual thrombosis. Anticoagulant treatment, started 12 months earlier, was then discontinued.

Almost 4 years after the initial worsening of her symptoms, the patient was referred to our Lymphology Clinic. She continued to present the same symptoms. Physical findings

showed swelling of the arm with a positive pitting test but no pathognomonic signs of primary lymphedema. The telangiectasias already described were clearly visible. On closer examination of her axilla, an abnormal structure on the lateral border of the pectoral muscle was palpable. This filiform structure crossed the axilla in the direction of the humerus. That finding pleaded in favor of a supernumerary muscle. We first thought of an arc of Langer but the course of the structure was not in accordance with Langer's description. Reviewing specific anatomical literature led us to conclude on the presence of a chondroepitrochlearis muscle.

The diagnosis was confirmed by a bilateral dynamic ultrasound and duplex showing the compression of the right axillary vein during shoulder adduction and protraction (*Fig. 3*).

A lymphofluoroscopy was performed in order to visualize the architecture and the

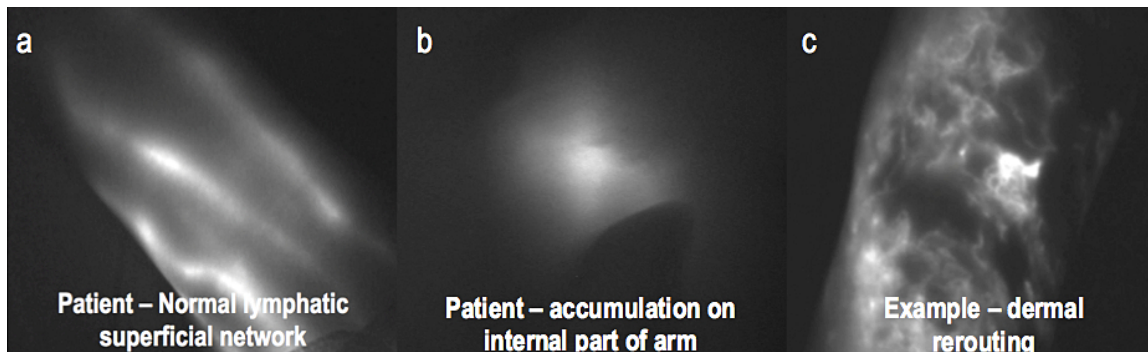


Fig. 4. Lymphofluoroscopy after an intradermal injection in the 1st interdigital web space showed a normal and functional superficial lymphatic network (a) in most of the arm with only one small abnormal area of dense and diffuse fluorescence found located in the internal part of the arm (b), just below where the chondroepitrochlearis muscle was observed. For comparison to 4a, a typical abnormal image of the superficial lymphatic system (c) is seen in patients with lymphatic insufficiency demonstrating an enlarged initial lymphatic network due to the dermal rerouting.

functionality of the superficial lymphatic network. This dynamic exam showed a normal and functional superficial lymphatic network all over the arm (*Fig. 4a*) with only a small area of dense fluorescence observed in the internal part of the arm, just below the chondroepitrochlearis muscle (*Fig. 4b*).

Surgery was planned under lymphofluoroscopy control in order to prevent damage to superficial lymphatic collectors. A 1.5 centimeters incision was performed over the abnormal muscle and showed a tendinous slip of the chondroepitrochlearis muscle above the axillary vein (*Fig. 5*). The slip was divided to release the compression.

Swelling and other symptoms including pain, cold hand, and impaired movements decreased significantly immediately after surgery. A mild consistent edema continued, which was treated and resolved combining exercises under multicomponent bandages and fluoroscopy-guided manual lymph drainage.

COMMENTARY

The chondroepitrochlearis is a very rare muscular variation and has been described with the coexistence of other muscular abnormalities in the axilla (1). This muscular slip

may arise from the pectoralis major muscle, the costal cartilages, or the aponeurosis of the external oblique muscle. It then courses across the axilla and inserts into the medial humeral epicondyle or the medial brachial intermuscular septum. It is considered as a weak adductor of the arm at the shoulder joint and contracts on forcible arm adduction and arm flexion. Its innervation originates from the medial pectoral nerve. Bergman et al in their book on anatomical variation named the muscle “costoepitrochlearis”, “chostoepitrochlearis”, “CET” or “chondrohumeralis” (2). Recently, Loukas et al suggested a revision of the current nomenclature and proposed the term of thoracoepicondylaris, which is more accurate according to them (3).

The chondroepitrochlearis has been associated with another aberrant muscle in the axilla called the axillary arch of Langer. The abnormal bundle originates from the upper edge of the latissimus dorsi, arches across the axilla in front of the vessels and nerves and inserts into the tendon of the pectoralis major and sometimes adheres either to the coracobrachialis muscle, biceps brachii muscle or long head of triceps brachii muscle, teres major muscle, coracoid process of scapula, or medial epicondyle of the



Fig. 5. Treatment first identified the tendinous slip of the chondroepitrochlearis muscle (arrow) which was divided to release the compression.

humerus (4). Chiba et al suggested that the chondroepitrochlearis is almost always associated with the arch of Langer, which itself occurs in 7 to 13% of the population (5) but other studies proved this assertion incorrect (6). Bergman et al suggested that the presence of the chondroepitrochlearis muscle is always associated with the absence of the normal twist of the pectoralis major tendon at its insertion (7). In mammals, the insertion is more distal and the chondroepitrochlearis is considered to be an atavistic anomaly attesting the ancestral form. The chondroepitrochlearis has been found at autopsy in infants with chromosomal defects such as trisomy 13 with a D/D translocation and trisomy 18. Barash et al suggested that during embryogenesis, the genetic loci for such atavistic anomalies are normally repressed but in trisomy 18, these loci are depressed (8). On the other hand, the phylogenetic significance of the arch of Langer is thought to be a remnant of the panniculus carnosus, a dermal sheath muscle that allows the movement of the skin independent of the movement of deeper muscle masses for example in shivering (9).

Clinical significance of the chondroepitrochlearis muscle has received increasing attention. Lin et al described a bilateral shoulder contracture due to chondroepitrochlearis muscles treated with surgery in order to release the affected muscles (10). Spinner et al showed an ulnar nerve entrapment caused by the accessory muscle (11), and cosmetic issues have also been described due to the webbing appearance of the anterior axillary fold. On the other hand, vascular complications have only been described for the arch of Langer. Sachatello et al (5) and Hafner et al (12) both reported intermittent obstruction of the axillary vein without venous thrombosis caused by an arch of Langer. In 2012, Magee et al first described a case of upper limb deep vein thrombosis caused by this abnormal muscle compressing the axillary vein (13).

In this case, we report what we believe to be the first known vascular complication caused by a chondroepitrochlearis muscle. We know now with enough proof (MRI, ultrasound, phlebography) that the hemangioma at birth was a misdiagnosis. To explain the telangiectasia and the lymphedema at birth, one can emphasize that she had an in utero thrombosis caused by the abnormal chondroepitrochlearis muscle resulting in the development of a collateral network. Her state remained quite stable over the years and her symptoms suddenly increased when she was 38 years old, during a strenuous workout at the gym. We hypothesize that she had an acute injury to the chronic deep vein thrombosis of the axillary vein caused by the repetitive movements. The compression of the axillary vein by the chondroepitrochlearis could have damaged the intima of the vessel, leading to the subsequent thrombosis as in the Paget-Schroetter syndrome (14). The repetitive insults over the years could have damaged the intima in a chronic way leading to the collaterals and the strenuous repetitive movements at the gym could have added an acute lesion resulting this time in an acute thrombosis.

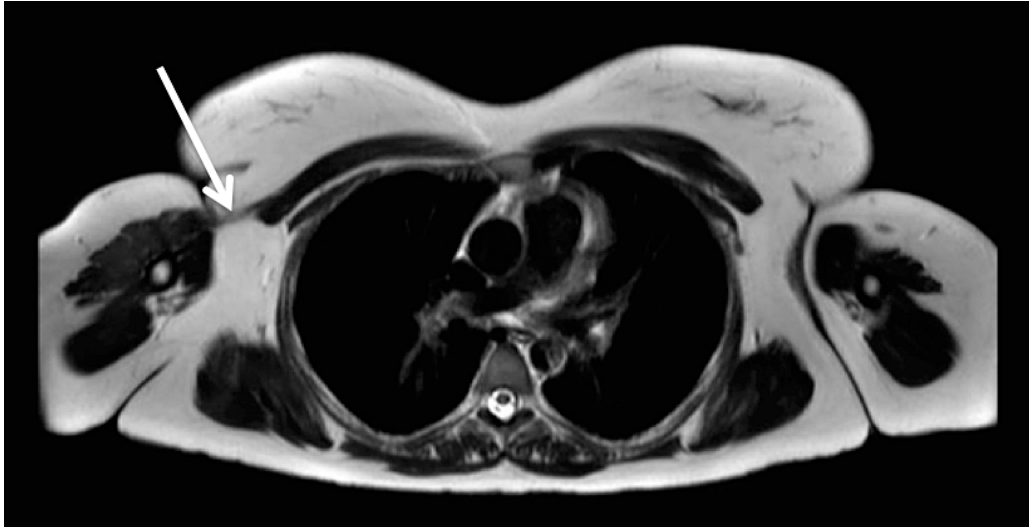


Fig. 6. An early MRI obtained from the patient's records demonstrated in this axial view that the malformation was seen as an individual slip, separated from the tendon of the pectoralis major (arrow).

In this case, once the suspicion of a chondroepitrochlearis muscle was made, a confirmation of the diagnosis was needed. In order to do that, a dynamic echo-Doppler was mandatory. The patient was asked to remain seated and to make an adduction and a protraction of the shoulder mimicking her daily activities. By using the gravitational force and the muscle contraction of the pectoralis major and the chondroepitrochlearis, the intermittent axillary vein compression could be demonstrated. That explains why the pathology was not assessed during standard ultrasound (the patient is in a half-lying position, both arms in abduction). By reviewing her old MRI, the malformation could be identified in the axial view as an individual slip, separated from the tendon of the pectoralis major (*Fig. 6*).

Knowing that the edema was present since her childhood, the presence of telangiectasias, and that filtered fluid returns to the circulation mostly by lymphatics (15), we proceeded to investigate also the superficial lymphatics.

Therefore, we decided to perform a lymphofluoroscopy. In the area of the hand

and forearm, it highlighted functional lymphatics with a normal architecture but just beyond the crossing of the chondroepitrochlearis muscle, the diffusion of the fluorescence suggested a lymphatic involvement (see video in online supplementary materials).

Lymphofluoroscopy was a helpful exam giving us the certainty that the edema was not related to a primary lymphedema. From a pathophysiological point of view, we can understand that the edema was produced by distal hyperfiltration as a result of intermittent compression of the axillary vein (16) reducing its flow and inducing a subsequent increase in capillary pressure. At the same time, the reduced transport capacity of the lymphatic system due to compression of lymph collectors by the abnormal muscle inhibited the ability of the lymphatic system to clear the edema.

Once the diagnosis was confirmed, the decision to perform surgery was made in order to release the abnormal muscle. A very small incision of 1.5 centimeters was performed directly over the abnormal structure in order to minimize the morbidity.

The slip was divided under fluoroscopy to preserve the lymphatic collectors presumably running in the incision area. After this minimally invasive procedure, the pain, paresthesia, and lymphedema disappeared, and a full range of motion was recovered.

CONCLUSIONS

In addition to cosmetic, functional, and neurological problems, the chondroepitrochlearis muscle can be the cause of deep vein thrombosis of the upper limb and should also be considered in the differential diagnosis of upper limb lymphedema.

Correct identification and thorough palpation of the relevant anatomy is mandatory for a clear diagnosis. A dynamic ultrasound in a half-seated position is a simple non-invasive radiologic exam that can highlight the intermittent axillary vein compression. The lymphofluoroscopy is a contributory exam to assess the involvement of the superficial lymphatic system in the pathology. In this case, surgical division completed by physical decongestive treatment was curative.

REFERENCES

1. Lama, P, BK Potu, KMR Bhat: Chondrohumeralis and axillary arch of Langer: a rare combination of variant muscles with unique insertion. *Rom. J. Morphol. Embryol.* 51 (2010), 395-397.
2. Bergman, RA, SA Thompson, AK Afifi, et al: *Compendium of Human Anatomic Variation*. Urban & Schwarzenberg. Münschen, Baltimore. 1988.
3. Loukas, M, RG Louis Jr, M Kwiatkowska: Chondroepitrochlearis muscle, a case report and a suggested revision of the current nomenclature. *Surg. Radiol. Anat.* 27 (2005), 354-356.
4. Landry, SO: The phylogenetic significance of the chondroepitrochlearis muscle and its accompanying pectoral abnormalities. *J. Anat.* 92 (1958), 57-64.
5. Sachatiello, CR: The axillopectoral muscle (Langer's axillary arch): A cause of axillary vein obstruction. *Surgery* 81 (1977), 610-612.
6. Chiba, S, T Suzuki, T Kasai: A rare anomaly of the pectoralis major-the chondroepitrochlearis. *Okijamas Folia Anatomica Japonica* 60 (1983), 175-186.
7. Bergman, RA: Doubled pectoralis quartus, axillary arch, chondro-epitrochlearis, and the twist of the tendon of pectoralis major. *Anatomischer Anzeiger.* 173 (1991), 23-26.
8. Aziz, MA: Anatomical defects in a case of trisomy 13 with a D/D translocation. *Teratology* 22 (1980), 217-237.
9. Besana-Ciani, I, MJ Greenall: Langer's axillary arch: Anatomy, embryological features and surgical implications. *Surgeon* 3 (2005), 325-327.
10. Lin, C: Contracture of the chondroepitrochlearis and the axillary arch muscles. *J. Bone Joint Surg.* 70A (1988), 1404-1406.
11. Spinner, RJ, SW Carmichael, M Spinner: Infraclavicular ulnar nerve entrapment due to a chondroepitrochlearis muscle. *J. Hand Surg.* 16B (1991), 315-317.
12. Hafner, F, G Seinost, T Gary, et al: Axillary vein compression by Langer's axillary arch, an aberrant muscle bundle of the latissimus dorsi. *Cardiovasc. Pathol.* 19 (2010), e89-e90.
13. Magee, C, C Jones, S McIntosh, et al: Upper limb deep vein thrombosis due to Langer's axillary arch. *J. Vascular Surg.* 55 (2012), 234-236.
14. Alla, VM, N Natarajan, W Rugmini, et al: Paget-Schroetter Syndrome: Review of pathogenesis and treatment of effort thrombosis. *West. J. Emerg. Med.* 11 (2010), 358-362.
15. Woodcock TE, Woodcock TM: Revised Starling equation and the glycocalyx model of transvascular fluid exchange: an improved paradigm for prescribing intravenous fluid therapy. *Brit. J. Anaesthesia* 2012 (2012), 384-394.
16. Stanton, AW, B Holroyd, PS Mortimer, et al: Comparison of microvascular filtration in human arms with and without postmastectomy oedema. *Exp. Physiol.* 84 (1999), 405-419.

C. Thomet, MD, JP Belgrado, PT, Ms., Sc.
Université libre de Bruxelles
CHU St. Pierre - Site César de Paeppe
Clinique de Lymphologie de Bruxelles
Rue de Alexiens 11
1000 Bruxelles, Belgium
Phone: 00 32 2 506 73 33
E-mail: celinethomet@gmail.com
belgrado@ulb.ac.be