

Thrombotic obstruction of the subclavian vein: Paget Schrötter syndrome in young adults

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Abstract

Thrombotic obstruction of the subclavian vein is a rare condition of a deep vein thrombosis of the upper limbs. Typical in young, active and healthy individuals, this condition is listed as Paget Schrötter syndrome. At primitive and secondary genesis, this disease results the most common state of the venous form of upper thoracic outlet syndrome (vTOS). Two cases of subclavian venous thrombosis in young adults have been briefly examined in this study, reporting in the discussion the clinical, diagnostic and treatment aspects of this pathology. Given the difficult comparison among the different adopted strategies, and despite the other pathologies that can sometimes show similar clinical events, it is always important to consider its existence because with the correct diagnosis, and the subsequent dedicated therapeutic procedure, it will be possible to obtain an excellent prognosis and a reduction of short and long term morbidity for this kind of patient.

Keywords

Subclavian venous thrombosis; paget schrötter syndrome.

Introduction

This condition is named after physicians James Paget, who first proposed the idea that venous thrombosis causes pain and swelling in the upper limbs, and Leopold von Schrötter, who later related the clinical syndrome to thrombosis of the axillary and subclavian veins [1,2].

In this study, the Authors briefly describe two cases of Paget Schrötter syndrome, that is a thrombotic obstruction of the subclavian vein in young adults in absence of immobility of the upper limbs, reporting in the discussion the clinical, diagnostic and treatment aspects of this pathology.

It is currently classified into primary and secondary; in the primitive form there are predisposing

factors in the determinism of the disease, such as intense efforts of the upper limbs or anatomical anomalies, in the secondary form mostly causal factors such as compressive tumor masses, coagulation alterations or venous catheters. The idiopathic form is extremely rare [3].

Venous stasis appears to be essentially correlated to an acquired cause of exertion during work and sports, although this pathology is referred to as the most common state of the venous form of the upper thoracic outlet syndrome (vTOS), anatomical anomaly that makes the onset of Paget Schrötter's syndrome more likely following continuous stress trauma [4]. Venous obstruction can reveal itself as a partial thrombosis, characterized by transient symptoms of pain, edema and cyanosis of the limb, or as a complete thrombosis, showing more visible and persistent symptoms. From a clinical point of view, the suspicion of subclavian venous thrombosis can be supported either by clinical provocation tests or by radiological investigations. Given the difficult comparison among the different adopted strategies, the treatment is usually the responsibility of multidisciplinary competence and makes use of options such as the conservative ones through anticoagulation therapy, surgical ones through decompression and rehabilitation through the study of postural mechanics and the correction of functional disorders, in order to improve prognosis and morbidity [5].

Clinical Case 1

Patient B.I., female, 28 year old, swimmer, with a history of atopic dermatitis and undefined contraindications to the use of estro – progestogens. She got to our emergency department causing edema and pain in her right arm, and showing signs of cutaneous cyanosis in her right hand, and functional impotence, as from about a 4 days time; the pain in the arm intensified with movement. Ross's maneuver is positive and color doppler ultrasound (Figure 1) shows a hyperechoic thrombus in the right subclavian vein, thrombosis that does not take flow even with activation of the arm muscle pump; thrombosis is also confirmed by angiotomography. Blood tests with D-dimer dosage were negative.



Figure 1: Subclavian Vein Thrombosis.

Clinical Case 2

Patient C. R., male, 26-year-old, worker, with mute medical history. He got to our emergency department for sudden onset of axillary and right arm edema, pain that radiated from the right armpit to the ipsilateral arm, and functional impotence, after sleeping for several hours lying on the right side; the pain

in the arm intensified with movement. Ross's maneuver is negative and color Doppler ultrasound (Figure 2) shows a hyperechoic thrombus in the proximal third of the right subclavian vein with edema of the arm muscles; thrombosis is also confirmed by angiotomography. Blood tests with the D-dimer dosage were negative.



Figure 2: Subclavian Vein Thrombosis.

Discussion

Paget Schrötter syndrome is a form of deep vein thrombosis in the absence of upper limb immobility, typically occurring in the axillary or subclavian vein. It is a relatively rare condition, it occurs in young and healthy patients, it is more frequent in males and it is often unilateral affecting the dominant upper extremity. It is mostly of functionally acquired etiology during sport or work, because it is induced by efforts and excessive and repetitive use of the arm, or it may occur as a sequel to the upper thoracic outlet syndrome due to a vascular venous compression at the level of the costo-clavicular space, 3-5% of cases between 20 and 50 years of age [6].

In this regard, the etiology is in any case multifactorial as precisely after repeated overload (and consequent muscle hypertrophy), after direct trauma (fracture, hematoma), after some rare acquired types of tumors or exostoses, or from congenital causes (supernumerary cervical rib, transverse magapophysis of the seventh cervical vertebra, morphological abnormalities of the first rib or clavicle, anomalies of the anterior scalene muscle) conditions that play a role in promoting the development of thrombosis. In fact, to varying degrees, singly or in combination, they can cause pressure, microtraumatism and/or local inflammation (with consequent fibrosis and stenosis) in correspondence with the subclavian vein which, moreover, is particularly vulnerable in the point out of the thorax [7]. The effect of causal factors seems to become evident when hyperabduction of the upper limb concomitant with the determining factor of effort occurs. Physical effort slows down the blood flow and causes stagnation of blood with subsequent intraluminal thrombus in the subclavian vein; the forced exhalation with closed glottis, which accompanies the effort, worsens the situation because it slows down the venous return [8].

As reported in the literature, about the Thoracic Outlet Syndrome (TOS) the disorders are different depending on whether the compression affects one or more of the three spaces included in this anatomical area located inferior to the neck, that is between the collarbone and the first rib, through the which passes

the neurovascular bundle (brachial plexus, subclavian artery and subclavian vein) [9]. As from the physiopathological point of view of the symptoms we can have cases of neurogenic etiology (nTOS), arterial vascular (aTOS) and vascular venous (vTOS), from a clinical point of view we will have symptoms depending on the anatomical structures involved and on the degree of compression. In fact, the potentially serious and disabling clinical symptomatology, although not easy due to the different nuances of clinical presentation and coexistence between the different forms and for the differential diagnosis with other specific pathologies, can did actually include the appearance of pain, functional impotence, edema, heat, redness or cyanosis from venous engorgement of the upper limb (venous thrombosis), pain, paleness, hypothermia, non-sphygmic pulse and paresis of the limb (arterial thrombosis), pain in the root of the neck, trapezius, shoulder or arm, numbness and/or a sense of weakness and/or heaviness in the arm or muscle spasms or paraesthesia/dysesthesia of the upper limbs and fingers (brachial plexopathy) [10]. The typical symptom of pain, in the course of the associated thoracic district syndrome, can manifest itself in different ways such as at the level of the root of the neck, in the arm or in the upper limb, either at rest or following movements with limb in elevation, or it can be caused by head rotations or deep inhalations.

Specifically, as regards the Paget Schrötter venous syndrome, the patient with partial thrombosis presents transient episodes of edema and cyanosis of the upper limb, often caused or aggravated by the elevation of the arm, or he can be asymptomatic if the partial obstruction is compensated by collateral venous network, even superficial (the vessels turgor of the arm and of the shoulder region are the expression of a vicarious circle of venous obstruction); this does not mean that partial thrombosis is not a predisposing factor for a complete thrombosis due to the continuous phenomena of perivenous inflammation and consequent fibrosis. On the other hand, regarding a complete obstruction the patient shows a more evident clinical profile and an acute course with more evident and persistent pain, edema and cyanosis of the arm [11].

Clinical suspicion of autonomous thrombosis of the subclavian vein can also be supported either by clinical tests or provocative maneuvers during physical examination, and by radiological study [12]. Although clinical test itself is not considered a fixed methodological criteria, and awaiting further comparative studies, we recall some more predictive clinical tests especially if thoracic outlet syndrome is concomitant (in this case the tests will be positive): The Ross test (patient standing with raised arms, abducted shoulders and flexed elbows at 90° for 3 minutes, slowly opens and closes the hands, the test is positive if the patient is unable to hold the position, resulting in ischemia of the limb; this test can be associated with the microvascular study by oximetry), the Adson test (patient is seated, the limb is pulled down and the examined wrist is grasped to appreciate the radial artery, the patient hyper extends and rotates the head towards the examined limb and performs a forced inspiration holding the breath, the test is positive if the radial pulsation disappears), the Halsted test (patient is seated the limb is pulled downwards and the examined wrist is grasped to appreciate the radial artery, the patient hyperextends and rotates the head to the opposite side, the test is positive if the radial pulse decreases or disappears), the Allen test (patient is seated with raised arm, abducted shoulder and flexed elbow by 90°, the examined wrist is grasped to appreciate the radial artery, the patient then rotates the head towards the opposite shoulder, the test is positive if the radial pulsation disappears) and the Wright test (patient is seated with the upper limb in maximum

abduction with extended elbow so that the forearm and hand are above the head in a frontal plane, the wrist to be examined is grasped to appreciate the radial artery, for 1 minute, the test is positive if the radial pulsation is reduced or disappears or if neurogenic symptoms such as paraesthesia appear) [13].

The least invasive but more consolidated diagnostic procedures for diagnostic purposes are ultrasound, angioTAC and MRI. Color Doppler ultrasound maintains a high sensitivity and specificity and represents the diagnostic investigation of first choice, as well as in the subsequent follow up, as inexpensive, easily repeatable and non-invasive, while the angioTAC exam or MRI are mostly used in doubtful cases or in the case of providing more anatomical details [14].

The treatment, often relates multidisciplinary competence, has the two fold objective: The dissolution of the thrombus and recanalization of the venous axis, or at least avoiding the extension of the thrombus, and the prevention of short and long term negative outcomes (acute pain, impotence functional, recurrence of thrombosis, pulmonary embolism); in fact, treatment options, evaluated on a case-by-case basis, can be conservative, surgical or rehabilitative [15]. Regarding conservative treatment, pain management is envisaged, pharmacological thrombolysis (fibrinolytics within 72 hours of the onset of thrombosis and administered for at least 48 hours), alternatively, even in the case of non-response or contraindications to fibrinolysis, by administering direct anticoagulation. This occurs by administering Low Molecular Weight Heparin (LMWH) at a therapeutic dosage for 7 - 20 days, using for example Enoxaparin with a dosage of 150 UI / Kg subcutis for uncomplicated patients, or 100 UI/ Kg subcutis twice a day in cases complicated, followed by indirect anticoagulation, with warfarin, for at least 3 months (in selected cases, therapy can continue by antiplatelet therapy); the subsequent follow-up at the hemostasis and thrombosis center in individuals with haematological disorders is useful [16]. Among the surgical options, in addition to mechanical thrombolysis for venous disobstruction in case of failure to respond to anticoagulant treatment, depending on the needs and for the etiological forms subject to surgical correction, surgical decompression with resection of the first rib (with traditional or minimally invasive thoracoscopic or robotic approach), scanelectomy/scalenotomy or venolysis with venoplasty are indicated [17].

In this regard, surgical timing is still debated. Decompressive treatment, when venous thrombosis is associated with thoracic outlet syndrome, is usually scheduled within 4 to 6 weeks to limit relapse and reduce morbidity, or should be early once the subclavian vein has been recanalized (mechanical thrombolysis that precedes decompression should be performed approximately within 7 - 14 days from the onset of thrombosis), or reserved both for those patients who come to observation after 7 days from onset, with complete thrombosis, and for those patients who are presenting refractory symptoms to conservative treatment; in all these cases, it is advisable to maintain oral anticoagulation therapy for at least another 3 - 6 months in the post-surgery period, followed by any antiplatelet therapy [18].

In addition to the appropriate lifestyle changes (arm position changes while working or the adjustment of athletic gestures in sport), physical therapy and dedicated rehabilitation are important with particular regard to postural mechanics and the correction of functional disorders (improvement of respiratory dynamics, passive stretching and static and dynamic improvement of the bachelors - thoracic district); these

therapies are mostly reserved for those subjects presenting a neurogenic form or those under surgery or with disabilities [19].

Rarely, subclavian vein thrombosis can cause fatal pulmonary embolism; pulmonary embolism is a rare complication especially in the course of extrinsic mechanical obstruction as it seems to prevent proximal embolization [20].

Conclusion

As from literature data, Paget Schrötter's syndrome is a deep vein thrombosis of the subclavian, mainly due to exertion and in young individuals, due by excessive repetitive athletic movements or by lifting of weights, and is more frequently combined with progressive mechanical compression of the thoracic district syndrome. Treatment of venous thrombosis must take into account the presence or absence of concomitant (primary form) or causal (secondary form) factors. Color Doppler ultrasound maintains a high sensitivity and specificity and represents the diagnostic investigation of first choice, as well as in the subsequent follow up. Given the relative rarity of this condition and the difficult comparison among the different adopted strategies, and despite there are other pathologies that can sometimes present similar clinical manifestations, it is important to always consider their existence since and make a case-by-case assessment of patients. For these patients, with a correct diagnosis and a subsequent dedicated therapeutic procedure, it is possible to obtain an excellent prognosis and a clear reduction of short and long term morbidity. In patients with subclavian thrombosis, we believe it is also necessary to investigate the risk factors of thrombophilic or prothrombotic status, for hypercoagulability of the blood, due to the fact that these conditions can determine an elevated thrombotic risk and which actually begins at a young age (before 45 years of age). Further randomized and controlled trials are needed to better define the management guidelines concerning this disease.

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