Caso Clínico

Thoracic Outlet Syndrome (TOS) Mimicking Takayasu’s Arteritis – Case Report

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Abstract

Thoracic outlet syndrome (TOS) is defined as a set of symptoms caused by the compression of the brachial plexus and subclavian vessels in the thoracic outlet region. Anomalies in musculoskeletal structures may be responsible for TOS, including prolonged transverse process of the seventh cervical vertebra, cervical rib, and first anomalous rib and clavicle fractures. The authors describe a case of a young woman with pain in the left forearm, accompanied by intermittent claudication, weight loss, myalgias and ischemic lesions in the fingers, with no pulses and no measurable blood pressure in the left arm, who was initially diagnosed as Takayasu arteritis. The chest radiography showed accessory cervical ribs and the dynamic vascular image tests (Doppler ultrasound and angiography) showed bilateral compression of the subclavian artery, confirming the diagnosis of TOS.

Keywords: Thoracic Outlet Syndrome; Cervical Rib Syndrome; Takayasu Arteritis.

Introduction

Thoracic outlet syndrome (TOS) is defined as a set of symptoms that may occur due to compression of the brachial plexus and subclavian vessels in the thoracic outlet region, between the neck and the axilla. Bone or soft tissue anomalies can be responsible for TOS, including prolonged transverse process of the seventh cervical vertebra, cervical rib, anomalous first rib, first rib or clavicle fracture's and fibromuscle abnormalities.1,2

In the literature there are some TOS classifications according to the compressed structures or the etiology of the lesion. Huang et al. classified TOS in three groups: compression of the brachial plexus (neurogenic TOS), compression of the subclavian vessels (vascular TOS) and another nonspecific form (pain and sensitive symptoms, without either objective signs of neurological compression or alterations in the neurophysiological tests).3

Colli et al. classified TOS into five groups. They...
subdivided the neurogenic TOS in two groups: one with classic signs and symptoms of neurogenic compression accompanied by specific eletroneuro-myography (ENMG) findings and other with clinical and ENMG nonspecific findings. The vascular TOS are also subdivided in: arterial or venous subclavian compression and the fifth group comprises patients with signs and symptoms of post-traumatic neurovascular compression.4

Sanders et al. classified TOS in three groups: neurogenic TOS, arterial TOS and venous TOS.5 Concerning the frequency of different forms of TOS, Huang et al. considered the nonspecific type as the most common and the vascular as the rarer form.3 Neurological symptoms occur over 90% of all TOS cases3,5,6 and the vascular form corresponds approximately to 5% (the arterial TOS represents less than 1%).5,6 The neurogenic and vascular forms may coexist and the distinction between these forms is not easy.3

There is no gold standard test for TOS diagnosis.2,7 Detailed anamnesis and physical examination are the most important tools for the TOS diagnosis. Provocative tests, such as Adson’s test, are nonspecific, and can be positive in asymptomatic individuals. The electro-diagnostic and image studies are not always useful for the diagnosis. Since there is no specific confirmatory test, TOS incidence is variable, ranging from 3 to 80 cases/1,000 inhabitants. It is more frequent in women with age ranging from 20 to 50 years3 possibly because generally women present weakened muscular structures which makes their scapulas lower, predisposing the compression of the structures involved in TOS.1

The differential diagnosis is wide and includes cervical disc lesions, osteophytes, Pancoast tumor, nerve sheath tumor, ulnar and median nerve entrapment, brachial plexitis, spinal cord tumor, shoulder’s diseases, fibromyalgia, multiple sclerosis, Raynaud phenomenon, acute coronary artery disease, venous thrombosis, micro-embolism, Takayasu arteritis, vasospastic disorders, complex regional pain syndrome, brachial plexus injuries and myofascial syndrome.1,3,6

There is no consensus for the best treatment of TOS and it depends on the etiology. The surgical treatment is usually indicated in cases of true neurogenic or vascular TOS and for patients with nonspecific form refractory to the conservative treatment. The best surgical approach in those patients is not defined yet.3

Takayasu’s arteritis (TA) is a chronic inflammatory disease that affects primarily large vessels like aorta and its branches.8 The inflammation leads to stenosis and occlusion of the involved arteries, aneurysms formation or both.9 It may cause the decrease or the absence of arterial pulses, cerebral ischemia, acute myocardial infarction, aortic insufficiency, cardiac congestive failure, limb claudication, hypertension, aneurysm and blindness.8,10 TA affects mainly women in the second and third decades of life8 and can cause premature death.9

Case Report

A 23 y.o. mulatto woman was admitted at our university hospital complaining about burning pain and intermittent claudication in the left forearm for 5 months. She had noticed her skin cold and pale on the left forearm and hand associated with a painful lesion on the second digital pulp in the last two months. The pain and intermittent claudication became worse and were associated with new necrotic lesion on the third finger of the left hand. She was being treated with cilostazol, acetylsalicylic acid and pentoxifylline without improvement. The patient also reported headache, dizziness, fever and a weight loss of 5 Kg. She referred a traumatic left clavicle fracture when she was child and had a history of heavy smoking, alcoholic habits and drug abuse with cannabis and cocaine. The patient had been attended in another hospital where it was prescribed 60 mg/day of prednisone due to the suspicion of TA.

When the patient was attended at our service, there was no left brachial and radial arterial pulse or limb edema. Blood pressure was 140 × 90 mmHg in the right arm, not measurable in the left arm and 160 × 90 mmHg in both lower limbs. A bruit over the left subclavian artery, pale skin in the left hand, mild hypotrophy in thenar and hypothenar regions and diminished temperature in the left forearm and hand were observed. She also presented necrotic lesions on pulp of the second and third fingers and on subungueal region in the fingers of the left hand (Figure 1). Hypoesthesia and paresthesias over the thenar region and palmar aspects of the first, second and third fingers of the left hand became worse with hyper-abduction and extension of the left shoulder. The Adson’s test, Roos’ test and Wright test were positive bilaterally. No episodes of fever were detected during the whole
inpatient period.

Laboratory tests: hemoglobin level was 11.6g/dL, white-cell count was 19,700/mm (1% bands, 78% segmented, 17% lymphocytes, 2% eosinophils, 2% monocytes) and erythrocyte sedimentation rate: 10 mm per hour. Serum creatinine, electrolytes, transaminases and the urinalysis were normal. Serologic tests for viral hepatitis B and C, HIV and syphilis and the anticardiolipin antibody test were all negative. Blood and urine cultures were also negatives.

The chest radiography showed bilateral accessory cervical ribs (Figure 2) and the Doppler ultrasound of the left upper limb exhibited reduced flow in subclavian artery due to costoclavicular entrapment at provocative test. The distal artery was fulfilled by collateral circulation and the subclavian and axillary arteries presented partial thrombosis, with a total thrombosis of the brachial artery.

The exam in the right upper limb also showed reduced subclavian artery flow at hyperabduction without thrombosis. The digital arteriography confirmed the alterations observed at Doppler ultrasound (Figure 3) and the echodopplercardiogram was normal.

The ENMG showed altered sensory and motor fibers conduction of all nerves in the left upper limb with no abnormality in the right one.

The patient was submitted to bilateral cervical rib excision and anticoagulation with improvement of all symptoms.

Discussion

We describe the case of a young woman with neurological and vascular symptoms mainly in the left arm who was erroneously diagnosed as TA. The patient fulfilled 5 of 6 American College of Rheumatology TA classification criteria (age at disease onset < 40 years, claudication of extremities, decreased brachial artery pulse, bruit over subclavian artery, difference of blood pressure between arms > 10 mmHg). The complaints of fever and weight loss also had suggested systemic disease, however, no episode of fever was observed during the hospitalization. The leucocytosis was attributed to the prednisone use and the weight loss was attributed to the ischemic pain and her appetite normalized and she recovered weight after the pain treatment. No signs of infection were observed either at physical exams or at laboratory tests.

The left subclavian artery is described as one of the most frequently affected vessels in TA, va-
rying from 46 to 67.8% in different parts of the world.9,12-15 However, digital ischemic lesions usually are not part of the clinical spectrum of TA. This finding was the first clinical sign to indicate other diagnostic possibility in our case. The vascular form of TOS is less frequently found than the neurological form, and in most of cases requires surgical treatment. We should be alert for this diagnosis in a young individual without high risk for atherosclerosis who presents ischemic manifestations in upper limbs.16

Arterial form of TOS usually presents as cold extremity, weakness, intermittent claudication, diffuse pain and decreased amplitude of the arterial pulse. Venous involvement usually presents with venous thrombosis, superficial vessels distension, edema and pain.6

Massionneure et al. classified arterial manifestations of TOS in 3 types: partial or complete thrombosis, post-stenotic dilatation and post-stenotic aneurysm of subclavian artery. These lesions may cause distal embolization, claudication, vasomotor phenomena and digital gangrene. Retrograde embolization from subclavian to vertebral or carotid arteries are rarely described.17 Thrombosis of the subclavian vein occurs most often in men with strenuous job and presents as edema and cyanosis of the upper limb or distended superficial veins of the shoulders or chest.3

Our patient presented partial thrombosis of the subclavian and axillary arteries and a total thrombosis of the left brachial artery, without aneurisms. The distal ischemic lesions could be due to embolism of proximal thrombus or insufficient collateral vessel for irrigation of distal regions.

There is no specific diagnostic method for none

**Figure 3.** Angiography on neutral position (3A) and hyper-abduction of the right arm (3B). Angiography of the left arm (3C and 3D) demonstrating cervical rib and thrombosis of subclavian and brachial arteries.
of TOS forms. On physical examination, various tests had been described, such as Adson’s test and hyper-abduction, but their sensibility and specificity are not still defined.\textsuperscript{18} Costa-clavicle or Hals-ted’s test, Roos’ test and Wright test are others maneuvers that can be used to evaluate TOS patients. However, false-positive or false-negative results can be found.\textsuperscript{7}

Cervical ribs are described in 10% of TOS patients and in 0.01 to 0.5% in the general population in which most of them are asymptomatic.\textsuperscript{3} Cervical ribs are more common in women than men and bilateral in more than 50%.\textsuperscript{19} The presence of cervical rib is not diagnostic for TOS and its absence does not invalidate the diagnosis.\textsuperscript{1} It is unknown why only few patients with cervical rib develop TOS. The format and the consistency of fibrous band that leagues this rib to the first rib seem to be one of the factors involved in the development of symptoms. Nevertheless, it seems to be related to some sports and jobs that demand a prolonged arm hyper-abduction.\textsuperscript{6}

In comparison to other image methods, Doppler ultra-sound and angiography in neutral and hyper-abduction of the arm are useful for the diagnosis of the vascular form of TOS.\textsuperscript{16} Doppler ultrasound has limitations in obese individuals and in areas in which bone structures overlie vessels. In these cases conventional or magnetic resonance (MR) angiography are useful\textsuperscript{20} specially to demonstrate fibrous bands and brachial plexus deflection in patients without cervical rib.\textsuperscript{7}

Nervous conduction studies and ENMG can help the diagnosis by revealing decreased senso- rial or motor action potential in ulnar and/or median nerves. ENMG can also reveal abnormalities in the intrinsic muscles of the hands.\textsuperscript{3}

The treatment of TOS is still controversial\textsuperscript{21} and can be conservative or surgical.

Conservative treatment includes behavioral changes by avoiding activities and positions that determine its appearance, beside rehabilitation with strengthening of pectoral musculature and postural positioning. The improvement after conservative treatment varies from 50 to 90% and depends on its etiology.\textsuperscript{3}

Surgical treatment involves surgical decompression and, when necessary, vascular reconstruction.\textsuperscript{4} Cervical rib excision and/or first rib excision; resection of cervical muscles, brachial plexus neurolysis and complementary vascular procedures are some of surgery procedures applied to TOS pa-

\textsuperscript{16} The presence of cervical rib per se is not an indication for surgery, unless there is failure in conservative treatment or incapacitating symptoms.\textsuperscript{19} Vascular reconstruction after decompression presents good results on a short and long term follow-up. Endovascular treatment with stents is also described, however long term studies showed high chance of re-stenosis. The treatment of venous involvement include early use of thrombolytic agents followed by anticoagulation and late surgical decompression, or thrombolytic therapy through catheter followed by early surgical decompression and anticoagulation followed by balloon angioplasty in cases of stenosis. These options had better results when compared to anticoagulation alone.\textsuperscript{16}

We described a young woman with neurogenic and vascular TOS characterized by intermittent claudication, bruit over subclavian arteries, absence of arterial pulse and non measurable blood pressure in the left arm. All of these features could be suggestive of TA. However, TA is a chronic arteritis and rarely causes ischemic cutaneous lesions as presented by our patient. This case emphasizes the importance of a detailed clinical examination and the differential diagnosis between these pathologies which have different treatment and prognosis. We should pay attention to suspicious cases of TOS, because some of its manifestations may be irreversible in case of late diagnosis.

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