CRITICAL UPPER LIMB ISCHEMIA CAUSED BY INITIALLY UNDIAGNOSED THORACIC OUTLET SYNDROME – CASE REPORT

ŁUKASZ DZIECIUCHOWICZ¹, WOJCIECH WŁODARCZYK², GRZEGORZ OSZKINIS¹

Department of General and Vascular Surgery, Medical University in Poznań¹
Kierownik: prof. dr hab. G. Oszkinis
Department of General Surgery with Services of Oncological and Vascular Surgery, Specialist Hospital in Piła²
Ordynator: dr n. med. K. Nowicki

A case of a progressive ischemia of the right upper extremity in a 57-year-old male caused by primarily undiagnosed compression of a subclavian artery by an accessory cervical rib is presented. Critical limb ischemia persisting for 7 years despite conservative treatment and a thrombectomy of upper extremity arteries and thoracic sympathectomy, led to a loss of three fingers and development of a non-healing ulceration of right upper extremity. Eventually a cervical rib resection together with a subclavio-brachial venous bypass graft permanently reversed the critical limb ischemia and allowed the ulceration to heal.

The presented case underscores the importance of an early surgical intervention in patients with upper limb ischemia and anatomic abnormalities in a thoracic outlet.

Key words: limb ischemia, thoracic outlet syndrome

Critical ischemia is most commonly found in the lower limbs. Critical upper limb ischemia is less common and can be caused by thrombophlebitis, posttraumatic changes, ergot intoxication (1), an old embolus or compression syndromes. Due to its rare occurrence, symptoms of critical upper limb ischemia can be misinterpreted, often as neurological abnormalities, which in turn may delay proper diagnosis and management (2, 3). In case of compression syndromes, early diagnosis is of significance since long-term compression of an artery, continuous or recurrent, may result in chronic occlusion of peripheral arteries due to peripheral thrombosis or embolism. In an initial period, due to relatively well developed collateral circulation, these symptoms can be mild, which also contributes to the delay in proper diagnosis.

The aim of this paper is to provide description of a critical ischemia of the upper limb caused by initially primarily undiagnosed compression of the right subclavian artery by an accessory cervical rib.

CASE REPORT

A 57-year-old male (R.H.) visited Outpatient Department of Vascular Surgery in January 2011 due to critical ischemia of the upper right limb. He complained of resting pain in the upper right limb. Clinical examination revealed unhealing wound after an amputation of the distal digit of the 2nd finger and status after amputation of distal fingers of the 1st and 5th fingers. Pulse was undetectable on the upper limb arteries. History regarding vascular risk factors demonstrated cigarette smoking, 20 cigarettes daily for 37 years and treated hypercholesterolemia. He did not have diabetes mellitus or arterial hypertension. He worked as a grinder. His brother was diagnosed with atherosclerosis of the lower limbs.
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First symptoms appeared seven years earlier. Initially the patient was treated at Outpatient Department of Surgery with a diagnosis of obliterating thrombophlebitis. Two months after the onset of symptoms the patient was consulted due to exacerbation of pain of the right upper limb at Outpatient Department of the Hospital Emergency Department where limb ischemia was diagnosed and medical treatment was recommended (acetylsalicylic acid, pentoxyphylline, bufomedil and ketoprofen). One month later the patient was treated at Department of General Surgery due to infected necrosis of the distal digit of the 5th right finger. Arteriography performed at that time demonstrated occlusion of distal segment of the brachial artery and disseminated occlusions of forearm arteries. Despite the fact that an accessory cervical rib was found, diagnosis of obliterating thrombophlebitis was sustained and medical treatment (acetylsalicylic acid, pentoxyphylline, fumaran bencyclane) was provided. One month later the patient was hospitalized at the Clinic of Vascular Surgery. AngioCT imaging confirmed the presence of occlusion of the brachial artery. Subclavian and axillary arteries were patent, without any signs of an aneurysm (fig. 1). Therefore thrombectomy of the brachial artery was performed and organized thrombi were removed, resulting in very good inflow of arterial blood. Fibrotic thrombi were also removed from the radial artery. However, no retrograde outflow from the palmar arch was achieved. A fibrotic thrombus was removed from the ulnar artery: a 5-cm segment in the wrist region, obtaining negligible retrograde outflow. Intraoperative arteriography (fig. 2) demonstrated occlusion of the palmar arch from the radial side and partial patency of the arch from the ulnar side, without contrast enhancement of digital arteries, which was a decisive factor against lack of possibility of vascular reconstruction on the forearm. After the surgical treatment blood supply to the hand improved and ischemic changes on the palmar surface healed and necrosis of distal digits of 1st and 5th fingers underwent gradual demarcation. Low molecular weight heparin and pentoxyphilline were prescribed and the patient was advised to avoid placing the right upper limb in the elevated and abducted position. Ischemia of the right upper limb was diagnosed as a result of brachial, radial and ulnar artery thrombosis and vibration-induced disease was suspected. The patient was transferred to Clinic of Thoracic Surgery to undergo surgical resection of the cervical rib.

Ultrasound imaging with double flow imaging demonstrated patency of subclavian, axillary artery and radial and ulnar arteries in the ½ proximal forearm. Reduction of flow velocity to 38 cm/s was demonstrated in the axillary artery behind the clavicle versus 99 cm/s in the subclavian artery, and in the functional position of the limb/hand under the head/ increased flow velocity in the axillary artery to 1.8 cm/s and morphological stenosis of this artery at the site of its appearance from under the clavicle. Despite such image, the patient was not qualified to resection of the cervical rib.

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Fig. 1. Computed tomography angiography of the right upper limb – a patent subclavian and axillary arteries and an occluded brachial artery can be seen

Fig. 2. Intraoperative arteriography of the right upper limb after thrombectomy of the brachial artery and forearm arteries – occlusion of the palmar arch from the radial side can be seen
cervical rib. One month later right sided thoracoscopic thoracic sympathectomy was performed in Clinic of Surgery of Military Clinical Hospital. After 10 days the patient was hospitalized at Department of Surgery and treated with alprostadil for 20 days. Five months later (in December 2004) partial amputation of right 1st and 5th fingers was performed. The patient was discharged home and was prescribed pentoxyphylline, buflomedil and ketoprofen.

Later the patient underwent periodic follow-up at Outpatient Department of Vascular Surgery. In December 2006 he underwent Doppler imaging with double imaging that demonstrated arterial patency of arteries up to ½ proximal forearm; in the radial artery at the level of the wrist the flow was low-resistance and no blood flow was found in the ulnar artery.

In 2010 worsening of blood supply to the hand appeared: dry necrosis of the distal digit of the 2nd finger resulted. The patient visited Hospital Emergency Department twice (in July and in September). He did not have any acute signs or symptoms of ischemia. The hand was warm, with preserved mobility and tactile sensation, without pulse on brachial, ulnar and radial arteries. Low molecular weight heparin was prescribed at therapeutic doses, acetyl-salicylic acid, sulodexid and follow-up at Outpatient Department of Vascular Surgery was advised.

Four months later (January 2011) ultrasound imaging with double flow imaging and angioCT of arteries were done (fig. 3) and demonstrated occlusion of subclavian artery 1.5 cm downstream to the bifurcation of brachiocephalic trunk, at the level of crossing the cervical rib, occlusion of axillary artery, filling of initial 2.5-cm segment of brachial artery (from which collateral vessels to the arm periphery originated) from the collateral circulation. Negligible contrast enhancement of very thin arteries was seen at the forearm. Waveless flow at the level of the wrist was seen at a velocity of 23 cm/s in the radial artery and 13 cm/s in the ulnar artery. The patient had signs and symptoms of hand ischemia and dry necrosis of the distal digit of the 2nd finger and was referred to Clinic of General and Vascular Surgery of Medical University of Poznań. Before hospitalization the patient underwent amputation of the distal digit of the 2nd finger in the outpatient setting. During the hospitalization at the Clinic, the cervical rib was removed through suprACLavicular access and the anterior scalenius muscle was resected, thrombectomy of subclavian artery was done and thrombectomy of an initial segment of right brachial artery was attempted. Very good blood inflow was achieved, however retrograde outflow from brachial artery was not achieved. Therefore a subclavian-brachial by-pass was implanted using a patient’s own vein, resulting in resolution of pain of the hand and healing of a wound after the amputation of the distal digit of the 2nd finger. The postprocedural period was uncomplicated. The patient was discharged home on day 2 after the surgical procedure. Double antiplatelet treatment was prescribed: acetylsalicylic acid at a dose of 75 mg daily and clopidogrel at a dose of 75 mg daily. The patient has been followed-up for eight months now. Angio-CT imaging has demonstrated patent subclavian artery, subclavian-brachial by-pass and interosseous artery on the forearm (fig. 4). Currently he does not complain of any pain of the limb, his hand has normal temperature, all wounds have been healed (fig. 5), pulse can be felt on the subclavian-brachial by-pass.

**DISCUSSION**

A case was described of a progressive, chronic ischemia of the right upper limb in a
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57-year old male. Resection of an accessory cervical rib and implantation of a subclavian-brachial by-pass resulted in inhibition of further progression of ischemia and healing if ischemic changes. Therefore one can assume that initially undiagnosed accessory cervical rib was the most probable cause of progression of ischemic changes. Later, despite detection of this accessory cervical rib, no association was found between this anomaly and progressive limb ischemia. Initially obliterating thrombophlebitis was diagnosed. In this disease entity surgical treatment is possible in very rare cases and does not provide satisfactory results. However it must be emphasized that the clinical presentation, except for a risk factor – cigarette smoking, did not meet generally accepted criteria of Buerger’s disease such as: symptom onset before the age of 45, lack of proximal source of emboli and typical angiographic image (4). The symptoms appeared at the age of 50 years and were limited only to a single limb. Localized symptom onset in Buerger’s disease is possible, but usually, as the disease progresses, symptoms appear in all four limbs, in a variable extent. Furthermore, arteriography did not demonstrate typical, unique symptoms for this disease such as corkscrew-like collateral blood vessels. Medical history did not involve migrating superficial phlebitis, typical for this disease, that could precede or coexist with signs and symptoms of limb ischemia or Raynaud’s sign that is present in approximately 40% of patients with Buerger’s disease (4, 5). Furthermore epidemiological studies indicate that prevalence of Buerger’s disease has recently been significantly reduced. In view of lack of occupational exposure to vibrations, diagnosis of vibration-induced disease in this patient was even less probable.

Despite the fact that an accessory cervical rib was described by Galen as early as in the 2nd century, Murphy and Mayo were the first to report association between occurrence of a cervical rib and injury of the subclavian artery in the early 20th century (6, 7, 8). In 1956 Peete et al. coined a term “thoracic outlet syndrome” referring to symptoms resulting from compression of a neurovascular band in the upper thoracic outlet (6). Signs and symptoms of upper limb ischemia related to the thoracic outlet syndrome may appear at the age of 13 – 74 years and most commonly are related to a presence of poststenotic aneurysm of dilation of the subclavian artery (9, 2). A factor that

Fig. 4. Computed tomography angiography – a patent subclavian-brachial by-pass can be seen

Fig. 5. Patient’s hand 8 months after resection of an accessory cervical rib and implantation of subclavian-brachial by-pass
could contribute to omission of a causal relation between an accessory cervical rib and progressive upper limb ischemia in this case, was lack of poststenotic dilation of the subclavian artery. Poststenotic aneurysm of the subclavian artery is classically reported as a source of peripheral embolism. Turbulent blood flow in the aneurysm results in formation of perimural thrombi that can detach and migrate as an embolic material to the upper limb arteries. Criado et al. found an aneurysm of the subclavian artery in 11 (85%) of 13 patients who developed an acute limb ischemia related to the thoracic outlet syndrome, while a poststenotic dilation was found only in the remaining two patients (2). Similarly, Durham et al. found peripheral embolism without the presence of a poststenotic aneurysm only in one of 27 upper limbs with arterial abnormalities related to the thoracic outlet syndrome. Therefore, embolism of peripheral arteries related to the thoracic outlet syndrome is possible without morphological changes in the subclavian artery, although it is rare.

Another factor that could contribute to omission of a causal relation between compression of the subclavian artery and progressive upper limb ischemia in this case, was the fact that compression of this artery was found in more than 10% of healthy young subjects with abduction and external rotation of the upper limb in the shoulder joint (10). An accessory rib has been also found to occur in 0.5-1% of the general population, but compressive symptoms occur only in 10% of subjects with an accessory rib (11). On the other hand, an accessory rib is the most common cause of arterial abnormalities in the thoracic outlet syndrome (2, 9).

Surgical treatment involves resection of an accessory cervical rib and, if needed, surgical reconstruction of the subclavian artery. These surgical procedures are performed under general anesthesia, from the supraclavicular access. In some cases, when a patient has a full-length cervical rib, a second incision under the clavicle is needed to fully remove it. It must be emphasized that it is a technically demanding procedure that requires very good knowledge of anatomy of this region and precise surgical technique, since there is a high risk injury of such structures and the subclavian artery and vein, brachial plexus, phrenic nerve or the thoracic duct.

CONCLUSIONS

1. Patients with chronic upper limb ischemia should always be tested for the thoracic outlet syndrome.
2. Detection of anatomical abnormalities that could result in compression of the subclavian artery, could be an indication to surgical intervention in these patients.

REFERENCES