

CONTINUING MEDICAL EDUCATION ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

Vascular Diseases Quiz – Case 44

A 46-year-old left-handed male with no significant past medical history was referred to vascular surgeon by his family physician for evaluation of a 3-month history of pain and cyanosis of the skin of the third, fourth, and fifth fingers of his left hand.

He also complained of small sore ulcers on his fourth and fifth fingers for over 6 weeks. His was a carpenter by occupation and he did admit to using his left hand as a hammer at times striking objects with the heel of the hand. He denied any tobacco or recreational drug use.

On physical examination he had normal and symmetrical radial and ulnar pulse, as well as blood pressure on both arms. There was bluish discoloration over the distal phalanx of the third, fourth and fifth fingers and superficial ulceration was noted on his fourth and fifth fingertips. Allen test was normal on both hands. Patient had full range of motion of the fingers, no tenderness and normal muscle strength. Thrombophilia and collagen vascular disease investigations done by his family physician were negative. A digital subtraction arteriogram (DSA) of the left upper extremity was performed and revealed ulnar artery aneurysm at the palmar level and occlusion of digital arteries of 3rd, 4th and 5th fingers (fig. 1).

What is the most likely diagnosis?

1. Raynaud's syndrome
2. Vasculitis
3. Thrombophilia
4. Arterial thoracic outlet syndrome
5. Hypothenar hammer syndrome (HHS)
6. Bulrger disease (thromboangiitis obliterans).

What is the "gold standard" diagnostic test for establishing the diagnosis of this condition?

1. Computed tomographic scanning
2. Magnetic resonance imaging
3. Angiography (DSA)
4. Duplex Doppler imaging
5. Allen test.

What is the best treatment in this case?

1. Calcium channel blockers
2. Antiplatelet agents or anticoagulation
3. Cold avoidance
4. Surgical resection and reconstruction



Figure 1. Aneurysm of the palmar ulnar artery with embolic occlusion of multiple digital arteries (black arrows).

5. Thrombolysis.

Comment

Family physician very often deals with patients seeking health care consultation for hand and finger pain. Most of the cases are attributed to musculoskeletal system disorder. On rare basis, however, hand and finger pain is caused by vascular insufficiency as a result of a variety of vascular overuse syndrome. An unusual vascular overuse syndrome that is being illustrated by this case is the HHS.

This clinical entity was first described by Guttani and Von Rosen. The term "hypothenar hammer syndrome" was suggested by Conn

et al in 1970 who proposed that repeated blunt trauma to the hypothenar portion of the hand can lead to ulnar artery damage. Typically, it occurs in men of mean age of 40 years, in occupations and sports where the heel of the hand is used as a hammer or is subject to repeated force. Occupations at risk include metal workers, machinists, mechanics, miners, sawmill workers, carpenters, bricklayers, butchers, bakers and those using vibrating tools. Occasionally, it can occur in various sports, including golf, mountain biking, baseball, softball, hockey and martial arts.

The ulnar artery is vulnerable in the distal portion of Guyon's canal, where it is not protected by the palmaris brevis muscle. At this site, the unprotected superficial palmar branch of the ulnar artery can be compressed against the bony hook of the hamate which may lead to arterial damage, thrombus formation, and aneurysm of the artery or microemboli leading to digital ischemia. Sensory branches of the ulnar nerve run nearby and their involvement may contribute to neurologic symptoms such as paresthesia and pain. Recent studies have suggested the involvement of focal fibromuscular dysplasia in the pathophysiology of the development of HHS in patients with occupational repetitive striking of the hands.

Clinical signs and symptoms may present dramatically or subtle and may be remote from the episode of injury. Typically, a male patient will present with ischemia of the second, third, fourth or fifth digits of the dominant hand. Patients may complain of digital pain with paresthesia, cold sensitivity, phasic blanching or discoloration of the fingertips, finger claudication, hypothenar pain or Raynaud's phenomenon. Occasionally, ischemic symptoms are present but not of sufficient severity for the patient to seek medical evaluation.

In HHS, findings on physical examination include hypothenar callus and tenderness, pulsatile hypothenar mass in the case of aneurysm formation, and unilateral episodic blanching or purplish discoloration of the second, third, fourth or fifth fingertips. A positive Allen test (test for ulnar artery patency by occluding radial and ulnar arteries and then releasing ulnar artery pressure) may provide important diagnostic clues when ulnar artery is occluded. In severe cases, gangrene, ulceration or eschar formation of the involved fingertips may occur.

Differential diagnosis incorporates large range of etiologies of vascular causes. It can be categorized into diagnoses that commonly affect the lower extremities (atherosclerosis, cardiac embolization, hypercoagulable states, and thromboangiitis obliterans), and those that are usually isolated to the upper extremities [Raynaud's syndrome, vasculitis (e.g., systemic lupus, rheumatoid arthritis), radiation arteriopathy, arterial thoracic outlet syndrome, and hypothenar hammer syndrome].

Thromboangiitis obliterans (Buerger's disease) is a non-atherosclerotic segmental inflammatory disease of small and medium vessels in the extremities, which is strongly associated with smoking. Inflammatory diagnoses (e.g., systemic lupus erythematosus,

rheumatoid arthritis) are excluded with normal findings of inflammatory or immunologic markers (i.e. acute phase reactants such as ESR, CRP, and autoantibodies, such as ANA, RF, and complement levels). Raynaud's syndrome is characterized by episodic attacks of vasospasm of arterioles of the distal extremities, commonly in female patients and usually with a bilateral pattern. In this case there is no history of cardiac arrhythmia and a negative hypercoagulable evaluation, making cardiac embolization or thrombophilia unlikely.

Arteriography remains the gold-standard diagnostic test, and can help direct surgical management. Ultrasonography may be useful in assessing patients noninvasively, and digital pressures can determine the patency of digital arteries. Ultrasound can also identify some ulnar artery aneurysms that may be missed by arteriography due to the presence of mural thrombus.

All patients with HHS should be counseled about minimizing further trauma to the hands. In case of asymptomatic ulnar artery occlusions non operative treatment is suggested. Padding and avoidance of further trauma (may require change of occupation) to the hands can have significant clinical improvement and avoid surgical intervention. Other non-operative care may include: cold avoidance, calcium channel blockers (nifedipine, diltiazem), antiplatelet agents or anticoagulation, local care of fingers with necrosis, and pentoxifylline to reduce blood viscosity.

Reconstruction is the preferred approach for patients with significant ischemic changes, particularly when there is ulnar artery occlusion or a patent embolizing source, as illustrated in this case. Vascular reconstruction with a short graft and normal inflow is often successful. Most patients with HHS can obtain a high degree of functional restoration. Thrombolysis may be beneficial in acute setting of recently embolized digital vessels. Occasionally, thrombolysis may uncover an unsuspected ulnar artery aneurysm. Patent and embolizing palmar ulnar arteries require either ligation (in the setting of a patent palmar arch without skin ulceration), or interposition vein reconstruction.

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Answers:
1. Hypothenar hammer syndrome
2. Angiography (DSA)
3. Surgical resection and reconstruction

