Case Report

Hypothenar Hammer syndrome: importance of MR and conventional angiographic findings

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Abstract: Hypothenar Hammer syndrome (HHS) is a rare diagnosis characterized by ulnar insufficiency, often accompanied by ulnar nerve neurologic deficit. This syndrome is caused by repetitive trauma to the ulnar artery in the Guyon's or ulnar canal, causing arterial intimal dissociation with possible aneurysmal formation, vasospasm, or even thrombosis. Hypothenar hammer syndrome is often suspected clinically using Allen's test, which is a sensitive but nonspecific test. Ultrasound can be used for initial evaluation; however conventional angiography has been the gold standard for diagnosis. Conventional angiographic findings include classic tortuous, corkscrew appearance of the ulnar artery. MR angiography is frequently used as a noninvasive imaging modality and has an advantage of multiplanar imaging, high soft tissue contrast, as well as lack of ionizing radiation. Accurate diagnosis of this syndrome is important to distinguish hypothenar hammer syndrome from other similar clinical entities such as Raynaud's phenomenon in order to appropriately treat this condition. This entity should be kept in mind when a cardiac patient with certain occupational hazards has to undergo radial artery access for catheterization. Management of hypothenar hammer syndrome is currently debated, though most commonly is treated conservatively, with or without added surgical management.

Keywords: Hypothenar Hammer syndrome (HHS); magnetic resonance angiography (MRA); ulnar artery insufficiency; Raynaud's phenomenon; hypothenar pain

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Introduction

Hypothenar Hammer Syndrome (HHS) is a rare diagnosis of ulnar artery insufficiency caused by repetitive blunt trauma to the hypothenar region of the hand. First coined by Conn *et al.* in 1970, HHS is most prevalent among middle-aged men with occupations that result in the frequent use of the hypothenar region of the hand to pound or squeeze hard objects, examples of which include construction workers, auto mechanics, and miners (1). There have also been incidences of HHS associated with overexposure to the use of vibratory tools and machinery, as well as in athletes from sports where trauma to the palm frequently occurs (2). Patients may present with unilateral Raynaud's phenomenon, tenderness to the hypothenar region, pain and discoloration of the digits, most commonly the 4th and 5th digits, and increased sensitivity to cold in the affected hand. Symptoms are usually episodic, and may or may not be precipitated immediately by trauma. In severe cases, ulceration and necrosis can result in gangrene of the digits (3). In primary care settings where HHS may be misdiagnosed as musculoskeletal overuse, increase awareness can lead to early intervention before ischemic changes become irreversible (4). In this case report, we highlight the imaging findings associated with HHS using angiography and diagnostic features that can distinguish HHS from other diseases with similar symptomatology.

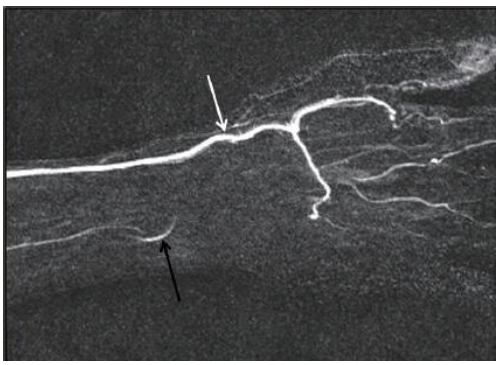

Figure 1 Distal radial artery (white arrow) is widely patent without significant stenosis. The radial artery supplies the princeps pollicis artery which is patent. Deep palmar arch appears patent. The second common digital artery is being supplied by the deep arch and is patent. A common trunk to the third and fourth common digital arteries appears to be patent. The medial aspect of the deep palmar arch terminates at the level of origin of a very faintly opacified short segment of the fifth digital artery which is not seen beyond its proximal 1–2 cm. The ulnar artery (black arrow) is faintly opacified in the lower forearm but occludes at the level of the distal forearm/proximal wrist region and is not identified. The superficial palmar arch is not identified.

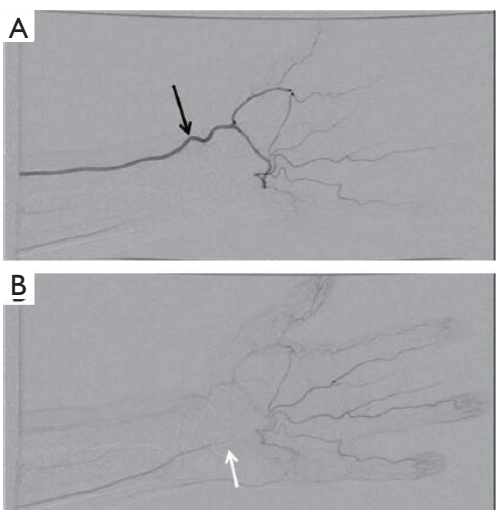

Figure 2 Digital subtraction angiogram image of two successive frames showing normal caliber radial artery (black arrow in A) and an abnormal and small caliber ulnar artery (white arrow in B) with delayed filling. Note deep palmar arch is supplied by the radial artery. Superficial palmar arch, which is mostly supplied by ulnar artery, was not seen.

Case presentation

The patient is a 46-year-old right-hand dominant male who works in auto upholstery presented to his primary care provider with a complaint of progressive left-hand pain and numbness. His symptoms involve the ulnar aspect of his hand, in particular, the left 4th and 5th digits, progressively worsening for the past 3 years. The patient states that he is regularly exposed to repetitive trauma to his palm regions. He was previously diagnosed with right-hand HHS, which was treated surgically with resection of a pseudoaneurysm and primary repair of the ulnar artery. The left-hand pain is not particularly relieved by any medications but was aggravated after a more recent trauma to his left wrist. He has a history of tobacco use, approximately 1 pack per week but quit 3 years prior to presentation. He is otherwise in a good state of health.

During his evaluation in the primary care physician's office, the patient was found to have palpable ulnar and median pulses, slightly diminished on the left. The Allen test is positive. Electromyography performed demonstrated normal conduction of the left ulnar and median nerves. At this time, the patient was referred to our imaging center for magnetic resonance angiography (MRA) due to high clinical suspicion for HHS. On MRA, the patient is found to have occluded left ulnar artery in the region of the ulnar canal, also known as Guyon's canal (*Figure 1*). There was no corkscrew of the distal digital arteries. There was no evidence of fracture of the carpal bones. Subsequently, the patient was referred to angiography of the left upper extremity, which demonstrated progressive narrowing and delayed opacification of the left ulnar artery with occlusion at the level of the carpal bones (*Figure 2*). Imaging findings were consistent with the HHS.

The patient was subsequently referred to a hand surgical specialist due to progressive symptoms. The patient was subsequently taken to the operating room for a reverse saphenous vein graft to bypass the thrombosed left ulnar artery. Intraoperatively, an occluded segment of the ulnar artery was noted in the Guyon's canal, noted to have some intimal disease. The patient tolerated the procedure well, with a good ulnar pulse.

Discussion

The ulnar artery branches off at the level of Guyon's Canal, with one branch forming the deep palmar arch while the other branch continues to exit Guyon's canal to

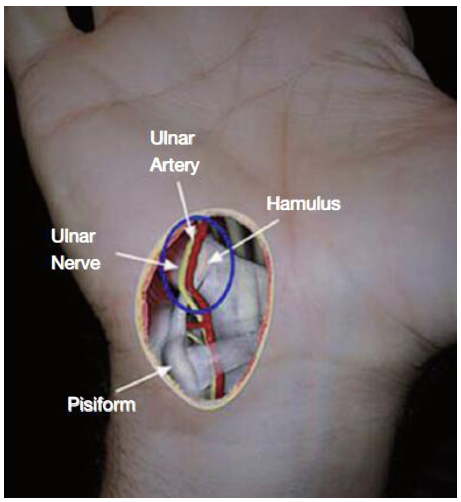

Figure 3 3D representation of the anatomical region affected in HHS. After exiting Guyon's canal, the superficial ulnar artery is at risk for compression against the hook of the hamate (With permissions from radsource.us. Weblink- http://radsource.us/hypothenar-hammer-syndrome/).

form the superficial palmar arch, both with radial artery contributions (*Figure 3*). Study of arterial pattern estimates that in 37% of individuals, the superficial palmar arch is derived solely from the ulnar artery (5). The course of the artery just anterior to the hamate bone coupled with the thin overlaying tissue makes this portion of arterial supply susceptible to injury. Repetitive trauma to this region compresses the ulnar artery against the underlying bone (hamate), resulting in intimal deterioration that encourages the formation of an aneurysm, thrombotic occlusion, and vasospasm (6). Similar mechanisms have been described in several other vascular disease patterns (7). Due to the proximity of the ulnar artery to the ulnar nerve, associated compression of the nerve either from the trauma or by aneurysm growth can cause additional symptoms such as paresthesia of the digits (8).

Diagnosis of HHS can be made with Allen test, Doppler ultrasound, and angiography. When performing Allen test, a patient's radial and ulnar arteries are compressed simultaneously to reduce blood flow to the hand. The pressure is released to one artery at a time to measure the time it takes for the color to return to the palm. A refill time of less than 5 secs is normal, negative for Allen's test. A positive Allen's test in HHS is characterized by normal filling by the radial artery and delayed filling by the ulnar artery, suggesting occlusion to blood flow. The sensitivity of the Allen test for HHS is 83% (2). However, a positive Allen's test can be found in wide variety of conditions such as incomplete anastomosis between the radial and ulnar artery branches. While Allen's test is easy to implement in a clinic setting, it lacks specificity and should be used in conjunction with other diagnostic tools.

Another diagnostic tool often used to study peripheral vasculature is Doppler ultrasound. This non-invasive procedure has been used in previous cases to assess thrombosis and aneurysmal dilatation to the ulnar artery (9). However, the gold standard and deterministic tool used in the diagnosis of HHS remain to be the conventional angiography. Features of HHS on angiography include the tortuosity, corkscrew-like appearance of the ulnar artery at the hypothenar eminence, aneurysmal dilation, occlusion, or stenosis in the same region that interrupts blood flow to the palmar arches (3). Computed tomography angiography (CTA) and MRA have also been used with the added benefits of being able to detect bony deformities and traumatic lesions, both being minimally invasive (10). The downside of using CTA and MRA to study ulnar patency is the inability to provide quantitative information on blood flow.

While the diagnosis of HHS is characterized by ulnar insufficiency, abnormalities in ulnar artery flow associated with trauma appear to be more common and can often remain asymptomatic. Ginn *et al.* studied the prevalence of ulnar artery abnormalities and ischemic changes to the digits in baseball players (11). Result suggests that catchers are more likely than other players to experience early vascular and ischemic changes to the ulnar artery despite having no report of symptoms, which is significant because catchers experience higher incidences of hand trauma from catching high-speed impact balls. Such observation is not unique to baseball. However, it is interesting to note not all individuals with a history of hypothenar trauma are diagnosed with HHS. A cohort study on 79 auto-mechanics who habitually use the hypothenar region of their hands revealed an HHS incidence of 14% (12). Another study of 300 individuals with occupational exposure to vibratory machinery found an HHS incidence of 9% (2). Histological examination of resected ulnar arteries from patients diagnosed with HHS suggests a higher incidence of vascular changes similar to fibromuscular dysplasia, which suggests an anatomical susceptibility that may predispose certain individual to HHS, which could explain the low incidence among at risk population (13). Additionally, there is also evidence showing a positive correlation with tobacco use in HHS patients (14).

The clinical presentation Raynaud's phenomenon and digital ischemia may result in misdiagnosing HHS for diseases with similar symptomatology and of higher prevalence. Differential diagnosis includes primary Raynaud's syndrome, connective tissue disorders, vasculitis, emboli stemming from large vessel disease, thromboangiitis obliterans (Buerger's Disease), and coagulation disorders (3). Primary Raynaud's is characterized by vasospasm in the digits leading to pain, discoloration, and cold intolerance in absence of underlying etiology. A form of reversible Raynaud's has been linked to exposure to vibratory tools, much like in the case with HHS, named hand-arm vibration syndrome (HAVS) (15). Their shared etiology linked to occupational overuse makes distinguishing the two syndromes difficult but important in clinical management. Unlike HHS, abnormal angiographic findings of primary Raynaud's and HAVS are typically limited to distal digital arteries, with reports of cork-screw appearances of collaterals (16,17). The digital discoloration associated with HHS also differs from the triphasic pattern of white-blue-red observed in Raynaud's. In a majority of HHS cases, symptoms occur only in the dominant hand, sometimes in both hands in 7% of cases (14). Therefore, bilateral involvement would suggest other causes. In addition, as HHS is linked to trauma from overuse of the hands, lower extremity symptoms are very unlikely, which are commonly seen in connective tissue disorders, vasculitis, and Buerger's. This entity should be kept in mind when a cardiac patient with certain occupational hazards has to undergo radial artery access for catheterization.

Treatment modalities for HHS focus on prevention of further trauma and restoration of ulnar arterial flow and range from percutaneous thrombin injection for ulnar artery aneurysm, which has similar success as for visceral arterial thrombin injection to surgical management with venous grafts (18). Due to the rarity of HHS diagnosis, there has yet to be sufficient evidence for a standard of care to be established. Conservative options include avoiding trauma to the hypothenar region which may require additional protection or change of occupation, smoke cessation, prescription of vasodilators, calcium-channel blockers, and antiplatelet therapy (9). Avoiding cold exposure is also recommended to minimize vasospasms and sympathetic aggravation. Surgical intervention may be indicated in addition to conservative management in severe cases for aneurysm repair and thrombi removal. Long-term follow-up of 12 patients who received vascular reconstruction following diagnosed with HHS showed persistent ulnar patency after in 75% of patients after 4.5 years with overall improvement of symptoms (19). The success of treatment will depend on the severity of ischemia, as well as the type of intervention and surgical procedure used. Until larger, prospective studies can take place to compare the efficacies of inventions, treatment of HHS will continue to vary case-by-case.

Conclusions

Clinical presentation of HHS includes Raynaud's phenomenon, pain, and tenderness to the digits and hypothenar region, and increased cold sensitivity. Especially at risk of developing HHS are individuals with a history of repetitive trauma to the hypothenar region of the palm, either occupational or recreational. Patients who present with these symptoms, along with a history working in but not limited to construction, auto repair, mining, carpentry, as well as those of athletic background, should be evaluated for the diagnosis of HHS. The gold standard diagnostic test is angiography which can confirm ulnar insufficiency due to aneurysmal dilation or thrombi formation. Clinical examination, angiographic findings, and patient history can distinguish HHS from other diseases with symptomatology. Correct diagnosis can ensure to proper and timely therapeutic response that can lead to improvement of symptoms and reverse ischemic changes. Although larger multicenter studies will be needed to determine the best treatment course, surgical interventions have been shown to have good outcomes in restoring patency to the ulnar artery. This entity should be kept in mind when a cardiac patient with certain occupational hazards has to undergo radial artery access for catheterization. Additionally, patients at risk of developing HHS given their background and history of tobacco use should be educated in primary care settings to improve hand protection and minimize preventable trauma.

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Footnote

Conflicts of Interest: Both authors have completed the ICMJE uniform disclosure form (available at http://dx.doi.org/10.21037/jxym.2017.10.01). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

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