

Hypothenar Hammer Syndrome in a Cyclist

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ABSTRACT

A case of hypothenar hammer syndrome with development of an ulnar artery aneurysm is reported in a recreational cyclist. The aneurysm was treated surgically and follow up Doppler ultrasound studies demonstrated persistent flow through the anastomosed artery. The pathology of the aneurysm is illustrated and the pathogenesis of the changes discussed.

Key Words: Hypothenar hammer syndrome; Guyon's canal; Cyclist; Pathology; Pathogenesis

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CASE REPORT

A 65 year old male presented with a mass in the region of the hypothenar eminence of the left hand (figure 1). The patient was an avid recreational cyclist who began cycling 40 years earlier and over the subsequent years accumulated in excess of 100,000 miles of cycling. The cycling was predominantly road cycling with a minimal amount of off road cycling. Although the patient always wore cycling gloves and the handle bar was wrapped, it was not padded. One year prior to presentation, he suffered a high speed bicycling accident with a fall on the right side and with abrasions on that side with a mild concussion. He did not recall particular injury to the left hand.

On physical examination, the lesion felt circumscribed and was just under 1 cm in size. The mass was moveable and did not pulsate. Although there was pain on pressure, the pain did not radiate in the distribution of the ulnar nerve. There were no sensory or motor deficits in the left hand. A CT scan was subsequently performed and interpreted as an ulnar nerve schwannoma located in Guyton's canal (Figure 2).

At surgery, an aneurysm of the ulnar artery was identified and resected. An end to end anastomosis of the ulnar artery was accomplished with good flow through the artery (Figures 3, 4).

The patient recovered without complications. The patient has taken aspirin, 80 mg/day following the surgery. A subsequent ultrasound and Doppler examination performed 14 months post-operatively showed persistent flow through the anastomosed ulnar artery (Figure 5).

Histologic evaluation of the resected aneurysm showed an aneurysm of the ulnar artery with central occlusive thrombosis (Figure 6). The wall of the artery showed circumferential attenuation. An elastic tissue stain demonstrated circumferential loss of the normal arterial wall elastic. A trichrome stain demonstrated circumferential fibrosis of the arterial wall. The occluded lumen showed a florid proliferation of endothelial cells highlighted by a CD 31 Immunohistochemistry stain. There was also a striking proliferation of myofibroblasts admixed with areas of mature collagenous fibrosis. The histologic features of the occlusive lesion are characteristic of a "Masson lesion". This lesion is well characterized as an exuberant form of organizing thrombus (Figure 7).

DISCUSSION

Hypothenar hammer syndrome (HHS) comprises a spectrum of pathologies in the ulnar artery with traumatic damage to the artery as the underlying etiology. The phenomenon was first described in a young factory worker in 1934^[1] for which a surgical resection of the thrombosed segment of the ulnar artery was performed and a pathologic examination was included. In 1970 the term “hypothenar hammer syndrome” was suggested by Conn, Bergan, Bell^[2] who proposed that repetitive microtrauma to the ulnar artery resulted in arterial wall damage.

HHS results from the unique anatomy of the ulnar artery as it passes through Guyon’s canal. Guyon’s canal, best regarded as a fixed space, is delineated by: (1) a palmar roof formed by the superficial antebrachial palmar fascia (palmar carpal ligament) together with fibers of the palmaris brevis muscle; (2) a radial edge where the roof tissues noted above attach to the flexor retinaculum



Figure 1 Left hand. A. Left hand with position of 1 cm mass circled.

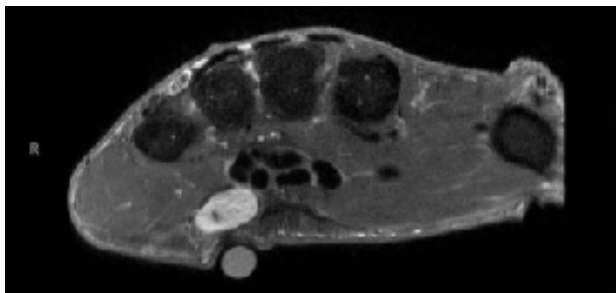


Figure 2 CT scan of left hand. CT scan with contrast demonstrates a well circumscribed lesion in Guyon’s canal which was initially interpreted as an ulnar nerve schwannoma.

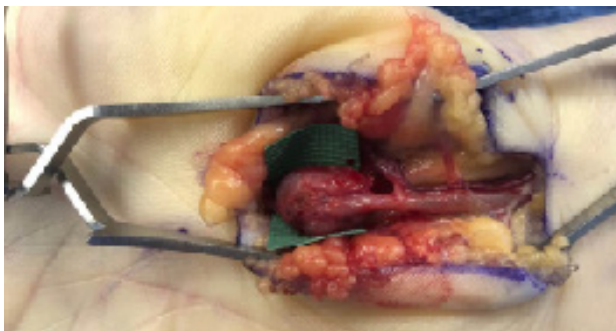


Figure 3 Left hand at surgery. Intraoperative demonstration of ulnar artery aneurysm.

(transverse carpal ligament); (3) a floor made up of the flexor retinaculum, a portion of the hook of hamate bone, and fibers from the muscles of the hypothenar muscle; and (4) an ulnar margin formed by the attachment of the fascial roof of the canal with the hypothenar muscle proximally and with the pisiform bone distally

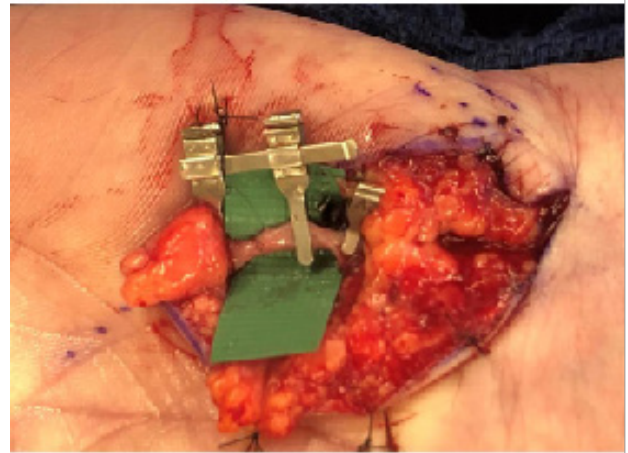


Figure 4 Left hand at surgery. Intraoperative demonstration of post-resection anastomosis of ulnar artery.

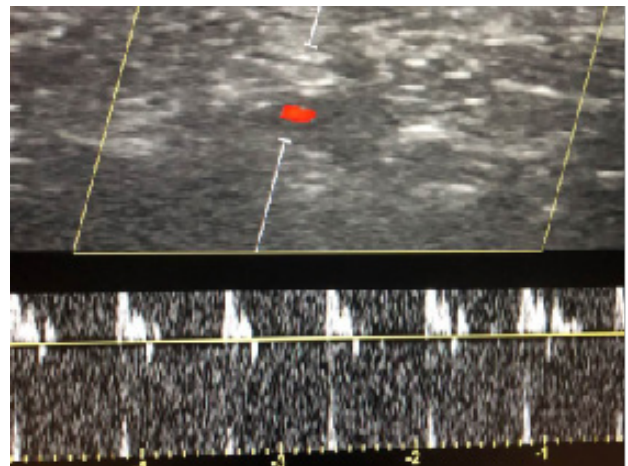


Figure 5 Ultrasound Doppler study of anastomosed ulnar artery. Red area shows flow through the anastomosed ulnar artery. Doppler study shows triphasic pattern, characteristic of flow through a high resistance vessel.

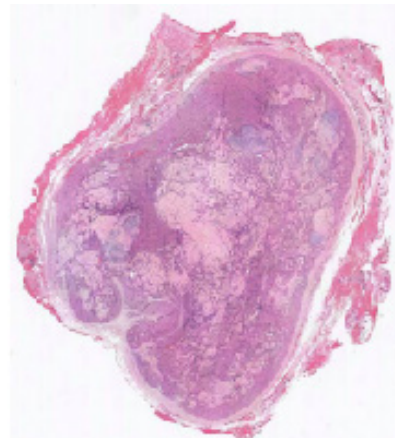


Figure 6 Scanned image of histologic section of ulnar artery aneurysm. This scanned image demonstrates dilated attenuated arterial wall with luminal thrombosis. There are areas of endothelial and myofibroblastic proliferation admixed with areas of dense collagenous fibrosis.

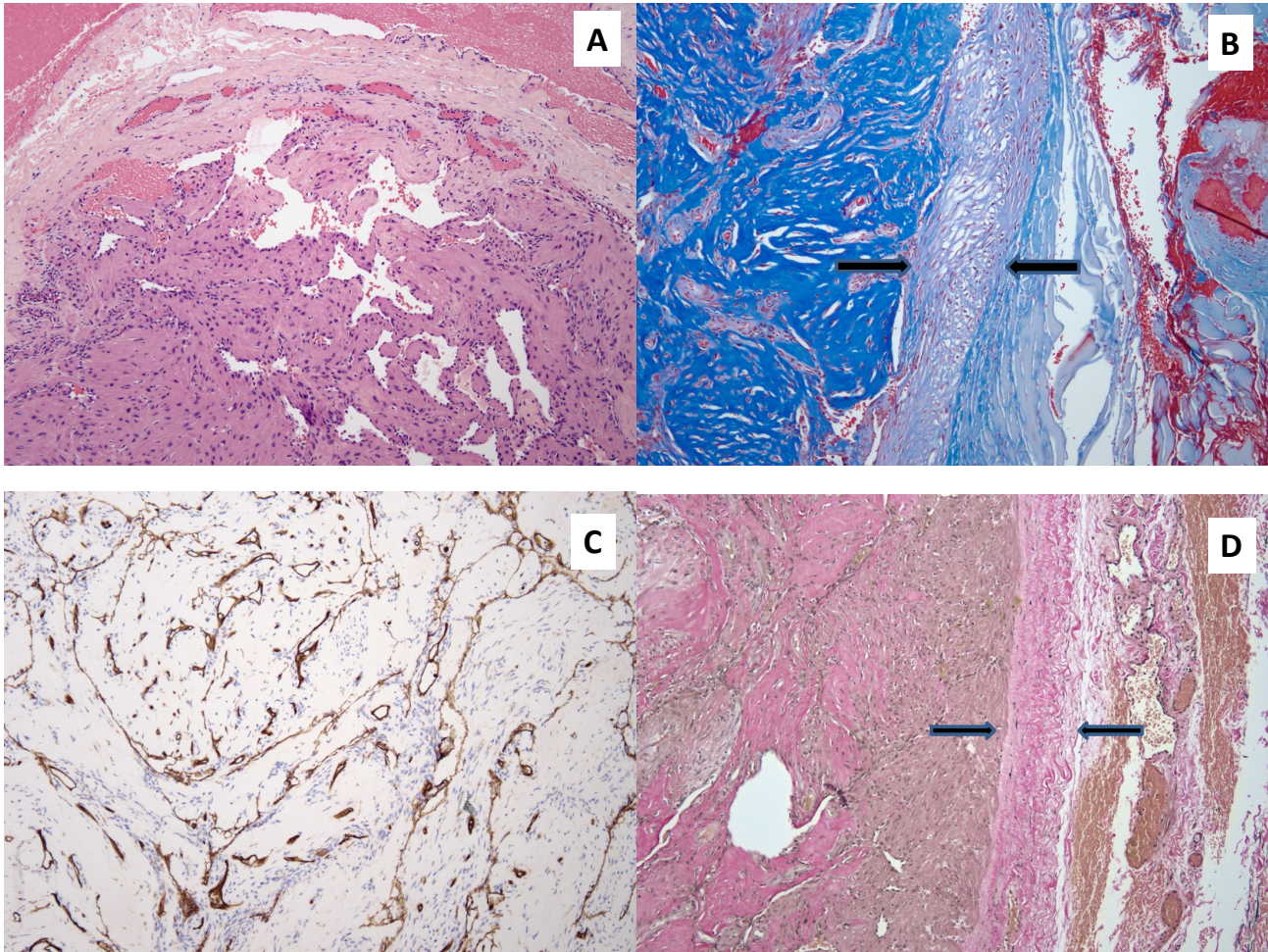


Figure 7 Photomicrographs of aneurysm of ulnar artery with thrombosis.

A. Intravascular area of endothelial proliferation with associated myofibroblastic proliferation. Hematoxylin and eosin stain; 100 x magnification.

B. Shows attenuated wall of vessel with collagenous fibrosis of wall (between arrows). Lumen contains areas of dense collagenous fibrosis and areas of endothelial proliferation. Trichrome stain; 100x magnification.

C. Immunohistochemical stain shows capillary proliferation highlighted by CD 31 IHC stain. Proliferating endothelial cells are admixed with myofibroblastic proliferation. Immunohistochemical stain for CD 31; 40 x magnification.

D. Attenuated wall of vessel shows loss of normal elastic fibers of the media (between arrows). EVG stain; 100 x magnification.

in the canal. The superficial branch of the ulnar artery crosses the surface of the hypothenar muscle where its superficial location makes it susceptible to traumatic injury^[3,4]. Specifically, the ulnar artery is trapped between the applied blunt force trauma to the palm and the relatively firm and unyielding surface of the hook of hamate bone. (Figure 8).

Repetitive microtrauma causes damage to the intima and vascular wall and results in the following pathologies: (1) damage to the intimal layer of the artery leads to recruitment of inflammatory mediators, aggregation of platelets, and vasospasm; (2) a more serious complication, arterial thrombosis may occur causing distal arterial ischemia which can also be associated with embolism to the digital arteries; and (3) cumulative damage to the vessel wall results in weakening of the vessel wall with a loss of elastic, fibrosis of the wall, and subsequent dilatation of the artery with aneurysm formation, with or without thrombosis of the aneurysm.

Clinical presentation classically includes pain at the hypothenar eminence. Other symptoms include digital pain with paresthesia, cold sensitivity, phasic blanching, discoloration of the fingertips, or finger claudication. The fifth digit is the most commonly involved though the third and fourth digits are often involved as well. Symptoms are unilateral and spare the thumbs, distinguishing HHS from Raynaud's phenomenon^[6]. If an aneurysmal segment is present, a pulsatile

palpable mass may be found.

Physical exam for HHS is often significant for hypothenar callus, tenderness, or mass (if aneurysm formation is present). Allen test is an important test that assesses the patency of the superficial palmar arch. In this test the radial artery and ulnar artery is compressed resulting in a blanched appearance of the hand. The ulnar artery is then released and the return of normal palm color is observed which should occur within 5 seconds. If the ulnar artery is occluded, palmar color will not return until the radial artery is released. Though the Allen test is widely used, Kaji^[7] found that 14% of patients with HHS had a negative Allen test and Mousavi^[8] found that 22% of healthy patients had a positive Allen test. Thus, the Allen test appears to be neither sensitive or specific.

Diagnosis of HHS can be confirmed with Doppler by showing ulnar artery thrombosis or aneurysm. However, the gold standard is angiography. Pathognomonic findings on angiography include ulnar artery tortuosity with corkscrew appearance, aneurysm, occlusion of the ulnar artery segment over the hook of hamate, occluded digital arteries in an ulnar artery distribution, and intraluminal emboli at sites of digital obstruction^[9]. Angiography is the gold standard because it can exclude subclavian artery embolic events, confirm the exact location of the vascular damage at the end of the ulnar artery, determine thrombosis vs aneurysm, and evaluate embolic events in

the digital arteries. It can also be useful in detecting early arterial wall thickening. The retrograde femoral Seldinger technique is preferred by several authors^[10,11] since it allows for better visualization of the subclavian and aortic arch. It is also safer in the rare case of an arteriographic complication.

Hypothenar hammer syndrome is a challenging diagnosis despite several publications concerning the disease. Differential diagnosis includes embolic/proximal vascular disease as well as local disease with or without associated systemic disease. Proximal etiologies include cardioembolic causes and large vessel disease such as atherosclerotic plaque and arterial thoracic outlet syndrome^[12]. Local etiologies with systemic manifestations include atherosclerotic disease, vasculitis, Raynaud's disease, connective tissue disease. Local etiologies without systemic manifestations include benign tumors of the hand such as lipomas, schwannomas, and vascular malformation^[13].

Many medical and surgical treatment options exist for hypothenar hammer syndrome. Treatment selection varies based on the level of ischemia present in the patient. For segmental occlusions without significant ischemia, nonoperative treatments are often sufficient. These include avoidance of trauma, smoking cessation, analgesia, calcium channel blockers, steroids, antiplatelet or anticoagulant agents, and pentoxifylline to reduce blood viscosity^[14]. Surgical intervention is usually indicated for patients with critical ischemia including intractable pain, vascular damage to multiple digital arteries and tissue loss. Surgical options include artery ligation, resection of thrombosed segment of the artery with end-to-end anastomosis, and resection and vascular reconstruction with graft vessel. Treatment strategies of HHS are unclear due to limited studies currently available. Many believe though that recovery of function is more likely with earlier aggressive and definitive treatment^[9]. In addition, surgical treatment will decrease the potential for serious thromboembolic complications in the fingers. In our case, the treatment strategy of resection and anastomosis was chosen. This selection was successful, evidenced by good flow in the ulnar artery observed on Doppler ultrasound study during post surgical follow up.

HHS is usually found in men of working age, mean of 40 years old^[10], involving the dominant hand with occupations involving repetitive blunt trauma to the hands. This includes workers who use tools such as hammers and those who push or squeeze hard objects. Metal workers, auto mechanics, lathe operators, machinists, miners, sawmill workers, butchers, bakers, brick layers, and carpenters are at most risk. HHS has also been reported in baseball, karate, badminton, mountain biking, golf, tennis, handball, volleyball, softball, weight lifting, break dancing and hockey^[9].

To our knowledge this is the first pathologically documented case of HHS occurring in a cyclist without any other occupational risk factors. In our case, it is likely that cycling on roads that are variably uneven caused vibrational forces to travel up the handlebars which were ultimately absorbed by the palms. All of the common hand positions assumed by our cyclist while riding resulted in direct pressure on the structures in Guyon's canal (Figure 9).

Cycling over a long period of time resulted in accumulation of microtraumas to the ulnar artery, causing intimal wall damage and medial wall fibrosis. Although HHS is generally thought of as a rare disease, it may be more common than generally described. For instance, HHS was found in 7% of 330 vibration exposed workers^[7]. Although HHS may be more prevalent than thought, many clinicians are unfamiliar with the etiology and presentation of HHS and consequently, HHS is often incorrectly diagnosed^[9,15]. When assessing a patient with HHS, careful history taking is important to

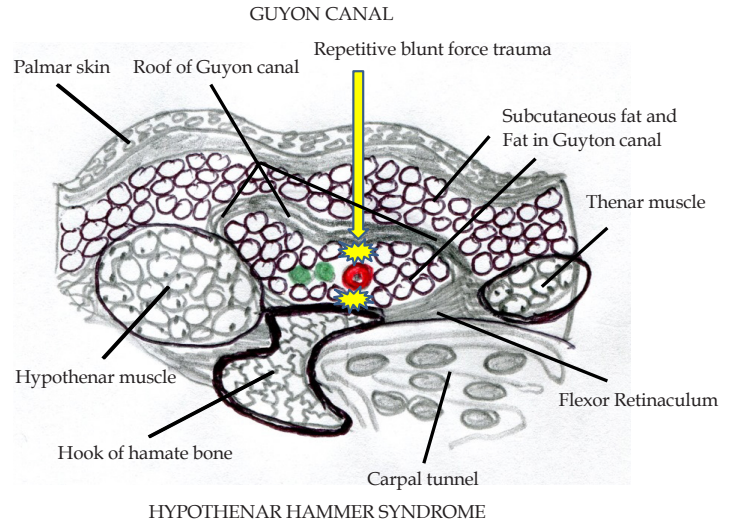


Figure 8 Diagrammatic sketch of Guyon's canal with pathogenesis of hypothenar hammer syndrome. Repetitive blunt force trauma to the palmar surface of the hand compresses ulnar artery against the hook of hamate bone in the floor of the Guyon canal. Ulnar artery shown in red, ulnar nerve in green. (Sketch adapted from photomicrographs of Cobb, Carmichael, Cooney^[6])

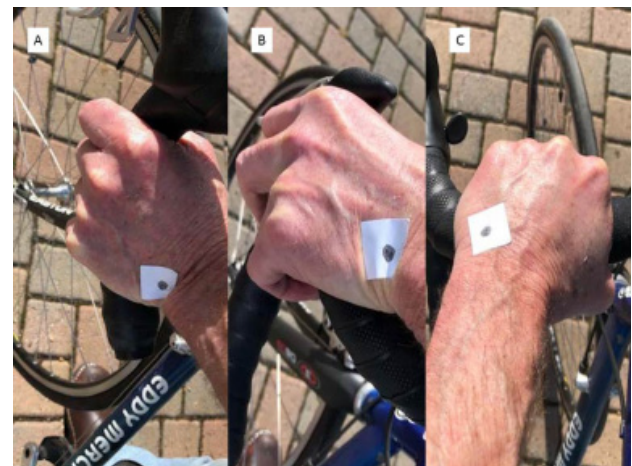


Figure 9 Most common hand positions while riding bicycle. Black dot on white rectangle marks area where plumb line dropped through hand intersects Guyon's canal with ulnar artery aneurysm. A) Hand position riding in "drops" of handlebar. B) Hand position riding gripping hoods of the brakes. C) Hand position on horizontal crossbar portion of handlebar. Note: In all 3 positions there is direct pressure on the structures in Guyon's canal.

identify occupational or recreational exposures to trauma that may cause vascular microtrauma to the ulnar artery. Prevention for our patient included the use of well padded gloves along with padded handlebars in order to dampen the force traveling to the ulnar artery.

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