Tortuous peripheral arteries: a cause of focal neuropathy

Case report

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Segmental ulnar neuropathy has been reported as a result of ulnar nerve compression due to posttraumatic false aneurysms of the ulnar artery and, more infrequently, due to thrombosis or true aneurysms of the same vessel. The authors present a case of segmental sensory ulnar neuropathy in the wrist which intraoperatively demonstrated impingement on the ulnar nerve by a tortuous ulnar artery. The symptomatic relief and electrophysiological improvement that occurred immediately following neurovascular decompression confirm that the vascular anomaly was the cause of neuropathy. Pulsatile pressure applied to the nerve trunk may have triggered ectopic stimulation of sensory fibers manifested by a tingling and burning sensation. There was immediate resolution of paresthesia following mobilization of the impinging vessel from the nerve. Subsequent rapid electrophysiological recovery may be explained by improvement in focal nerve microcirculation following vascular decompression. Tortuosity (megadolichoectatic anomaly) of intracranial arteries has been related to cranial nerve or brain-stem dysfunction; however, this appears to be the first report in the literature of a case in which such association has been found to occur extracranially, involving a peripheral nerve.

KEY WORDS • ulnar nerve • vascular compression • ulnar artery • wrist entrapment neuropathy

Entrapment of peripheral nerves at strategic appendicular sites (such as the cubital tunnel, carpal tunnel, Guyon's canal, or tarsal tunnel) is a frequent cause of focal neuropathy. Although most commonly idiopathic, entrapment can also result from compression of the nerve by osteoarthritic processes, soft-tissue tumors, synovial cysts, and bursal enlargement; for the upper extremities, a combined compression of nerve and blood vessels can be found as well. Unlike cranial nerves, which have been reported to be compressed by elongated, tortuous, and dilated arteries (megadolichoectatic anomaly), only aneurysmal or thrombosed ulnar arteries have been identified as impinging on peripheral nerves.

Our report focuses on a case of compression of the ulnar nerve by a tortuous ulnar artery at the distal forearm. We review the concept of pulsatile compression of nerves and, based on our clinical experience, we propose to include impingement by arterial vessels in the list of etiological factors of peripheral nerve compression.

Case Report

This 62-year-old man was referred to our neurosurgery clinic with a 2½-year history of intermittent tingling, numbness, and burning sensation along the hypothenar eminence and the fourth and fifth digits of his dominant right hand. During these short-lived paroxysms (lasting a few minutes, but never longer than 10 minutes) the skin on the affected area would become painful to the slightest touch. The patient compared his symptoms with the feeling of "a freezing hand immersed in hot water." His medical history was remarkable for a Jeep injury sustained in World War II, with a right-hand fracture and ulnar nerve palsy becoming apparent shortly thereafter. This resolved without operation.

Examination. Examination elicited no tenderness at the cubital tunnel; however, it revealed a 1 × 1-cm tender, elastic, nonmobile, nonpulsatile mass, proximal to the creases on the volar aspect of the wrist. No Tinel's sign was elicited. There was slight diminution of the hypothenar eminence muscle mass, with mild weakness of the fifth digit abduction. There was no weakness of the interosseous muscles. Sensation to touch and pinprick was diminished in the ulnar distribution of the hand.

X-ray films of the wrist and hand revealed old fractures of the base of the first and second metacarpal bones, and degenerative radiocarpal joint disease. Elec-
trodiagnostic studies (nerve conduction velocity) did not demonstrate ulnar nerve segmental delay across the elbow or distally; however, a sensory nerve action potential (SNAP) study of the ulnar nerve did reveal a significantly prolonged distal latency of 5.8 m/sec (normal 1.6 to 3.1 m/sec) and a decreased amplitude of 6.9 μV (normal 15 to 50 μV) (Fig. 1). Electromyography showed minimal chronic neuropathic changes in the hypothenar muscles.

Operation. Exploration of the ulnar nerve at the wrist was performed utilizing magnifying loupes and intraoperative nerve stimulation. An 8-cm longitudinal incision was made between the flexor carpi ulnaris and the palmaris longus tendons and was extended to a point just proximal to the wrist crease. A bluish nonpulsatile mass was immediately apparent in the subcutaneous plane proximal to Guyon's canal. Following dissection of the fibrous superficial planes of the mass, an S-shaped ulnar artery protruded ventrally into the operative field as a "box-spring," exposing underneath it an impinged ulnar nerve (Fig. 2 upper left). Each time the ulnar artery was mobilized radially to de-press the nerve, a small branch running perpendicular and medially to it would stretch over the nerve (Fig. 2 upper right). This branch was severed and the ulnar artery was tacked away radially from the nerve, by suturing the fibrous periadventitial tissue with the fascia superficialis. The ventral aspect of the nerve was indented corresponding to the niche left by the arterial vessel impingement (Fig. 2 lower).

Postoperative Course. Following the operation, the patient had no more episodes of tingling, numbness, and burning sensation. Examination demonstrated good sensation to pinprick and touch on the ulnar aspect of the hand. An electrodiagnostic study on the 10th postoperative day already revealed improvement in the distal latency of ulnar nerve SNAP (3.0 m/sec) which was sustained at 4 weeks (3.2 m/sec) and 6 weeks (3.4 m/sec), as illustrated in Fig. 1. The patient was referred to occupational therapy and, at 6 weeks postoperatively, was again able to do carpentry.

Discussion

Arterial enlargement, elongation, and dilatation, which may include vessel tortuosity, have been specifically described in relation to the intracranial vessels. The vertebrobasilar system is more commonly affected, the term "megadolichoaneurysm" being coined in 1964 by Boeri and Passerini to describe enlargement and elongation. The term "dolichoectasia" (enlargement and fusiform dilatation) was used later regarding any involved intracranial artery. Kerber, et al., described symptomatic "tortuous vertebrobasilar systems" (just elongation). Defects in the internal elastic lamina and reticul fiber deficiency in the muscular layer of the vessel wall have been the common features on microscopic examination, and Hegedin suggested that they (not atherosclerosis) constitute the morphological basis of ectasia. Furthermore, the presence of this arterial anomaly in children and in adults without histological trace of atherosclerosis suggests an alternative cause for the putative elastic lamina defect. In addition, in atherosclerotic aorta walls, increased collagenase activity was found only in the areas of aneurysmal dilatation. Our patient, aged in his late 60's, did not have hypercholesterolemia, a history of vascular disease, or hypertension. At surgery, the magnified view of the ulnar artery (as seen in Fig. 2) did not indicate atheromatous changes.

Vascular compression of cranial nerves has been identified as a causal factor in numerous clinical syndromes. Trigeminal neuralgia, hemifacial spasm, acoustic nerve dysfunction, and glossopharyngeal neuralgia have all been relieved by microvascular decompression near the entrance of the cranial nerves into the brain stem. Regarding peripheral nerves, a good number of reports describe compression of the ulnar nerve due to posttraumatic false aneurysms of the ulnar artery of the wrist and hand. This pathology has also been reported with thrombosis of the ulnar artery as it emerges from Guyon's canal, usually due to repetitive occupational trauma and, more rarely, as a result of acute blunt trauma or anomalous arterial anatomy. Very infrequently, compression by true ulnar artery aneurysms such as arteriosclerotic or thrombosed cirsoid aneurysms has been described. However, symptomatic "megadolichoectasia" anomaly of peripheral arteries has not been reported. Release of two
fibrovascular bands coursing from the ulnar artery to the distal belly of the flexor carpi ulnaris muscle, entrapping and grooving the ulnar nerve, resulted in the reversal of electrical block, complete relief of pain, and a full neurological recovery during the ensuing 6 months in a 74-year-old patient reported by Holtzman, et al.\textsuperscript{12} Unlike our patient, no enlargement or tortuosity of the artery was found.

The delayed onset of sensory ulnar neuropathy in our patient (45 years) makes it difficult to invoke the right-hand blunt trauma he sustained in his World War II Jeep accident as a direct etiological factor. We may not be able to rule out a pathogenic relationship between the trauma and the development of tortuosity of the ulnar artery. However, the radiographs of the wrist and hand only revealed old fractures of the proximal second metacarpal and base of the first metacarpal bones, while no signs of skeletal injury in relation to the course of the ulnar artery were found.

The rapid improvement in sensory latency to 3.0 m/sec as early as the 10th postoperative day and maintained at 12 weeks indicates a mild compression (first degree nerve injury)\textsuperscript{33}). Nevertheless, this classification is based on models of transient and prolonged experimental cuff nerve compression\textsuperscript{9,36} We have studied and reported physiological changes associated with acute and chronic experimental models of pulsatile compression of the ninth, 10th, and 11th cranial nerves,\textsuperscript{28-30} but, to the extent of our knowledge, no models of pulsatile nerve compression of peripheral nerves have been developed. The clinical data available regarding vascular compression of peripheral nerves comes from combined neurovascular injuries resulting in posttraumatic aneurysms. This makes it very difficult to ascertain whether a primary traumatic nerve injury, ischemia due to vessel injury, or progressive pressure from a subsequently enlarging aneurysm was responsible for the neuropathy. These data indicate that posttraumatic aneurysms are associated from the time of injury with severe pain, sometimes causalgic; it may worsen with further enlargement of the aneurysm, and motor function is usually preserved.

We may assume that in our patient, the sharp 90\degree double-bent S-shaped but patent ulnar artery impinged on the ventral aspect of the ulnar nerve for at least the 2½ symptomatic years. The constant “hammering” of the nerve at the pulse rate may explain the most prominent symptom: paresthesia. Immediate resolution of the tingling and burning sensation by simply moving aside the pulsatile pressure suggests that this may have triggered ectopic activation of sensory nerve fibers. Improvement in focal nerve micrcirculation by the neurovascular decompression could not be ruled out either. The funicular arrangement of the ulnar nerve at the distal wrist consists of cutaneous fibers occupying the anterolateral portion of the nerve, while the muscular fibers are located posteromedially.\textsuperscript{31} The vascular compression in our patient was anterolateral and this would explain why he had no motor deficit.

Acknowledgments

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References