Ulnar nerve compression in the proximal forearm by an intraneural arteriovenous malformation

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Compression of the ulnar nerve is most commonly caused by ganglions, lipomas, anomalous tendon and muscles, trauma related to an occupation and arthritis in the upper limb; however, nerve compression in the upper limb and, more importantly, in the proximal forearm by a vascular lesion is rare. A rare case of ulnar nerve compression in the proximal forearm by an arteriovenous malformation in a 45-year-old man who presented with six-year history of gradually increasing swelling in medial aspect of proximal forearm is reported. After excision of the tumour, it was found to be an arteriovenous malformation.

Key Words: Arteriovenous malformation; Forearm; Ulnar nerve

Chronic nerve entrapment syndromes in the upper extremity are common and their occurrence is likely to increase as risk factors such as diabetes, obesity and advanced age become more prevalent in the general population. Ganglions, lipomas, anomalous tendon and muscles, trauma related to an occupation and arthritis can cause ulnar nerve compression in the upper limb. However, ulnar nerve compression in the upper limb and, more importantly, in the proximal forearm by a vascular lesion is rare.

Ulnar artery aneurysm, tortuous ulnar artery, hemangioma and thrombosis have been reported in the literature as vascular lesions. In most cases, the diagnosis can be readily made on the basis of history and physical examination. For more atypical presentations, consideration should be given to other neurological conditions that mimic nerve entrapment syndromes such as mononeuritis, Parsonage-Turner syndrome and motor neuropathies; not only will these conditions not be properly considered, but they may exacerbate the symptoms.

CASE PRESENTATION

A 45-year-old man presented with six-year history of gradually increasing swelling in the medial aspect of proximal forearm, approximately 4 cm below the medial epicondyle, with tingling sensation of fourth and fifth digit in his right hand. He had no history of diabetes mellitus and other medical history was unremarkable. A clinical examination revealed paresthesia and hypesthesia of the fifth digit and medial region of the fourth digit. Hypothermia atrophy and weakness of the hypothenar muscles were also noted. A thorough history was taken including the onset of symptoms, presence of grip or pinch weakness, numbness, aggravating and alleviating activities, comorbidities (ie, diabetes, peripheral neuropathies) and previous elbow trauma. The single most important feature on history, however, was the chronicity of the symptoms.

A sensory conduction study of the right ulnar nerve was normal; however, a motor conduction study of the right ulnar nerve from the elbow and wrist to the abductor digiti minimi muscle revealed a small amplitude. There was also an abnormal spontaneous activity in the flexor carpi ulnaris. These electrophysiological findings were consistent with an ulnar nerve lesion at or around the proximal forearm or elbow. Magnetic resonance imaging was not performed because of the strong suggestion of ulnar nerve compression at the proximal forearm according to clinical presentation and electrophysiological studies.

A presumptive diagnosis of ulnar nerve compression at the proximal forearm was made and it was decided to explore the ulnar nerve in the proximal forearm under general anesthesia. Under microscopic magnification, the ulnar nerve was explored and a vascular anomaly encompassed by the nerve tissue was found. A prominent vascularity suggestive of a vascular malformation was apparent on exploration of the ulnar nerve. Engorged, purple veins as well as small arterial feeders were noted, which were affecting the ulnar nerve. One prominent draining vein adjacent to the nerve fascicles was resected. The overlying nerve was thinned and compressed by the arteriovenous malformation (Figure 1). External neurolysis as well as limited internal neurolysis of the ulnar nerve was undertaken.

Histological examination revealed the lesion to be an arteriovenous malformation (Figure 2). At six-month and one-year follow-ups, the patient’s pain had improved. He continues to use over-the-counter nonsteroidal anti-inflammatory agents for pain control on an as-needed basis. His motor and sensory examinations were normal at the six-month follow-up.

DISCUSSION

The elbow is the most common site for ulnar nerve compression (1,2). The etiology of ulnar nerve compression at the proximal forearm includes compression, traction and repeated irritation, lipoma, vascular anomalies, bone fractures, ulnar artery disease, anomalous tendon and muscles, and trauma related to an occupation (3-7).

The region of the nerve affected by compression is most commonly diagnosed based on patient-reported symptoms and clinical signs because of the morbidity associated with nerve biopsy; current understanding of the histopathological changes associated with nerve entrapment syndromes is derived from animal model studies. A consistent finding in these studies is that the severity of pathological changes is dependent on the magnitude and duration of compression. An initial breakdown in the blood-nerve barrier is observed, followed by endoneurial edema. Along with perineural thickening, this edema may lead to the deposition of Renaut bodies within the substance of the nerve, further increasing endoneurial pressure. Increased endoneurial pressure disrupts the microvascular circulation, thereby inducing dynamic ischemia in the nerve.

Depending on systemic factors as well as the location and degree of compression, patients with nerve entrapment syndromes can present...
with a constellation of symptoms. Given the wide variability in symptoms, numerous attempts have been made at creating standardized diagnostic approaches to these conditions.

Shea and McClain (8) classified ulnar nerve lesion into three types according to anatomical location. Type I is motor and sensory abnormalities with the lesion of Guyon’s canal or proximal to it (30%). Type II is motor abnormalities with weakness of the intrinsic muscle (52%). Type III is sensory abnormalities on the palmar surface of the hypothenar eminence and in the fourth and fifth digit (18%). Our case corresponds to type I according to the Shea and McClain classification.

After clinical suspicion of ulnar nerve compression, although nerve conduction tests can diagnose exact location of nerve involvement, magnetic resonance imaging should be performed for accurate diagnosis.

Although many causative lesions of ulnar nerve compression in proximal forearm have been reported in the literature, vascular lesion should be considered as a possible etiology.

CONCLUSION

We report a rare case of ulnar nerve compression in the proximal forearm caused by an arteriovenous malformation. Although many etiologies may cause ulnar nerve compression in the forearm, vascular lesion should be considered as a possible etiology.

REFERENCES

1. Erkin G, Uysal H, Keles I, Aybay C, Ozel S. Acute ulnar neuropathy at the wrist: A case report and review of the literature. Rheumatol Int 2006;27:191-6.
2. Papathanasiou ES, Loizides A, Panayiotou P, Papacostas SS, Kleopa KA. Ulnar neuropathy at Guyon’s canal: Electrophysiological and surgical findings. Electromyogr Clin Neurophysiol 2005;45:87-92.
3. Aguilar PH, Bov-Seng-Shu E, Gomes-Pinto F, et al. Surgical management of Guyon’s canal syndrome, an ulnar nerve entrapment at the wrist: Report of two cases. Arq Neuropsiquiatr 2001;59:106-11.
4. Bozkurt MC, Tagil SM, Orçakar L, Ersoy M, Tekdemir I. Anatomical variations as potential risk factors for ulnar tunnel syndrome: A cadaveric study. Clin Anat 2005;18:274-80.
5. Erkin G, Uysal H, Keles I, Aybay C, Ozel S. Acute ulnar neuropathy at the wrist: A case report and review of the literature. Rheumatol Int 2006;27:191-6.
6. Jose RM, Bragg T, Srivastava S. Ulnar nerve compression in Guyon’s canal in the presence of a tortuous ulnar artery. J Hand Surg Br 2006;31:200-2.
7. Koch H, Haas F, Pierer G. Ulnar nerve compression in Guyon’s canal due to a haemangioma of the ulnar artery. J Hand Surg Br 1998;23:242-4.
8. Shea JD, McClain EJ. Ulnar-nerve compression syndromes at and below the wrist. J Bone Joint Surg Am 1969;51:1095-103.