Cubital Tunnel Syndrome Caused by Ulnar Nerve Schwannoma in a Patient with Diabetic Sensorimotor Polyneuropathy

Meng-Ta Liu[1], Jiunn-Tay Lee[2], Chih-Hsin Wang[3], Yu-Chieh Lin[4,5], Chung-Hsing Chou[2]

Abstract-

Purpose: An uncommon ulnar nerve schwannoma should not be neglected in a diabetic patient with cubital tunnel syndrome, the second most common cause of entrapment neuropathy.

Case Report: A 61-year-old woman with a past history of type 2 diabetes mellitus complicated with sensorimotor polyneuropathy presented with progressive numbness and weakness of the left ring and little fingers for one year. A provisional diagnosis of cubital tunnel syndrome was made according to physical examination and electrophysiological studies. The magnetic resonance imaging showed a nodular lesion over ulnar aspect of the left elbow, which was demonstrated to be a schwannoma by histopathology. The patient had moderate improvement after surgical decompression of the left cubital tunnel.

Conclusion: This case illustrates the heterogeneous group of pathologies causing peripheral neuropathy. The diagnosis of ulnar nerve schwannoma with cubital tunnel syndrome, superimposed with diabetic sensorimotor polyneuropathy, was made carefully according to clinical manifestations as well as a series of electrophysiological, imaging, and pathological studies.

Key Words: cubital tunnel syndrome, ulnar nerve, nerve compression, schwannoma, diabetes

Acta Neurol Taiwan 2016;25:60-64

INTRODUCTION

Cubital tunnel syndrome (CUTS) is the second most common peripheral nerve entrapment syndrome in the human body, after carpal tunnel syndrome resulting from median nerve entrapment[(1)]. An underlying mass causing nerve entrapment should not be overlooked in diabetic patients since diabetes makes a nerve more vulnerable to compression due to local ischemia and interference with the innate metabolism of the nerve[(2)]. A series of

From the [1]Department of Internal Medicine, Hualien Armed Forces General Hospital, Hualien, Taiwan; [2]Department of Neurology, Tri-Service General Hospital, National Defense Medical Center, Taipei, Taiwan; [3]Division of Plastic Surgery, Department of Surgery, Tri-Service General Hospital, National Defense Medical Center, Taipei, Taiwan; [4]Department of Pathology, Tri-Service General Hospital, National Defense Medical Center, Taipei, Taiwan; [5]Department of Pathology, Taoyuan Armed Forces General Hospital, Taoyuan, Taiwan.
Received May 16, 2016.
Revised & Accepted June 14, 2016.

Correspondence to: Chung-Hsing Chou, MD. Department of Neurology Tri-Service General Hospital National Defense Medical Center, No.325, Sec.2, Chenggong Rd., Neihu District,Taipei City 11490, Taiwan.
E-mail: choutpe@yahoo.com.tw

electrophysiological examinations play as an assessment tool for diagnosis making and outcome prediction. We here describe an uncommon ulnar nerve schwannoma as the cause of cubital tunnel syndrome in a diabetic patient with sensorimotor polyneuropathy who got partial recovery after surgical decompression.

CASE REPORT

A 61-year-old, right-handed, Taiwanese female presented with progressive numbness and weakness of the ring and little fingers in the left hand for one year. She had a history of type 2 diabetes mellitus (DM) for 12 years, complicated with sensorimotor polyneuropathy diagnosed 5 years ago. A recent glycated hemoglobin A1c level was 9.2%. Clinical examination demonstrated wasting of the hypothenar and intrinsic muscles causing claw hand deformity, with positive Froment's sign, and involuntary abduction of the little finger, the Wartenberg's sign. Tinel's sign was positive at the left elbow, with a score 2 on the McGowan grading system for ulnar nerve neuropathy. The range of motion of the left elbow was not limited.

T1- and T2-weighted magnetic resonance imaging (MRI) documented a well-defined nodular lesion about 1.3 × 1.3 × 1.8 cm over ulnar side of the left elbow, suggestive of a neurogenic tumor (Figure 1A and 1B). The nerve conduction velocity and electromyographic studies indicated left ulnar and median sensorimotor neuropathy, with prolonged distal latency, reduced amplitude, and significant reduction in conduction velocity over the cross-elbow segment of the left ulnar compound motor action potentials (CMAPs), reduced amplitude of left median and radial CMAPs, as well as reduced conduction velocity and reduced amplitude of the left ulnar sensory nerve action potentials (SNAPs), and prolonged distal latency, reduced conduction velocity, and reduced amplitude of left median SNAPs. Increased duration and amplitude of motor unit potential of the left flexor carpi ulnaris (FCU) and the left abductor pollicis brevis (APB) muscles were recorded (Figure 2), suggesting the presence of conduction block of the ulnar nerve proximal to the stimulation site[3].

The patient underwent surgical decompression of the left cubital tunnel by microscopic neurolysis with excision of neuroma and fat graft of the left elbow. The excised mass was measured 1.3 × 1.3 × 1.5 cm (Figure 3A and

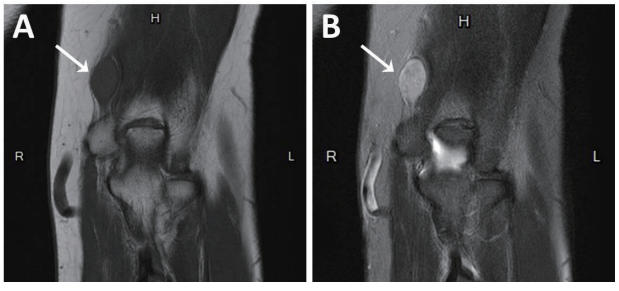

Figure 1. (A) T1-weighted spin echo and (B) T2-weighted fat-suppression fast spin-echo magnetic resonance imaging revealed a well-defined nodular lesion about 1.3 × 1.3 × 1.8 cm over ulnar aspect of the left elbow. H = the humerus.

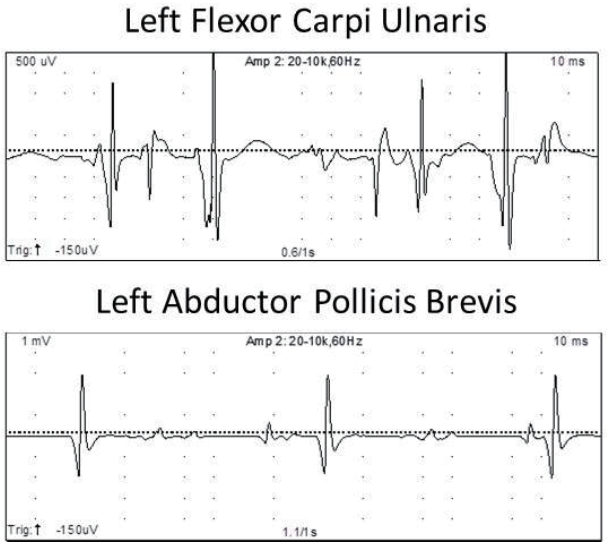

Figure 2. Increased durations and amplitudes of compound motor action potential of the left flexor carpi ulnaris and abductor pollicis brevis.

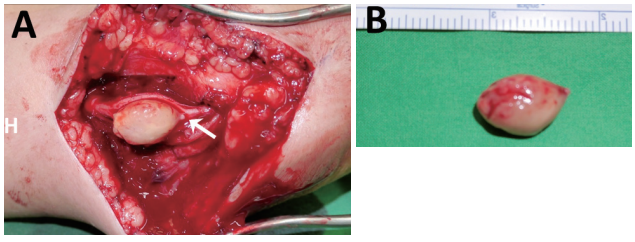

Figure 3. (A) Intraoperative view of the tumor inside the epineurium of the ulnar nerve. (B) The solid mass was 1.3 × 1.3 × 1.5 cm in size. H = the humerus.

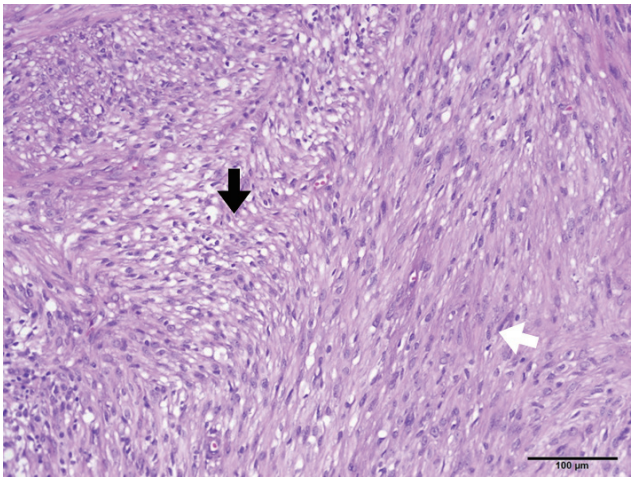

Figure 4. The tumor was composed of cellular spindle cells arranged into intersecting fascicles (Antoni A, black arrow) and hypocellular (Antoni B, white arrow) elements, indicating benign schwannoma. (hematoxylin and eosin stain 200X)

3B). Histopathology confirmed schwannoma, with Antoni A and Antoni B regions (Figure 4). The patient underwent rehabilitation and had moderate improvement in numbness and weakness of the fingers. Electromyographic study showed normal duration and amplitude of the sampling muscles with improved nerve conduction velocity of the left ulnar nerve at the postoperative 6-month follow up.

DISCUSSION

Focal mononeuropathies, depending on the distribution, are believed to be more frequent in diabetic patients. In a study of consecutive outpatients, for example, limb mononeuropathies, such as radial, ulnar and peroneal nerve paralysis, were more frequent in diabetic patients than in the general population, but the frequency of symptomatic carpal tunnel syndrome (CTS) and Bell's palsy was not higher in diabetic patients[4]. Once the peripheral neuropathy involves isolated damage to more than two separate nerve areas, it is defined as mononeuritis multiplex that consists of painful, asymmetrical, asynchronous sensory and motor components. Indeed, distribution of altered sensation or weakness due to diabetic neuropathy and its exact etiology may vary between individuals. Diabetes duration, poor metabolic control, and presence of microvascular complications have been proposed to be contributing factors of peripheral neuropathy[5,6]. The peripheral nerve is vascularized by a longitudinal vascular network with many anastomoses, allowing the blood flow to continue when segmental arteries are occluded. A major impairment in blood flow is therefore required to produce experimental ischemic nerve injuries, and these studies of experimental diabetes demonstrated the increased susceptibility of diabetic nerves to injury[7,8]. The abnormal susceptibility of diabetic nerves to compression could be accordingly attributable in part to chronic ischemic injuries.

The manifestations of a diabetic hand may not just be ulnar neuropathy. The definition of diabetic hand traditionally consists of limited joint mobility, Dupuytren's contracture and trigger finger[9]. Stenosing flexor tenosynovitis (SFT) and CTS were further included into the spectrum of diabetic hand syndrome (DHS) in a population-based cohort study in Taiwan, which demonstrated a higher risk of DHS in men and younger patients with DM[10]. Ulnar neuropathy can certainly be found in diabetic patients with hand pathology due to compression or ischemia of the ulnar nerve. It is relatively rare and usually found in patients with long diabetes duration[11,12]. The exact probability of an increased frequency of limb mononeuropathies in diabetic patients however is not easily to confirm by epidemiological studies because mononeuropathies due to other factors are relatively frequent in the general population[13]. A prospective electrodiagnostic study suggests that the ulnar nerve is very susceptible to focal entrapment, with a diagnostic rate of 34%, in patients with type 2 DM[14]. Furthermore, one-fifth of those with ulnar neuropathy at the elbow did not have polyneuropathy[14]. In our case, electrophysiological studies showed the pattern of neuropathy beyond the common diabetic hand syndrome. She denied a history of trauma or osteoarthritis that may contribute to tardy ulnar palsy[15], and instead her ulnar neuropathy at the elbow result from a heterogeneous group of pathologies causing peripheral neuropathy.

Cubital tunnel is the most common location where ulnar nerve is compressed around the elbow, compared to arcade of struthers, medial intermuscular septum, medial epicondle, and deep flexor sponeurosis[1]. Interestingly, the clinical manifestations of CUTS may vary by age

at presentation, according to a demographic analysis[16]. Sensory symptoms seem to be more dominant in younger patients who present with more irritation of the nerve. The presentation of CUTS in older patients however is more insidious and predominantly motor. The heterogeneous clinical manifestations may be a result of differing reasons for pressure on the ulnar nerve within the cubital tunnel. Our patient was diagnosed to have a schwannoma which render the onset insidious, rather than traumatic injury or compression by anconeous epitrochlearis that is more common in young patients.

Schwannomas (neurilemmomas) are the most common benign neoplasm of peripheral nerves, but they account for only 5% of soft tissue tumors in adults[17-19]. The tumors commonly develop as slow-growing, and they can remain painless swelling for several years before being diagnosed. Among all the cases with schwannomas in the extremities, only 30-70% complained of pain and 20% had paresthesia on presentation[20]. Similarly, our patient presented with muscle wasting and weakness and initially attributed altered sensation to a concurrent diabetic polyneuropathy. The MRI study serves as a useful tool for preoperational diagnosis, and schwannomas are isointense to muscles in T1-weighted images and hyperintense in T2-weighted ones[21].

CONCLUSION

Diabetes makes a nerve more vulnerable to compression due to a microvascular injury causing local ischemia or by interfering with the innate metabolism of the nerve. The concurrent compressive injury should not be neglected in patients with diabetic polyneuropathy. Electrophysiological and neuroimaging studies were in line with our clinical diagnosis for a patient with CUTS, who was further diagnosed to have a schwannoma at the cubital tunnel by histopathology.

REFERENCES

1. Cutts S. Cubital tunnel syndrome. Postgrad Med J 2007;83:28-31.
2. Sytze Van Dam P, Cotter MA, Bravenboer B, Cameron NE. Pathogenesis of diabetic neuropathy: focus on neurovascular mechanisms. Eur J Pharmacol 2013;719:180-186.
3. Ross MA. Electrodiagnosis of peripheral neuropathy. Neurol Clin 2012;30:529-549.
4. Stamboulis E, Vassilopoulos D, Kalfakis N. Symptomatic focal mononeuropathies in diabetic patients: increased or not? J Neurol 2005;252:448-452.
5. Estrella JS, Nelson RN, Sturges BK, Vernau KM, Williams DC, LeCouteur RA, Shelton GD, Mizisin AP. Endoneurial microvascular pathology in feline diabetic neuropathy. Microvasc Res 2008;75:403-410.
6. Malik RA, Tesfaye S, Thompson SD, Veves A, Sharma AK, Boulton AJ, Ward JD. Endoneurial localisation of microvascular damage in human diabetic neuropathy. Diabetologia 1993;36:454-459.
7. Brown MJ, Sumner AJ, Greene DA, Diamond SM, Asbury AK. Distal neuropathy in experimental diabetes mellitus. Ann Neurol 1980;8:168-178.
8. Schmidt RE. Autonomic neuropathy in experimental models of diabetes mellitus. Handb Clin Neurol 2014; 126:579-602.
9. Papanas N, Maltezos E. The diabetic hand: a forgotten complication? J Diabetes Complications 2010;24:154-162.
10. Chen LH, Li CY, Kuo LC, Wang LY, Kuo KN, Jou IM, Hou WH. Risk of Hand Syndromes in Patients With Diabetes Mellitus: A Population-Based Cohort Study in Taiwan. Medicine (Baltimore). 2015;94:e1575.
11. Boulton AJ, Malik RA, Arezzo JC, Sosenko JM. Diabetic somatic neuropathies. Diabetes Care 2004;27:1458-1486.
12. Boulton AJ, Vinik AI, Arezzo JC, Bril V, Feldman EL, Freeman R, Malik RA, Maser RE, Sosenko JM, Ziegler D; American Diabetes Association. Diabetic neuropathies: a statement by the American Diabetes Association. Diabetes Care 2005;28:956-962.
13. Acosta JA, Hoffman SN, Raynor EM, Nardin RA, Rutkove SB. Ulnar neuropathy in the forearm: A possible complication of diabetes mellitus. Muscle Nerve 2003;28:40-45.
14. Rota E, Zavaroni D, Parietti L, Iafelice I, De Mitri P, Terlizzi E, Morelli N, Immovilli P, Guidetti D. Ulnar entrapment neuropathy in patients with type 2 diabetes mellitus: an electrodiagnostic study. Diabetes Res Clin Pract 2014;104:73-78.
15. Jeon IH, Oh CW, Kyung HS, Park IH, Kim PT. Tardy

ulnar nerve palsy in cubitus varus deformity associated with ulnar nerve dislocation in adults. J Shoulder Elbow Surg. 2006;15:474-478.
16. Naran S, Imbriglia JE, Bilonick RA, Taieb A, Wollstein R. A demographic analysis of cubital tunnel syndrome. Ann Plast Surg 2010;64:177-179.
17. Perrotta R, Virzi D, Tarico MS, Napoli P. An unusual case of symptomatic Schwannoma on the elbow. Br J Neurosurg 2011;25:306-307.
18. Tang CY, Fung B, Fok M, Zhu J. Schwannoma in the upper limbs. Biomed Res Int 2013;2013:167196.
19. Forthman CL, Blazar PE. Nerve tumors of the hand and upper extremity. Hand Clin 2004;20(3):233-242, v.
20. Ichikawa J, Sato E, Haro H, Anayama S, Ando T, Hamada Y. Posterior interosseous nerve palsy due to schwannoma: case report. J Hand Surg Am 2008; 33:1525-1528.
21. Hems TE, Burge PD, Wilson DJ. The role of magnetic resonance imaging in the management of peripheral nerve tumours. J Hand Surg Br 1997;22:57-60.