Focal hand dystonia in a patient with ulnar nerve neuropathy at the elbow

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Published online: 15 June 2010
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Abstract We describe a patient who presented with dystonia of her small finger secondary to entrapment neuropathy of the ulnar nerve at the elbow. Pre operative electrophysiological studies suggested that the locus of entrapment was located proximal to the medial epicondyle. This was confirmed intraoperatively by the presence of a thickened and prominent arcade of Struthers. Surgical decompression resulted in a rapid and dramatic improvement of the dystonic pattern as well as an improvement in nerve conduction. A review of literature has not revealed any other reports of such a clear cut association between ulnar nerve entrapment and non task-specific focal hand dystonia.

Keywords Ulnar nerve · Cubital tunnel syndrome · Compressive neuropathy · Dystonia

Introduction

Ulnar nerve neuropathy at the elbow (UNE) is the second most common entrapment neuropathy in adults accounting for 20% of focal neuropathies demonstrated by electrophysiological studies [1]. The most common symptoms are sensory disturbances along the ulnar nerve distribution,

pain at the elbow, and weakness of ulnar-innervated intrinsic hand muscles. The extent and pattern of motor compromise in the hand is not always uniform and varies depending on which fascicles are actually compromised within the ulnar nerve. A well-known pattern involves greater weakness of the adductor digiti minimi (i.e., third palmar interosseous muscle) relative to the abductor digiti minimi. Consequently, in such cases, the small finger stays abducted, giving rise to the well-known Wartenberg's sign [2]. Selective clawing of the ulnar two digits is also a common finding. While such relative imbalances are known and usually lead to static deformities of the fingers, very few reports of focal dystonia following entrapment neuropathies have been published. The commonest association appears to be a form of task-specific dystonia in musicians [3]. We report a case that establishes a clear association between entrapment neuropathy of the ulnar nerve with focal non-task-directed dystonia of the small finger.

Case Report

A 62-year-old left-hand-dominant lady presented with a history of weakness of her left hand along with altered sensation in the ring and small fingers. She also indicated that her small finger exhibited a tendency to involuntarily and intermittently “override” her ring finger. There was no history of antecedent trauma to the wrist or elbow. She was not performing any activities that involved repetitive movements of the hand. There was no history of diabetes mellitus or thyroid problems.

Clinical examination revealed a patchy motor loss affecting the ulnar-innervated muscles of the hand. The adductor digiti minimi (third palmar interosseous) had a

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Figure 1 Preoperative picture showing dystonic contraction of the small finger.

power of 3/5 while the abductor digiti minimi and fourth dorsal interosseous muscles showed no demonstrable strength (0/5). Sensory loss was also patchy over the ulnar-innervated area with the two point discrimination being very variable between 9 and 20 mm. Interestingly, Tinel's sign was equivocal over the cubital tunnel. Deep tendon reflexes were symmetrical in the upper extremities. In addition to these, the left hand exhibited a tendency to episodic dystonic adduction and flexion of the small finger so that it moved over to and stayed on the palmar aspect of the ring finger for several minutes at a time (Fig. 1). This

Figure 2 Preoperative nerve conduction study showing slowing of conduction velocity proximal to the elbow.

apparently interfered with effective use of her dominant left hand.

Electrophysiological studies were conducted to more clearly delineate the problem. There was significant focal slowing of motor conduction in the ulnar nerve across the elbow. The inching method was used to more specifically localize the region of slowing. This revealed that the maximum slowing occurred in a 2-cm segment immediately proximal to the medial epicondyle, with the elbow kept in 90° of flexion (Fig. 2). Needle electromyography (EMG) showed decreased motor units and increased polyphasic units in the ADM, FDI, and flexor carpi ulnaris (FCU); the opponens, flexor, and the adductor digiti minimi also showed similar changes without any myokymic discharges. The flexor digitorum superficialis and extensor digitorum communis muscles in the forearm showed motor unit recruitment corresponding to the dystonic movement.

Based on these clinical and electrophysiological findings which indicated a diagnosis of ulnar nerve neuropathy at the elbow, the patient was taken up for surgery. A longitudinal incision was made over the cubital tunnel between the medial epicondyle and the olecranon. This was carefully deepened down through skin and subcutaneous tissue. Various cutaneous nerve branches crossing the incision were identified, retracted, and protected throughout the procedure. The ulnar nerve was then identified and the cubital tunnel was released progressively by dividing the ligament of Osborne after which the aponeurosis between the two heads of the FCU was divided. The dissection was

Figure 3 Postoperative picture showing resolution of dystonic contraction of the small finger.

then carried proximally. It became evident at this point that the patient had a very thick arcade of Struthers and this was also divided. Once this was done, the ulnar nerve was examined. It was found to be significantly hyperemic but

did not show any signs of hourglass constriction. After completing the release, the elbow was taken through a full range of movement. It was noted that the ulnar nerve did not subluxate, and hence a decision was made not to proceed with anterior transposition.

Postoperatively, the patient was placed in a long arm splint with the elbow in 30° of flexion for a period of 2 weeks. On her first postoperative visit at 2 weeks, the patient demonstrated dramatic motor recovery, though her sensory deficits persisted. Muscle strength in the previously weak muscles was 4/5, and there was complete resolution of her dystonic movements (Fig. 3). Electrophysiological studies 6 weeks after surgery showed significant improvement in motor conduction in the ulnar nerve across the elbow (Fig. 4).

Discussion

Focal dystonic posturing can occur as a primary problem with no clear and demonstrable cause or secondary to a number of central nervous system disorders like Parkinson's disease or rarely follow some form of nerve injury (i.e., trauma or entrapment). The most familiar forms of focal dystonia seen by a hand surgeon are the “writer's cramp” and “musician's cramp” which are examples of task-specific dystonia. Though a vast majority of these cases appear to be “primary dystonias,” about 10% of patients with writer's

Figure 4 Postoperative nerve conduction study showing improved conduction velocity proximal to the elbow.

cramp report previous hand trauma [4, 5]. However, typical posttraumatic dystonias, described in the literature, have features significantly different from primary dystonias. They tend to be “fixed dystonias” unlike the action dystonias and mobile dystonic postures seen in primary dystonia [6]. Other differences include limitation of passive movements, contractures, absence of sensory tricks, and presence of chronic regional pain syndrome [7–10].

Review of literature suggests that peripheral nerve lesions may cause dystonias, sometimes with features of primary dystonias. Charness et al. [3] in 1996 reported the presence of ulnar nerve neuropathy in 40% of musicians with occupational cramps. Near nerve recording technique showed focal slowing of motor conduction in 79% of patients while surface recording was abnormal in only 29%. The same authors noted that ulnar neuropathy was present in 77% of patients with flexion dystonia of the ring and small fingers and in only 10% of patients with other patterns of dystonia. They reported improvement in task-specific dystonia of musicians in 3–12 months after ulnar nerve decompression. Ross et al. demonstrated a dystonic pattern of antagonistic bursting in patients with ulnar nerve neuropathy and suggested that ulnar neuropathy may initiate or sustain a specific dystonia, by inducing a central disorder of motor control [11]. They studied surface EMG of the extensor digitorum communis and flexor superficialis muscles to the ring finger during tapping movements of the ring finger in patients with ulnar neuropathy as well as normal subjects. They were able to demonstrate an abnormal burst pattern and loss of the silent period in those with ulnar neuropathy. Girlanda et al. [12] conducted a more extensive study and speculated that a peripheral nerve injury can potentially induce a rearrangement of reciprocal inhibition circuits at the spinal cord level, which may predispose to the development of dystonia. However, as their patients did not show clinical dystonia, those authors suggested that some unknown factor may be required to alter the intracortical circuits and produce clinically evident dystonia. We found one report of focal hand dystonia with wrist and finger flexion in a patient with thoracic outlet syndrome by Quattorone et al. [13]. In this case, the dystonia apparently improved 12 h after surgery to resect a cervical rib. Becker et al. [14] in 2002 reported shoulder girdle dystonia in selected patients with cervical disk prolapse. They concluded that peripheral trauma to the nerve roots may precipitate dystonic movements in susceptible patients.

Our case demonstrates a case of non-task-specific focal dystonia that occurred secondary to an entrapment neuropathy that was demonstrable clinically and electrophysiologically as well as intraoperatively. This relationship was further demonstrated by the dramatic recovery of both clinical and electrophysiological parameters after surgical decompression. To our knowledge, this is the first reported

case that establishes a clear-cut relationship between an ulnar nerve entrapment neuropathy at the elbow and non-task-specific dystonia of the hand.

This case has led us to reflect on the possible reasons for the rarity of such dystonia in entrapment neuropathies of the upper extremity. Since this case clearly establishes that peripheral nerve compression can potentially cause changes in motor control circuits leading to dystonia, the question remains as to why similar dystonia has not been reported following carpal tunnel syndrome which is the most common entrapment neuropathy in the upper extremity. Has it to do with the fact that the ulnar nerve innervates the bulk of the intrinsic hand muscles? Even if that were to be the case, why is dystonia rare in ulnar nerve neuropathy afflicting nonmusicians as in this patient? If significant changes in motor control circuits cause the dystonia, why the rapid improvement in dystonia following surgical decompression? Perhaps more reports such as these will lead to an increase in awareness for the potential for dystonia in peripheral nerve disorders. This may lead us to be on the lookout for this clinical sign and eventually a better understanding of its pathophysiology. It is also important to identify not only entrapment neuropathies, but also hand dystonias in patients with repetitive overuse syndrome of the upper limbs, so that appropriate therapeutic measures can be instituted [15].

Disclaimer No benefits, financial or otherwise, have been received by any of the authors for whole or part of this report.

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