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Case Report

A rare case of proximal arm neurofibroma causing ulnar nerve compression: successful surgical intervention and recovery

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ABSTRACT

Compressive neuropathy of the ulnar nerve due to a neurofibroma above the elbow is extremely rare, with few cases reported worldwide. A young male presented with a progressively enlarging swelling and increasing pain in the right upper arm. Over time, the patient developed symptoms indicative of ulnar nerve compression, including numbness, tingling, and weakness in the hand and forearm. Physical examination and diagnostic imaging revealed the presence of a neurofibroma compressing the ulnar nerve above the elbow. The patient also exhibited generalized features consistent with neurofibromatosis, such as multiple café-au-lait spots and cutaneous neurofibromas. Given the severity of the symptoms and the confirmed diagnosis, the patient underwent surgical excision of the neurofibroma along with neurolysis to decompress the ulnar nerve. The surgical intervention aimed to relieve pain and improve sensory and motor function. The surgical procedure was successful, resulting in significant pain relief and marked improvement in sensory and motor functions. Postoperatively, the patient reported substantial symptomatic relief and regained functional use of the affected limb. This case underscores the importance of considering rare causes of ulnar nerve compression, such as neurofibromas, particularly in patients with underlying neurofibromatosis. It also highlights the effectiveness of surgical removal and nerve decompression in resolving symptoms and restoring function in such complex cases.

Keywords: Neurofibroma, Surgical excision, Ulnar nerve, Neurofibromatosis

INTRODUCTION

The ulnar nerve is a major peripheral nerve of the upper limb, originating from the brachial plexus with contributions from the C8 and T1 nerve roots. It traverses a complex anatomical pathway, descending along the medial aspect of the arm, crossing behind the medial epicondyle of the humerus, and extending into the forearm and hand. The ulnar nerve is particularly vulnerable to compression at various anatomical sites, leading to neuropathy characterized by sensory and motor deficits. Ulnar pain can originate from compression of a variety of places such as the cervical nerve roots as they exit the spinal cord, the brachial plexus, the thoracic outlet, or further down the upper extremity in the arm, elbow, forearm, or wrist.[1] Key anatomical landmarks along its course include the medial intermuscular septum, where the nerve runs in the medial compartment of the arm; the cubital tunnel at the elbow, where the nerve passes behind the medial epicondyle; and the forearm, where it traverses between the two heads of the flexor carpi ulnaris muscle before entering the hand via Guyon's canal. Ulnar neuropathy can occur at various levels along the nerve's course. Compression at the elbow, known as cubital tunnel syndrome, is the most frequent site of ulnar nerve entrapment. Here, the nerve can be compressed by fascial bands or the flexor carpi ulnaris muscle, causing numbness and tingling in the ring and little fingers, hand weakness, and muscle wasting. Compression at the wrist, known as Guyon's canal syndrome, typically affects the intrinsic

muscles of the hand, leading to similar sensory symptoms and muscle weakness but sparing the forearm muscles. Compression in the arm is rare but can occur due to anomalous muscles, fibrous bands, or space-occupying lesions such as neurofibromas. Symptoms of compression in the arm include pain, paresthesia, and motor deficits similar to those observed in more common sites of compression but originating higher along the nerve pathway.

CASE REPORT

A 28-year-old male carpenter presented to OPD with a one-year history of slowly progressing pain in the proximal right arm, accompanied by pain and numbness in the hand. Over the past two months, his symptoms had significantly worsened. Physical examination revealed multiple café-au-lait spots and cutaneous neurofibromas, indicative of neurofibromatosis, along with features of generalized ligamentous laxity. A hard swelling was palpable over the medial proximal arm. There was notable wasting of the hypothenar muscles, ulnar clawing of the fingers (Figure 1), and weakness of the interossei and lumbricals, with a positive Froment's sign (Figure 2). The patient's grip strength was severely compromised, and he had poor sensation in the ulnar nerve distribution in the forearm and hand, significantly impacting his ability to perform daily activities. The Tinel sign was positive over the swelling. Ophthalmological consultation revealed multiple healed ulcers on ophthalmoscopy and Lisch nodules on the bilateral iris on slit lamp examination.

X-ray imaging of the affected area showed no bony involvement. Blood investigations, including calcium, vitamin D, and liver function tests, were all within normal limits. A nerve conduction study confirmed ulnar nerve neuropathy. MRI of the arm and shoulder revealed multiple confluent fusiform T2 hyperintense and T1 hypointense lesions, the largest measuring 6.7×3.4 cm in the mid-arm along the ulnar nerve. Multiple other similar lesions were also noted (Figure 3). Given the involvement of the ulnar nerve and the severity of the symptoms, surgical intervention was undertaken for symptomatic relief. The swelling was excised through a medial zig-zag incision over the upper medial arm. The lesion was found to be compressing the ulnar nerve, and careful excision was performed along with nerve neurolysis (Figures 4 and 5). The excised swelling was sent for biopsy. Macroscopically, the lesion appeared as a grey-white mass that enlarged the nerves in a tortuous fashion (Figure 6). Microscopic examination showed multinodular, plexiform bundles of bland spindle cells with elongated, tapered nuclei and wavy contours. There was no evidence of atypia, necrosis, or increased mitosis (Figure 7). Postoperatively, the patient underwent physiotherapy sessions. At a six-month follow-up, the patient showed complete recovery of motor and sensory function with a good functional grip strength, allowing him to resume all routine activities (Figure 8). This case underscores the importance of considering rare causes of ulnar nerve compression and demonstrates the effectiveness of surgical treatment in achieving symptomatic and functional improvement.

DISCUSSION

Neurofibromas are the most prevalent peripheral sheath tumours.[2] They comprise of Schwann cells, perineural cells and mast cells in myxoid back ground. Majority of them occur sporadically. Out of localised, diffuse and plexiform types, plexiform neurofibromas carry high chance of malignant transformation.[2] In this report, we present a rare case of ulnar nerve compression in the arm due to a neurofibroma. Neurofibromas are benign nerve sheath tumours that can cause significant morbidity when compressing peripheral nerves. This case underscores the importance of considering fewer common sites of compression in patients presenting with ulnar neuropathy symptoms, particularly when standard diagnostic approaches for cubital tunnel syndrome or Guyon's canal syndrome fail to identify the source of nerve compression.

Plexiform neurofibromas are common in neurofibromatosis type 1 patients (26.7%) and are often a source of functional and cosmetic impairment. It involves diffuse involvement of nerve and its branches which defines its plexiform nature (mass like appearance).[3] There is often associated soft tissue involvement and is massive that often hinders its complete resection and diagnosis of early malignant transformation. It can arise almost from any nerve and its part that may be superficial cutaneous nerves, cranial nerves, spinal nerves, deep peripheral nerves, brachial or lumbar plexus.[3] Depending on the region of nerve involvement it can be visible on surface or can remain as deep-seated mass.[3] Thoracic and lumbar plexus involvement is often accompanied by massive limb hypertrophy. Lesions can grow longitudinally or circumferentially around the nerve with two specified growth periods of early child hood and puberty. Plexiform mass often shows the presence of centrally passing intact nerve fibres. Compressive neuropathy may take decades to present in spite of massive tumour presence shown by muscle atrophy.[4] Patients often present with chronic, severe progressive pain in compressive neuropathy. These along with cutaneous manifestations leads to suspicion of plexiform or nodular neurofibromas.[5]

Initial investigation is a USG of peripheral nerve. High resolution ultrasonography shows nerve swelling proximal to compression site and focal change in nerve texture.[9] Ultra sonography is useful mainly for distal entrapment neuropathies.[10] Although EMG is the investigation for localizing the site of compression, it may reveal false negative results and non-localizing findings.[8] MRI is the Investigation of choice for proximal entrapment neuropathies.[3] Magnetic resonance imaging of peripheral nerve provides a non-invasive and accurate diagnosis.[6] Target sign is characteristic that shows hyper intense periphery and hypo intense centre in T2 weighted images. Plexiform neurofibromas are more heterogeneous and

infiltrative than nodular neurofibromas. Histological examination shows small elongated spindle cells with characteristic wavy or comma shaped nuclei arranged in myxomatous stroma and are S-100 positive. Surgical excision remains the definite treatment.[6] The indications include pain, neurological deficit, cosmetic impairment and malignant degeneration Malignant degeneration occurs in 20-29% of patients.[7]

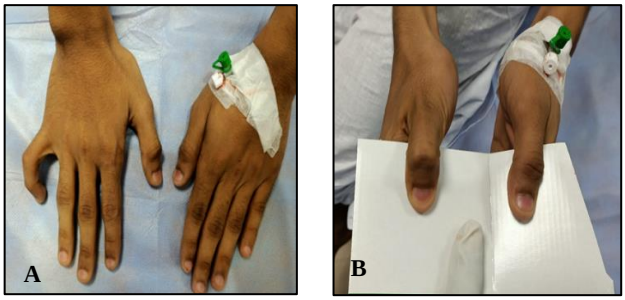

Figure 1: (A) Ulnar clawing of the right hand, (B) illustrates positive froments sign with overactive FPL muscle.

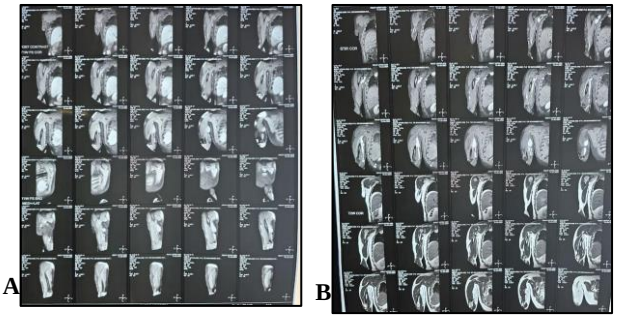

Figure 2 (A and B): MRI fims revealing multiple neurofibromas around the right arm and shoulder.

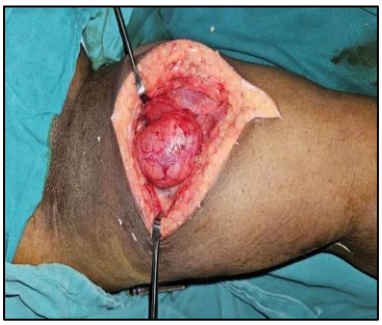

Figure 3: Intraoperative image of the swelling over upper medial aspect of arm.

Diffuse plexiform neurofibromas grows intra and extra fascicular and hence complete resection is not possible. Pain relief is imminent but there can be neural deficit due to nerve manipulation. Radical surgical removal often requires removal of a large nerve trunk without which recurrence is the rule followed by neurological deficit. Interferon alpha has been used in plexiform neurofibroma with variable results.[1] FDA has approved the use of mitogen activated protein kinase inhibitors (selumitinib) for paediatric patients above 2 years with inoperable plexiform neurofibromas.[11] Neurofibromas pose major challenge in deciding the time of excision with surgeons advising removal mass enbloc in child hood and another majority to wait till the lesion is symptomatic. In our case after patient had a very good post-operative outcome with relief of symptoms.

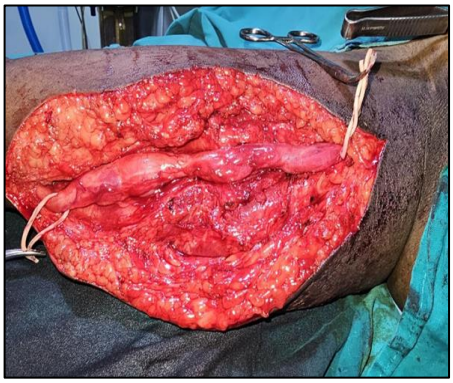

Figure 4: Intraoperative image showing ulnar nerve after excision of the swelling.

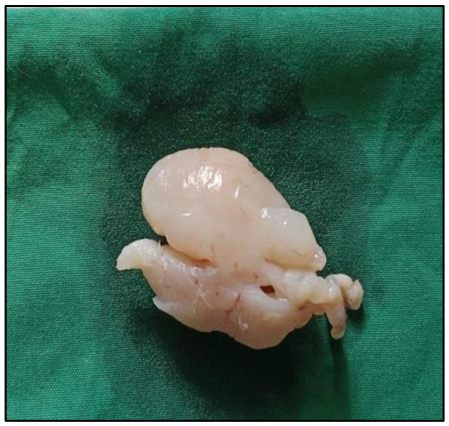

Figure 5: Macroscopic picture with grey-white mass that enlarged the nerves in a tortuous fashion.

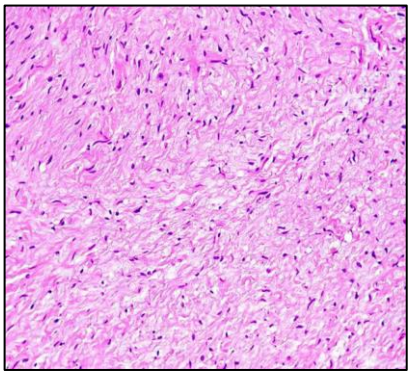

Figure 6: Microscopic picture with H and E Staining demonstrating multinodular, plexiform bundles of bland spindle cells with elongated, tapered nuclei and wavy contours.

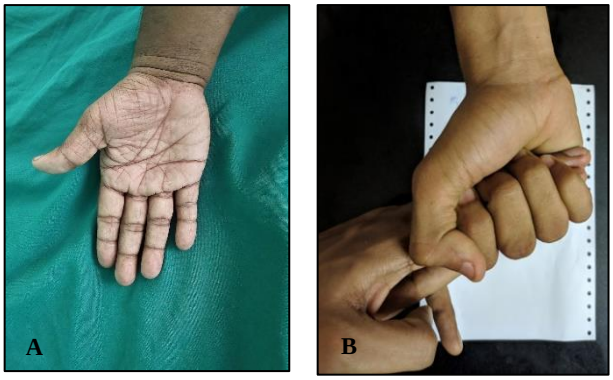

Figure 7 (A and B): Clinical follow up image of hand 6 months after surgery with no hand clawing and good grip strength.

CONCLUSION

This case underscores the exceptional rarity of ulnar nerve compression caused by a neurofibroma in the proximal arm, especially in patients with neurofibromatosis. The successful surgical excision and neurolysis not only provided significant symptomatic relief but also facilitated full functional recovery, allowing the patient to resume normal activities. This report highlights the importance of considering uncommon etiologies in cases of atypical neuropathy and demonstrates the efficacy of prompt surgical intervention. The favourable outcome emphasizes the necessity for thorough diagnostic evaluation and individualized treatment strategies in managing peripheral nerve tumors, ultimately enhancing patient care and prognosis.

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