

# Neurofibroma of The Median Nerve Producing Carpal Tunnel Syndrome in Young Adult

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## ABSTRACT

Carpal tunnel syndrome usually presents bilaterally and a secondary nature should be suspected in patients with unilateral symptoms, especially in children and young adults. Solitary peripheral nerve tumours are rare and difficult to diagnose. Schwannoma being the most common followed by neurofibroma and both produces similar clinical signs and symptoms. Complete resection of tumour is nevertheless recommended so decision making during treatment is very difficult. We presented a case of 19 year boy presented with mass in the volar aspect of right wrist and features of carpal tunnel syndrome treated by decompression and incision biopsy preserving the nerve function.

**KEY WORDS:** carpal tunnel syndrome; median nerve; neurofibroma; tumour.

## INTRODUCTION:

Isolated neurofibromas in the hand are rare and pose both diagnostic and treatment challenges for the surgeon. Preoperative imaging can be useful to delineate the anatomical relationships of a soft tissue lesion but is not always diagnostic.<sup>1</sup> Carpal tunnel syndrome (CTS) is by far the most frequent compression neuropathy and women are affected twice as often as men and manifestation usually occurs over the age of 30. In young adults typical diagnostic clues, may be absent leading to misdiagnosis and late treatment.<sup>2</sup> Whenever a patient presents with unilateral symptoms and unilateral neurophysiologic impairment, the possibility of a space-occupying lesion compressing the median nerve should be kept in mind in the differential diagnosis.<sup>3</sup> It is established that the neurofibroma may become malignant and that removal involves resection of the nerve but the radical resection is nevertheless recommended and

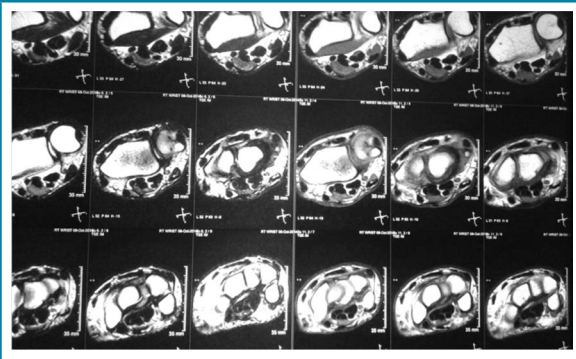
the defect in the nerve can be overcome by free nerve transplantation.<sup>4</sup> We did decompression of carpal tunnel micro-dissection of tumour mass for incision biopsy by considering the resection latter on if required; now the patient is symptom free till 6 month of follow up.

## CASE REPORT:

A 19- year boy presented in the outpatient department with lump located in the volar side of right wrist and complains of wrist pain, parasthesia and decreased sensation over thinner aspect of hand, and 2<sup>nd</sup> -4<sup>rd</sup> fingers for 1 year duration. On examination 3x2cm lump at the level of palmar crease (zone 5) was found. Lump was non tender and firm in consistency, positive Tinnel sign and Phalens sign without objective motor deficit was noted. Patient was thoroughly examined for stigmata of neurofibromatosis and there were no family history suggestive of neurofibromatosis as well. USG revealed 20x15 mm mass in close approximation with median nerve. MRI suggests soft tissue mass on the volar aspect of right distal forearm in close apposition and inseparable from the median nerve D/D fibroma, neurofibroma. (Figure 1)

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**Figure 1. Right wrist MRI showing intraneural mass of the median nerve.**

Patient was advised surgical treatment, purpose of surgery and possible options with their advantages and disadvantages. Decompression of carpal tunnel was done, median nerve was found swollen, inseparable intraneural mass of 3x2 cm originating from the median nerve itself was found. Micro dissection of the tumour mass was tried but it was coming out from the nerve itself so that only incision biopsy from the mass was taken and sheath closed by 6-0 prolene suture preserving the nerve and sent for histopathology. (Figure 2)



**Figure 2. Intra –operative photograph of the swollen median nerve with mass at wrist level.**

Histopathology revealed un-encapsulated proliferations of interlacing bundles of spindle cells with hyperchromatic serpentine nuclei intermixed with dense bundles of collagenous fibres suggestive of neurofibroma. Patient was advised for possible malignant transformation of tumour, its features options of treatment i.e resection and nerve transplantation. He was relieved of pain and parasthesia postoperatively, no neurological deficit and completely symptoms free till 6 month of follow up.

## DISCUSSION:

The carpal tunnel syndrome (CTS) is by far the most frequent compression neuropathy but in young adults typical diagnostic clues suggestive of a CTS, may be absent leading to misdiagnosis and late treatment.<sup>2</sup> Whenever a patient presents with unilateral symptoms of carpal tunnel syndrome and unilateral neurophysiologic impairment, the possibility of a space-occupying lesion compressing the median nerve should be kept in mind in the differential diagnosis.<sup>5</sup> Because of its relative rarity and a wide variety of clinical manifestations (lipoma, fibroma, neuroma, ganglion, cystic lesion etc.) peripheral nerve tumours often present to the specialists from widely different disciplines, thus often resulting in delayed diagnosis and a non-cohesive pattern of management.<sup>6</sup> Neurofibromas are rare benign peripheral nerve tumour and management depends on symptoms. They do not typically need to be surgically removed; exploration and excision may be required under the following circumstances: diagnosis, pain, cosmetic, progressive neurological complications, compression of adjacent tissues and suspicion of a malignant tumor.<sup>7</sup>

Complete resection of solitary nerve associated with benign tumour of extremities is possible and associated with an improved outcome had been reported but preservation of nerve continuity is the underlying goal of the therapeutic strategy, irrespective of the type of tumor.<sup>8</sup> Extricable tumours (schwannomas, intraneural lipomas) displace nerve fiber bundles without penetrating into the bundle itself and can thus be resected without interrupting nerve continuity. Inextricable tumours (solitary neurofibromas, hemangiomas of the Schwann sheath, neurofibrolipomas) infiltrate the structural elements of the nerve fibers making complete excision impossible without altering the nerve function.<sup>9</sup> Surgical excision is the most effective therapy, however some authors recommend excision of only symptomatic tumours or those demonstrating enlargement during follow up.<sup>9</sup> We stress on the importance of imaging studies in patients with unilateral symptoms that are usually not used in carpal tunnel syndrome. The reported patient was evaluated and magnetic resonance images revealed an intraneural space-occupying lesion. Although rare, the surgeon should include in the differential diagnosis of carpal tunnel syndrome, the unusual cause of tumours and tumour-like lesions,

especially when the patients profile is not typical (young, male, no repetitive stress or manual labour). In addition, the presence of a palpable mass at the distal forearm or palm dictates the need for imaging studies. The extent, location and aggressiveness of the mass will determine the approach and type of procedure.<sup>10</sup>

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