An Ulnar Neurofibroma: Management with Sural Graft Reconstruction

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Received: 10 March 2015 / Accepted: 25 May 2015 / Published online: 4 June 2015
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Introduction

Neurofibromatosis type 1 (NF1) is a common inherited disorder that affects about 1 in 2500 to 1 in 3000 people worldwide, irrespective of sex or ethnic origin. Individuals affected by NF1 are prone to develop benign and malignant tumors of the central and peripheral nervous system, in addition to malignant diseases affecting other parts of the body [1].

The tumor with highest prevalence in patients with NF1 is the neurofibroma, which is primarily composed of Schwann cells and fibroblasts with numerous perineural, endothelial, and mastcells [2]. Ten different histopathological types of neurofibromas—with no significant difference in their clinical presentation—have been reported in patients with NF1. Except from plexiform neurofibroma, which is pathognomonic for NF1, other histopathologic variants may occur sporadically or in association with neurofibromatosis [3].

Neurofibromas and schwannomas are common benign nerve sheath tumors which are currently considered as distinct entities. Neurofibromas diffusely expand the involved nerve and encompass included axons, whereas schwannomas commonly arise from intracranial nerves. Histologically, neurofibromas contain cells with oval to spindled, wavy nuclei in a variable collagenized stroma. S100 immunoreactivity is generally present. In neurofibromas, although it is usually

restricted to a smaller proportion of cells, it is less uniform than in schwannomas. The incidence of malignancy is significantly greater in NF1 patients with neurofibromas compared with NF1 patients with schwannomas [4]. Around 10 % of patients affected by NF1 will develop a malignant peripheral sheath tumor (MPNST) during their lifetime, and nearly 50 % of patients with MPNST have NF1 [4].

A Case Report

Our case was a 45-year-old woman, affected by NF1. She noticed a lump in the medial third of the ulnar side of her right forearm 2 years before being first diagnosed. She consulted for dysesthesia in the ulnar portion of the right hand and 5° finger; soon after, she noted loss of strength in the 5° finger and in the interosseus muscles of the same hand. The lump grew and her clinic signs rose.

Physical exam revealed a mild atrophy of the hypothenar muscles and the first interdigital area and paresis of the abductor digiti minimi muscle in the affected hand. There was hypesthesia in the cutaneous area of the ulnar nerve at the hand level. A mass was evident in the medial third of the ulnar side of the patient's forearm, which was extremely painful under mechanical stimulation. Moreover, Tinel's sign was positive along the territory of the ulnar nerve. Examination of all the limbs revealed multiple café-lait spots and cutaneous nodules. No other tumors were detected. Preoperative MRI revealed a multilobulated mass along the medial third of the forearm. This mass extended along ulnar nerve distribution with the probable diagnosis of neurofibroma. Preoperative EMG noted a mild mixed affectionation of the ulnar nerve.

During surgery, the right ulnar nerve was exposed in the medial third of the forearm (Fig. 1). Using microsurgery techniques with the microscope, we discovered a big tumor from

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Fig. 1 Ulnar neurofibroma, in continuity with a fasciculus

one fascicle of the nerve. We removed the mass ($7.5 \times 5 \times 3.5$ cm) preserving the stumps of the fascicle (Figs. 2 and 3). There were not extraneural tumors. We decided to bridge the defect with an interfascicular graft of the sural nerve, which had a similar diameter. We exposed the donor nerve by a small longitudinal incision behind the lateral malleolus. The nerve was found just behind the small saphenous vein. We removed 10 cm from the sural nerve and prepared it for a one fasciculus, which was the defect of the nerve necessary in this case. The ends of the graft were meticulously coapted to the corresponding end of the fasciculus using microscope. The stumps of the donor nerve were cut to obtain a more optimal epineurium. We carried out an epi-perineurium suture without tension. One suture of 10-zero nylon was used at each end of graft. We passed the suture through the perineurium of the fasciculus and the epineurium of the graft and knotted it very loosely [5]. The histopathological exam revealed a neurofibroma tumor, with a several mixoid component.

Disease-related pain disappeared after 4 weeks from surgery. We found evidence of nerve regeneration 6 months after surgery. After 6 months from surgery, Tinel's sign was tested negative at the ulnar forearm (this can be modified for a more accurate statement), thus suggesting nerve regeneration. Furthermore, there was neither strength loss nor dysesthesia in the hand and no further tumors were discovered.

Fig. 2 The ulnar neurofibroma dissected and united to its fasciculus

Fig. 3 Ulnar neurofibroma, HE

Discussion

Neurofibromas have been reported in the thumb [6], palm [7], index finger [8], median nerve [9, 10], fifth finger [11], and wrist [12, 13]. Interestingly, six cases have been reported for the ulnar nerve, although they were treated with different approaches [14–19].

Caroli et al. [14] reported improvements on clinical signs, with no impact on nerve function, after surgical removal of a small ulnar neurofibroma in the Guyon canal. For a neurofibroma of the ulnar nerve found next to the elbow, Lohmeyer et al. [15] decided to remove the tumor and to reconstruct the nerve with an interfascicular graft of the sural nerve. Authors reported nerve regeneration 6 months after surgery. Brihate et al. [16] published a case report from a patient with gigantism who presented a plexiform neurofibroma in the epitrochlear sulcus. Authors decided to excise the tumor. Since its anastomosis was not possible, pain syndrome could only be avoided through amputation of the fifth finger and a partial excision of the soft tissue hypertrophy of the hypothenar. Marchetti et al. [17] opted for a radiosurgical treatment in a case of a small ulnar neurofibroma case, achieving satisfactory clinic results. Gosein [18] reported a neurofibroma bigger than usual (11 cm) in the ulnar nerve that was not completely removed. Although the patient showed no postoperative motor deficits, he reported tingling along the ulnar nerve distribution. Finally, Gonzalez-Darder and Pseudo-Martínez [19] reported a case of multiple ulnar neurofibromas, classified as NF5, with many masses along the ulnar nerve. Authors could remove only three tumors, which were easily resectable. However, remaining tumors were not possible to remove since they originated from multiples fascicles. Alternatively, anterior transposition of the ulnar nerve in the elbow was performed at the end of the microsurgical procedure. Surgery resulted in pain removal, and the patient could return to his previous occupation.

Therefore, to the best our knowledge, only Lohmeyer et al. [15] reconstructed the nerve with an interfascicle graft, despite neurofibroma being involved in the nervous driving, due to

the affection of the nerve fascicles. We decided to treat great ulnar neurofibroma by reconstructing the nerve, since evidence suggest that nerve reconstruction is the best procedure in order to reestablish nervous connections by a new neural tube [20]. We chose the sural nerve because of our knowledge of this donor nerve and because of its similar size to the fasciculus removed.

We have obtained satisfactory results using a nervous graft, despite the fact that we had to carry out two different approaches, on the forearm and on the leg, however, our experience using this donor nerve is satisfactory and other options were dismissed, for example, the end-to-side technique. End-to-side nerve repair offers a promising technique for repair of peripheral nerve injuries where the proximal nerve stump is unavailable or a significant nerve gap exists. End-to-side holds great appeal in a wide range of clinical scenarios as it eliminates morbidity associated with nerve grafts, minimizes the number of suture lines crossed by regenerating axons, and decreases total time to reinnervation by shortening axon regeneration distance [21, 22]. However, we assessed the ulnar nature and its distal structure, where the fasciculus are more intertwined, with motor and sensory fibers united in the same fasciculus [23]. It agrees with the theory of Kayikcioglu [24], which has achieved to show no motor or sensory reinnervation at 20 months follow-up after immediate repair end-to-side of median to ulnar nerve with an epineurial window. For this reason, we prefer the use of nerve graft technique that in addition is more dominated by us.

In conclusion, neurofibroma is a mass which affects always a nervous fasciculus; so in our opinion, to reestablish the continuity of the nervous tube is essential to return to the normal nervous function. Sural nerve graft is a suitable option when the ulnar nerve is affected, although end-to-side technique could be an optimal surgery of the median nerve.

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