Plexiform Neurofibroma from Palmaris Longus with Carpal Tunnel Syndrome

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ABSTRACT

Plexiform neurofibromas (NF) involving the palmaris longus tendon are rare diseases difficult to diagnose when the classical manifestations, e.g. skin pigmentation, subcutaneous nodules, lisch nodules, family history, etc are absent. We report a case of 26 years male with plexiform NF of palmaris longus tendon which is a relatively rare site. Plexiform NF commonly involve the cranial nerves.

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INTRODUCTION

Plexiform neurofibroma (NF) is a benign tumor arising from axon, schwann cell or fibroblast and eventually leads to enlargement of involved nerve trunk. NF is a group of autosomal dominantly inherited disorder arising from neural tissue. There are two types. Neurofibroma-1, being the most common type (90% of the cases) and affects 1 in 3,000 newborns with 50% mutation and almost 100% penetrance. The genetic abnormality of NFI is mapped to chromosome 17q11.2. Neurofibroma-2 is much less common and affects 1 in 40,000 individual, the genetic mutation of NF-2 is mapped to chromosome 22 which bilateral acoustic neuromas are present leading to hearing loss starting as early as the teen age years.

Plexiform NF is common over the branches of trigeminal and cervical nerves over the face and relatively less common in extremities. Some authors have reported an incidence of 0.8% for NF in the hands and from 10 to 15% for NF-1. We are reporting a rare case of plexiform NF arising from nerve branches supplying palmaris longus tendon.

CASE REPORT

A 26-year-old man came to our institute with chief complaint of tingling sensation and numbness in left index, middle and ring finger since 1 year. It was associated with a painless swelling (4 × 3 cm) over volar aspect of wrist just proximal to proximal wrist crease. On examination, there was a 1×1 cm size swelling, smooth, well defined, firm, and mobile in direction of tendon, with feel of bag of worm. No skin pigmentation, nodules, iris nodule or hearing problem were noted. Family history was not significant.

X-ray was normal. Electromyogram (EMG) and nerve conduction velocity (NCV) showed median nerve compression at the site of scar mark. Intraoperatively there was a swelling on palmaris longus tendon measuring 5 × 3 cm compressing the median nerve. The swelling was resected and sent for histopathological examination (Fig. 3). Postoperatively median nerve function was normal. And symptoms reduced after surgery. Histopathology report shows a well circumscribed tumor compromising of both the neural and fibrous tissue. The individual neural cells are elongated with oval nuclei having eosinophillic fibrillary cytoplasm suggestive of plexiform NF.

DISCUSSION

Neurofibromatosis type I (NFI) was first described by Frederick von Recklinghausen in 1882 and for that reason has also been known as von Recklinghausen’s neurofibromatosis. The diagnosis of NF-1 is made on the basis of a group of clinical manifestations following the established NIH criteria proposed in 1988 (11). It includes cutaneous neurofibromas, café-au-lait spots (five or more of 1.5 cm size and larger in adults and 0.5 cm or larger in puberty and under), axillary freckling (Crowe’s sign) and Lisch nodules among many others. Café au late spots are the first manifestation present in 90% of patients; 50% of patients develops lesions during their first year of life. Cutaneous nodules also are common manifestation is present in 90% of patients; 50% of patients develops lesions during their first year of life. Cutaneous nodules also are common manifestation is present in 40% of patients. The tumor arising from neural element is not confined to skin and subcutaneous tissue but it also involves some viscera. Plexiform NF can be of two types, nodular and diffuse. Diffuse type is also known as elephantiasis neurofibromatosa. The patient which we have described is a case of nodular plexiform NF. He developed swelling in adulthood and presented to us with the symptoms...
resembling carpal tunnel syndrome. NCV confirmed the diagnosis of median nerve compression.

Although NF is a benign condition, it can undergo malignant transformation into a malignant peripheral nerve sheath tumor.\textsuperscript{4-6} In some reports the incident rate has been up to 2 to 5%.\textsuperscript{4} Only two forms of NF, plexiform and localized intraneural NF, are significant precursors of malignant peripheral nerve sheath tumors.\textsuperscript{5}

The management of NF is not well-defined being a genetic etiology its management is aimed at mostly controlling symptoms. Excision of the tumor is the only available therapy. And to completely excise tumor from nerve there is possibility of partial or complete loss of function. According to Seddon, tumor resection is not necessary, unless the tumor causes pain, shows exuberant growth, or impairs function. Surgery was indicated, since patient was having symptoms of median nerve compression.\textsuperscript{7} Swelling was localized to tendon of palmaris longus and was not from median nerve so complete removal was possible as we removed it with tendon. Excised swelling and tendon were resected successfully and the swelling was sent for histopathological reporting. Swelling was from nerve to palmaris longus branch of median nerve.

CONCLUSION

Diagnosis of plexiform NF is difficult without associated lesions like café-au-lait spot, irish nodule, axillary freckling, biopsy is must. As such no treatment is available but surgery is indicated when it causes functional impairment.

REFERENCES