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Plastic Surgery

PLEXIFORM NEUROFIBROMA OF THE MEDIAN NERVE IN THE PALM

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ABSTRACT **INTRODUCTION:** Plexiform Neurofibromas occur commonly in the cervical and trigeminal nerve distribution but presentation in extremities is rare. This study is a review of 4 cases of plexiform neurofibroma in the palm.

MATERIALS AND METHODS: Between January 2016 to December 2017, four cases of Plexiform Neurofibroma involving the median nerve in the palm underwent excision and reconstruction as warranted. Postoperative outcome monitored.

RESULTS: All patients underwent complete excision. Nerve grafting was done in two patients. All wounds healed without any postoperative wound complications. None had motor weakness. At the end of 6 months, sensation was S3+ in all patients.

CONCLUSION: Management of plexiform neurofibroma requires proper diagnosis, meticulous surgical technique and postoperative physiotherapy to produce a good postoperative result with adequate hand function.

KEYWORDS : Plexiform neurofibroma, Median nerve, Neurofibromatosis

INTRODUCTION

Plexiform Neurofibroma is a benign tumour arising from axon, Schwann cell or fibroblast and eventually leads to enlargement of involved nerve trunk [1]. The disorder can be isolated to a region or associated with other anomalies which include pigmented cutaneous lesions, namely, café-au-lait spots, generalized tumors of neural crest origin, Lisch nodules, defects of cranial vault, kyphoscoliosis, optic gliomas, focal gigantism, and vascular lesions. Plexiform neurofibroma occurs commonly in the cervical and trigeminal nerve distribution over the face with relatively less common occurrence in the extremities. [2]. This study is a review of 4 cases of plexiform neurofibroma involving the median nerve in the palm.

AIM

To study the occurrence and management of plexiform neurofibroma of the median nerve in the palm

MATERIALS AND METHODS

This study included four cases of Plexiform Neurofibroma involving the median nerve in the palm, who presented to our hospital from January 2016 to December 2017. All patients complained of a swelling in the palm with paresthesia involving the median nerve distribution. Thorough history was taken from all patients and meticulous examination done. Physical examination included examination of motor and sensory systems.

Preoperatively, MRI was done for all of the patients to know the extent and plane of swelling and to rule out vascular malformation.

All patients were operated under general anesthesia with pneumatic tourniquet control and under 3.5X loupe magnification. The incisions were marked and made along palmar creases avoiding straight lines. Meticulous dissection was done to preserve the sensory and motor branches of the median nerve along which the neurofibroma was located. [Figure 1]

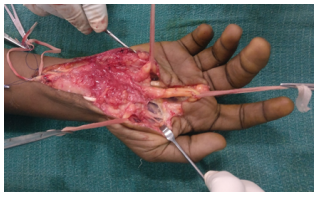

Figure 1: Plexiform Neurofibroma along the sensory branches of Median Nerve

In all patients, excision of the entire neurofibroma mass was done. In two patients, a segment of sensory branch of the median nerve could not be dissected separately from the tumor, and hence was excised. Sural nerve graft was harvested, reversed and nerve anastomosis was done in those cases where segmental sensory nerve loss was present. Motor branches were preserved in all cases. All wounds were closed with drain. The specimens were sent for histopathological examination.

Postoperatively the drain was removed after 48 hrs. The wound was assessed on the 2nd and 5th day. Physiotherapy was started after 48 hrs. Tinel's sign was assessed after 21 days. Motor examination was done after 5 days and 14 days and 21 days. Detailed Sensory evaluation including crude and fine touch, pressure sense, static and dynamic two-point discrimination (2 PD) testing, vibration sense and cold and hot testing were done on days 21, 30, 60, 90, 120, 150, 180. Preoperative and postoperative sensations were graded using the MRC scale [3]. The progress in tinel's sign was marked. The minimum follow up duration for patients was 6 months and the maximum was 18 months

RESULTS

Age of the patients ranged from 15 – 25 years. Three of our patients were female and one was male. All patients complained of a swelling in the palm with paresthesia involving the median nerve distribution. Preoperatively, sensation was S2 in three patients and S3 in one patient. Complete excision of the tumour was done in all. In two patients, a segment of sensory branch of median nerve was excised. In one patient, a 5 cm segment of common palmar digital nerve to second webspace was excised. [Figure 2]

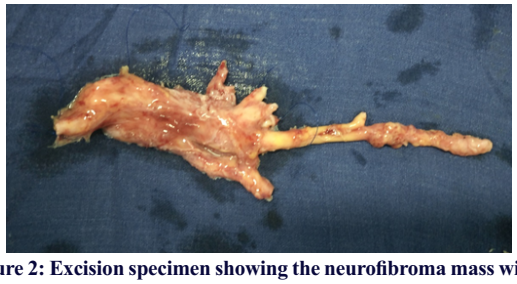

Figure 2: Excision specimen showing the neurofibroma mass with a 5 cm segment of common palmar digital nerve to second webspace.

In another patient, a 4 cm segment of common palmar digital nerve to the third webspace was excised. The histopathological report for all

patients was plexiform neurofibroma. All wounds healed without any postoperative wound complications. None of the patients exhibited any motor weakness postoperatively on day 5 ,14 and 21. Sensory examination was also done on 5th postoperative day. Two patients in whom all the branches were preserved did not experience any sensory deficit. In the two patients who underwent excision of sensory branches of median nerve, there was S1 sensation as per MRC scale. There was no improvement in the sensations on day 14 and 21. In the two patients who underwent nerve grafting, Tinel's sign was positive on day 21 and was progressive on subsequent examinations and progressed to fingertips by 5 months. Both patients showed improvement to S2 on day 90. At the end of 6 months, sensation was S3+ in all patients.

DISCUSSION

Neurofibromatosis was first described by the German pathologist Frederich Von Recklinghausen in 1882.[4] . It is a hereditary neurological disorder. Neurofibromatosis-1-or Von Recklinghausen's disease is an autosomal dominant disease transmitted on long arm of chromosome 17q11.2 and is mostly caused by mutation of NF-1 gene with incidence of 1 in 3000 live births. Neurofibromatosis-2 results mainly from a mutation of NF-2 gene and transmitted on chromosome 22 with incidence of 1 in 40,000 live births [5]

In 1987, a conference at National Institutes of Health had agreed to 7 points diagnostic criteria of patients with NF-1. [1]

1. Six or more café-au-laid spots larger than 5 mm in greatest diameter in prepubertal individuals and 15 mm in greatest diameter in post-pubertal individuals.
2. Two or more neurofibromas of any type or one plexiform neurofibroma.
3. Freckling in the axillary or inguinal regions.
4. Optic glioma.
5. Two or more Lisch nodules (iris hamartomas).
6. A distinctive osseous lesion, such as sphenoid dysplasia or thinning of the long bone cortex, with or without pseudoarthrosis.
7. A first-degree relative with NF-1 according to the above criteria.

Plexiform neurofibromas are benign peripheral nerve sheath tumors that can be seen in either type of neurofibromatosis. They are diffuse, large, elongated fibromas, histologically similar to discrete neurofibromas and commonly seen on the face and neck and sometimes can lead to massive enlargement of a limb or some other part of body (elephantiasis neuromatosa) [6, 7]. Their growth rate is highly variable. Often, overlying hyperpigmentation ("giant café-au-lait spot") or hypertrichosis can be seen [8]. In this tumor, the nerve is converted into convoluted mass and on palpation feels like "bag of worms". Isolated cases of plexiform neurofibroma without any association with NF1 have been reported in literature [9-11].

Plexiform neurofibroma is commonly seen involving the branches of trigeminal and cervical nerves over the face, but plexiform neurofibroma affecting the digital nerves in palm is very rare. Nagey et al. reported a neurofibroma of the palmar cutaneous branch of the median nerve, presenting as palmar mass with an overlying abnormality of the skin [12]. Basheer et al. reported four cases of neurofibroma affecting digital nerves and achieved excision of the tumor by meticulous dissection in three out of four cases [7]. Jones and Tonkin reported a unique case of 44-year-old gentleman presenting with a lump within his right palm that spanned several decades. This tumor turned out to be emanating from both ulnar and median nerves, the histopathological diagnosis of which was a plexiform neurofibroma [13]. This series of 2 patients presented with isolated mass on the wrist and palmar region without any other symptoms of neurofibromatosis.

Treatment of plexiform neurofibroma is primarily surgery. Surgical excision is probably the only therapy available because there is no medication that can prevent or treat plexiform neurofibromas. The results of surgical excision can also be poor and the procedures can be complicated due to the size, location, vascular status, neural involvement, microscopic extension of the tumor, and the high rate of tumor re-growth [9].

In our series there was good surgical recovery and post-surgical functions were good in all patients. There was no motor deficit and good recovery of sensory function after the surgery. This enabled them to lead their normal daily activities without any restrictions.

CONCLUSION

The Plexiform neurofibroma of the palmar region is a rare isolated tumour without any other signs of the disease complex and can be debilitating for the patients in terms of functions of the hand. Proper diagnosis, meticulous surgical technique and postoperative physiotherapy are necessary to produce a good postoperative result with adequate hand function.

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