e - ISSN - 2349 - 8005

INTERNATIONAL JOURNAL OF ADVANCES IN CASE REPORTS

Journal homepage: www.mcmed.us/journal/ijacr

PLEXIFORM NEUROFIBROMA OVER LEFT PALM: A CASE REPORT

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Article Info
Received 15/12/2015
Revised 27/12/2015
Accepted 10/02/2016

Key words:
Plexiform neurofibroma, Hyperpigmentation or hypertrichosis

ABSTRACT
Plexiform neurofibromas are benign peripheral nerve sheath tumours involving single or multiple nerve fascicles, often arising from branches of major nerves. They elicit a 'wormy' sensation on palpation. Often there is overlying hyperpigmentation or hypertrichosis. Plexiform neurofibroma over palm is rare & affects fine motor function of hand. Here, we report a case of plexiform neurofibroma over the palm

INTRODUCTION

Neurofibromatosis (NF) comprises a distinct genetic disorder characterized by benign growths of peripheral neurofibroma, nerve sheaths and café-au-lait macules (CALM), associated with various other cutaneous and systemic manifestations. Other diagnostic features of NF 1 include axillary or inguinal freckling, Lisch nodules, optic glioma, sphenoid dysplasia and NF 1 in a first degree relative (parent, sibling or offspring). The inheritance is autosomal dominant with variable penetrance. The gene for NF 1 is located in chromosome 17q and encodes for protein neurofibrin [1]. The incidence of NF1 is 1 per 2500-3300 births [2]. Riccardi has described many clinical types of neurofibromatosis.

CASE REPORT

A 25-year-old female presented to skin opd with single soft, asymptomatic swelling over middle of left palm of 14 years duration. The onset of the swelling was spontaneous and gradually progressive. There was no past history of trauma or surgery over left palm or similar complaints in family. Examination revealed a single 3×2 soft, light brown colored growth over left palm (fig 1) with feel of 'bag of worms' on palpation. CALM were present and there were no other nodules present on the body. Under local anaesthesia, excision of swelling was done and sent for HPE (histopathological examination). Histopathology of the tumor showed bundles of spindle-shaped cells (Schwann cells) arranged in whorl pattern in deep dermis (fig 2), which confirmed the clinical diagnosis of plexiform neurofibroma.

DISCUSSION

NF 1 is inherited as an autosomal dominant condition with incidence of 1 in 3000 live births
Diagnostic Criteria for Neurofibromatosis 1(NF1):
Two or more criteria must be satisfied for diagnosis [3]

1. Six or more CALM's >5 mm in the greatest diameter in prepubertal individuals; 15 mm in greatest diameter in postpubertal 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 thinning of the long bone cortex with or without pseudoarthrosis or sphenoid dysplasia
7. A first-degree relative (parents, siblings or offsprings) with NF-1 according to the above criteria.

Plexiform neurofibromas are generally present at birth or become apparent during first several years of life. It may lead to disfigurement, blindness or organ dysfunction by compression of vital structures. There may be overlying hyperpigmentation (‘giant *café-au-lait spot*’) or hypertrichosis [4]. The growth- rates of plexiform neurofibromas is highly variable. Periods of rapid growth alternating with long periods of quiescence are common. Histologically, they are benign nerve sheath tumors that appear as discrete masses arising from peripheral nerves and it consists of Schwann cells, mast cells, fibroblasts, and perineural cells.

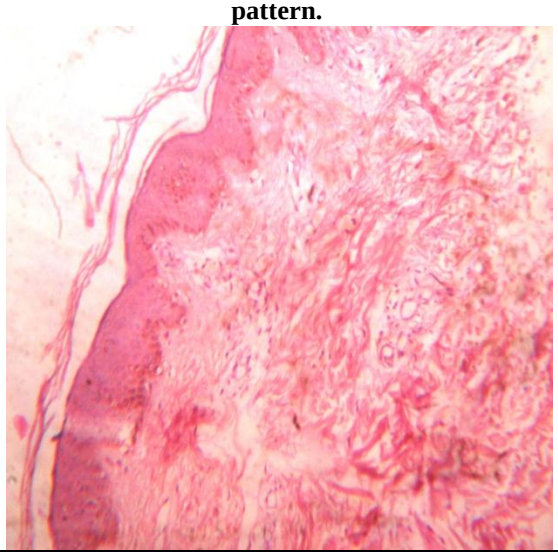
Plexiform neurofibromas are of two types, nodular and diffuse. The diffuse type is also known as elephantiasis neurofibromatosa.

The management of plexiform neurofibroma is aimed mostly at controlling symptoms. Surgical excision is probably the only therapy available so far.

Excision of benign plexiform neurofibromas is mostly very difficult due to tumor invading the surrounding structures and nerves and its inherent vascular nature. Life threatening hemorrhages are common with facial plexiform neurofibromas. Plastic surgeon’s advice is essential before removal. A number of agents (including, antiangiogenesis drugs, farnesyl transferase inhibitors and fibroblast inhibitors) are being used in various clinical trials to assess the drug’s effect on the growth of plexiform neurofibromas [5]. Currently, there is little evidence favoring the use of any of the drugs in patients with symptomatic plexiform neurofibromas.

Figure 1. Clinical photograph showing a single 3×2 soft, light brown growth over left palm.

Figure 2. Histopathological section showing bundles of spindle shaped cells arranged in deep dermis in a whorled pattern.



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