Scholars Journal of Medical Case Reports (SJMCR) **ISSN 2347-6559 (Online)**
Abbreviated Key Title: Sch. J. Med. Case Rep. **ISSN 2347-9507 (Print)**

A United of Scholars Academic and Scientific Society, India

Giant Neurofibroma of Right Cheek

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Article History
Received: 26.03.2018
Accepted: 13.04.2018
Published:30.04.2018

DOI:
10.36347/sjmcr.2018.v06i04.005

Abstract: Neurofibroma is a tumor of neural origin derived from the cells that constitute the nerve sheath. It's an uncommon benign tumor found as a solitary tumor or as a partial manifestation of neurofibromatosis type I (NF1, also called von Recklinghausen's disease). The cause of solitary neurofibroma is still unknown; it is defined by Marocchio *et al.* as a hyperplastic hamartomatous malformation rather than neoplastic. NF1 is an autosomal dominant genetic syndrome caused by mutations in genes coding for neurofibromin, it is characterized by cutaneous manifestations as cafe-au-lait spots with a large number of nervous system tumors. The hereditary factors and systemic symptoms present in the disseminated neurofibromas are absent in the solitary type.
Keywords: Neurofibroma, cheek, benign tumour.

INTRODUCTION

A neurofibroma is a benign nerve sheath tumor in the peripheral nervous system. In 90% of cases they're found as stand-alone tumors, while the remainder are found in persons with neurofibromatosis type I (NF1), an autosomal dominant genetically inherited disease, they can result in a range of symptoms from physical disfiguration and pain to cognitive disability.

Neurofibromas arise from nonmyelinating-type Schwann cells that exhibit biallelic inactivation of the NF1 gene that codes for the protein neurofibromin [1]. This protein is responsible for regulating the RAS-mediated cell growth signaling pathway. In contrast to schwannomas, another type of tumor arising from Schwann cells, neurofibromas incorporate many additional types of cells and structural elements in addition to Schwann cells, making it difficult to identify and understand all the mechanisms through which they originate and develop.

CASE REPORT

A 45 year old male came to surgery OPD with swelling over right cheek from past 15 years. He complains of initially the swelling was small and gradually increased to present size of 19 x14 cm in diameter [Figure 1]. Clinically the y diagnosed it as nerve sheath tumour and sent for Fine needle aspiration cytology (FNAC). FNAC was done using 23 guage needles and hemorrhagic material was aspirated. On cytology we reported it as benign spindle cell lesion and advised for biopsy. Surgery was done and specimen was sent for histopathological examination. Grossly we recieved a large specimen measuring around 20x14x10 cm diameter, which is soft to firm in consisitency. Cut secion show geryish white areas, multiple sections were taken and submitted. On microscopy showed many spindle to elongated cells with tappering nuclei, which resembled the appearance of shredded carrot like. No evidence of necrosis or mitotic figures identified [Figure 2]. It was a diagnosed as Neurofibroma of cheek.

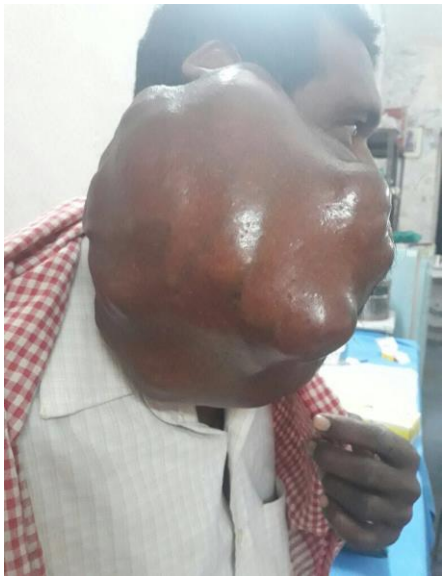

Fig-1: Clinical photograph of swelling over right cheek

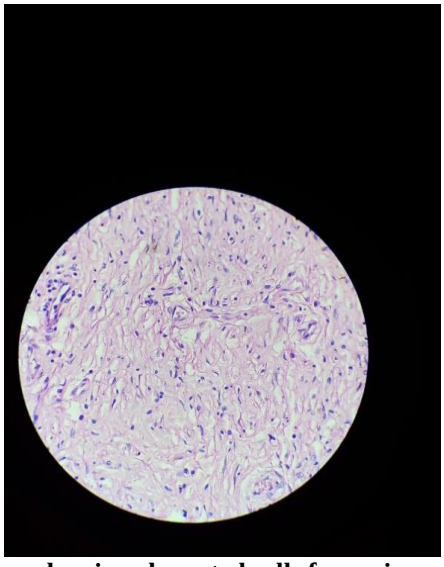

Fig-2: Sections showing elongated cells favouring neurofibroma

DISCUSSION

Neuro fibroma may occur as a solitary, fusiform cutaneous tumour of a single nerve, but more often are multiple associated with Von Recklinghausen's disease. Solitary neuro fibroma is a tumour of adults but multiple neurofibromas or neurofibromatosis is a hereditary disorder with autosomal dominant inheritance. Solitary Neuro fibroma is generally asymptomatic but patients with Von Recklinghausen's disease have a triad of features like multiple cutaneous neuro fibromas, numerous pigmented skin lesions and pigmented iris hamartomas. Neurofibromatosis type 1 is a genetic disorder having mutation in chromosome 17 while type 2 has mutation in chromosome 22.

Neuro fibroma is known to mainly affect the cutaneous nerves of the trunk, neck, and head. Rare cases in the deep organs or in the peritoneal cavity were also reported. Concerning the retroperitoneal location, exceptional cases of benign nerve sheaths tumors were reported in large series of primitive retroperitoneal tumors. To our knowledge, the presence of solitary neuro fibroma in the preperitoneal space has never been reported in the literature [2, 3].

Neuro fibroma appears to affect adolescents and young adults without a gender preference. It grows along the peripheral nerves as a non-encapsulated focal mass with well-defined margins.

The clinical manifestations of solitary neurofibromas are not specific and change according to their locations, their effect on gastrointestinal motility and their possible impingement on contiguous structures, resulting in palpable masses, abdominal pain, and transit disorders due to extra-luminal pressure. The deep seated neurofibroma on peripheral nerves and

spinal roots frequently leads to neurological disability [4].

Neurofibromas have tendency for local recurrences after excision. Neurilemmoma virtually never turns malignant, while sarcomatous transformation in neurofibroma, particularly in neurofibromatosis, is not unusual. It is estimated that about 3% of patients with Von – Recklinghausen's neurofibromatosis develop malignant transformation of one of the nodules. Rarely, neurogenic sarcoma may develop spaontaneously in the absence of pre-existing Von –Recklinghausen's disease [5].

Malignant Peripheral nerve sheath tumour is a poorly differentiated spindle cell sarcoma of the peripheral nerves occurring most often in the adults. The tumour may arise devo or from malignant transformation of a pre-existing neuro fibroma than a schwannoma, generally at an early age. About 50% of the tumours are seen in patients with neurofibromatosis type 1with chromosomal deletion 17p and p53 gene mutations, while some develop at sites of previous irradiation.

Preoperative imaging is usually insufficient to establish the diagnosis with certainty, only the histology can do. In fact, the preoperative histological evidence can influence the treatment modifying the surgical approach, conservative versus aggressive. Surgical removal is the only treatment option.

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