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PLEXIFORM NEUROFIBROMA OF CHEEK- A RARE CASE REPORT

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ABSTRACT Plexiform neurofibromas (PNs)are benign tumors that usually originate from the nerve sheath of branches of visceral or subcutaneous peripheral nerves. It is a least common form of neurofibromatosis which occurs in 5% of cases. Plexiform neurofibroma is usually recognized as a pathognomonic criterion of Neurofibromatosis-1; it may also occur as a solitary lesion arising in a nerve root. Although they are benign, they have a 2%–5% potential for malignant transformation. Here, we present a case of 9 years old female who presented with a left cheek plexiform neurofibroma without any other signs of Neurofibromatosis 1, since a period of 4 months. Patient was operated and the tumor mass was completely excised. The patient is now on follow up and no signs of recurrence have been recorded till present.

KEYWORDS : Plexiform neurofibroma, Neurofibromatosis 1

INTRODUCTION

Neurofibroma is a disease of the peripheral nervous system and occurs most commonly in the extremities. Several forms have been described: cutaneous neurofibromas (both localized and diffuse types), intraneural neurofibromas (localized and plexiform), massive soft tissue neurofibromas (solitary or multiple), and sporadic neurofibromas or those associated with neurofibromatosis-1 (NF-1) [1,2]

Plexiform neurofibromas (PNs) are benign tumors that usually originate from the nerve sheath of branches of visceral or subcutaneous peripheral nerves. It is a least common form of neurofibromatosis which occurs in 5% and is suspected to arise during gestation which can be an early source of disfigurement, disability, and lethality [3]. They are often described as rare histological variants of neurofibroma (NF), consisting of a proliferation of cells in the nerve sheath and involving multiple nerve fascicles. Plexiform neurofibroma is usually recognized as a pathognomonic criterion of NF-1 (or Von Recklinghausen's disease); it may also occur as a solitary lesion arising in a nerve root [4]. Usually described as a "bag of worms," PNs are non-circumscribed, baggy and thick in shape.

The pediatric population is favorably affected since PNs usually grow in early childhood at variable rates. A minority of tumors in the head and neck are as of children stemming from either the peripheral portion of the facial nerve within the parotid gland or its terminal branches [4].

Approximately 50% of children with internal PNs have tumor-related symptoms in early childhood that correlate with tumor size and location [5].

Although they are benign, they have a 2%–5% potential for malignant transformation [6]. Patients with NF-1 and plexiform neurofibromas have a higher mortality rate when compared with patients with or without asymptomatic plexiform neurofibromas [7].

HISTOPATHOLOGY

Macroscopically, the tumor appears as a solid, tan-gray, glistening mass that is translucent to light [8]. Histopathologically, the pattern may be that of distorted masses of myxomatous peripheral nerve tissue still within the peripheral sheath, seen scattered within a collagen rich matrix. This is considered to be virtually diagnostic of NF1, even in the absence of other manifestations.

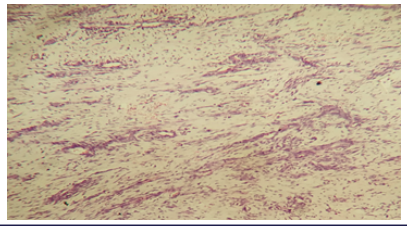

FIG 1. HPE OF SPECIMEN

S -100 is a useful marker for schwann cells, and the lesional cells are uniformly positive signifying that they originate from neural crest derived tissue [9]. EMA-positive staining for perineural cells is always suggestive of plexiform neurofibroma [10].

CASE REPORT

HISTORY

We present a case of 9 years old female who presented with a left cheek swelling since a period of 4 months. It was insidious in onset, initially there was reddening of the skin over the left cheek and size had increased progressively within a short duration. It was not associated with pain, tenderness, discharge or bleeding from the area, no history of fever, not associated with difficulty in swallowing or opening of mouth. There is no history of any such swelling in any other part of the body. There was no history of deviation of angle of mouth or difficulty in closure of eyes. No history of loss of weight. There is no history of any major illness in the past. There is no any significant family history.

CLINICAL EXAMINATION

There was a solitary swelling over the left cheek, size approx.- 5 x 4 x 3 cm. Skin over the swelling appeared normal, not adherent to underlying structures. It was firm in consistency, regular well- defined margins, mobile, non-reducible, non-pulsatile, no tenderness, no local rise of temperature.

- Cervical lymph nodes were non-palpable. Facial nerve examination was normal.
- There were no significant findings in examination of oral cavity, Ear, Nose and Throat.
- There were no any significant swellings of café-au lait spots in any other part of the body.

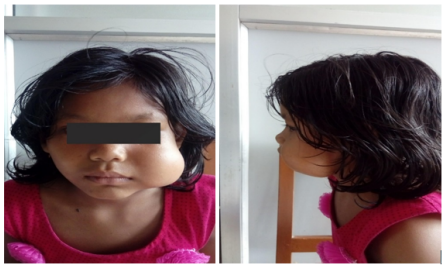

FIG 2. **FIG 3. PREOPERATIVE PHOTOS**

INVESTIGATIONS-

Routine blood investigations (Routine blood examination, platelet count, Blood sugar, S. Creatinine, Bleeding profile, Viral markers) were done and all were found to be within normal limits. FNAC of the

aspirate from the swelling was done and smears showed predominantly singly lying short spindle shaped cells having scant cytoplasm and round to oval hyperchromatic nuclei, which was suggestive of Benign spindle cell lesion.

High resolution Ultrasonography of the swelling was done in which it was observed to be a well- circumscribed oval shaped mass lesion of approximately 5.5 x 3.8 cm in the left cheek. It was primarily in the buccal pad of fat. The adjacent muscles are displaced. No obvious adjacent structure infiltration, bony erosion or internal vascularity was noted. It was separate from parotid and submandibular glands and showed a possibility of plexiform neurofibroma.

OPERATION

Patient was operated under General anesthesia and excision of the mass was done. Incision was given from behind the tragus along the angle of mandible to the chin on left side. Mass was seen to be separate from parotid tissue, it was predominantly present in the buccal pad of fat, extending to the pterygopalatine fossa above the medial pterygoid. Facial Nerve and its branches were preserved. Specimen was sent for Histopathological examination.

FIG 4.

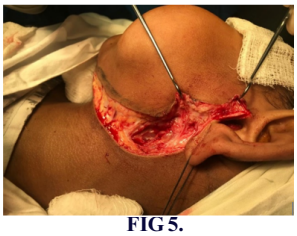

FIG 5.

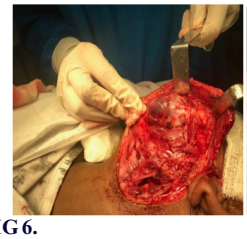

FIG 6.

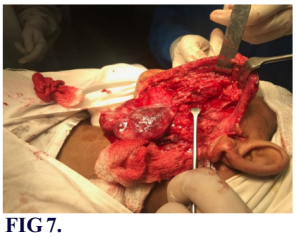

FIG 7.

INTRAOPERATIVE PHOTOS

FIG 8.

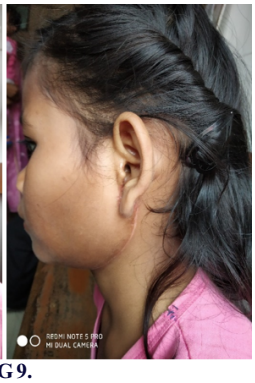

FIG 9.

POST-OPERATIVE PHOTOS

FOLLOW UP

Patient was discharged postoperatively on 5th day and is now on regular follow up. Postoperative wound is healthy and patient doesn't have any fresh complaints.

Postoperatively HPE mentioned an encapsulated mass, whitish, homogenous with few brownish areas. On microscopic examination findings were suggestive of neurofibroma.

DISCUSSION

Plexiform neurofibromas represent 14% of all benign mesenchymal tumours and 10% of non-epithelial salivary gland tumours [11]. The term "plexus" refers to a combination of interlaced parts or a network. Plexiform neurofibromas are uncommon and occur almost exclusively in about 5-15% patients with neurofibromatosis-I. Two types of plexiform neurofibromas have been recognized: (a) a diffuse type/ elephantiasis neurofibromatosa and (b) a nodular type [12]. Plexiform neurofibromas demonstrate an intricate microenvironment composed of irregularly growing Schwann cells, large numbers of degranulating mast cells, perineural cells, collagen-secreting fibroblasts, and blood vessels. To date, management of progressive symptomatic PNs remains medically challenging. PNs appear to be radio-insensitive tumors, and radiation is avoided given the known risk of radiation-induced secondary malignancies [13]. In this case complete excision of tumor mass was done and no other adjuvant treatment was prescribed. Recurrence is reported in 20% of the patients with plexiform neurofibroma after complete resection and increases to 44% with incomplete resection (when there is involvement of vital structures) [14]

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