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Surgical Management of Plexiform Neurofibroma: Case Report

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Abstract Neurofibroma is an autosomal dominant, multi-system disorder affecting approximately 1 in 3500 people. We present a case of plexiform neurofibroma of the head and neck and elucidate our experience with the surgical resection and reconstruction to restore the patient's quality of life, boost confidence and exclude the possibility of malignant transformation.

Keywords: Neurofibromatosis, Microvascular Reconstruction, Head-And-Neck Surgery

Introduction

Neurofibromatosis (NF) first described in 1882, is an autosomal dominant, multi-system disorder affecting approximately 1 in 3500 people1, which makes it one of the most common genetic diseases in man; There is a positive family history in only half the cases 2. It presents in the third or fourth decade of life 3. The penetrance approaches 100% by age 20; If the patient has the mutation, they will exhibit manifestations1. 90% of cases of NF are NF14. They arise from non-myelinating type Schwann cells exhibiting biallelic inactivation of NF1 gene that codes for protein neurofibromin.

The incidence of head and neck involvement in this disease has been reported from 22%-47%5. Intra-oral involvement is not common, Preston et al. (1952) quotes 6%, Farmer and Lawton (1966) 2%, while Gorlin et al. (1976) put it at between 4-7% of cases6. Tongue is the commonest intra-oral site, closely followed by intra-osseous lesions, followed by buccal

mucosa, palate, and gingiva, all seem to be affected in approximately equal proportions6.

Its presence in the head and neck interferes with the airway, eyelids, vision, mastication, and lip competence, in addition to producing a cosmetic deformity. Sarcomatous transformation of NF1 has been reported, particularly when associated with von Recklinghausen's disease, incidence being placed approximately 15%7.

Case Report

A 26-year-old male patient visited the hospital with the chief complaint of a swelling on the left side of the face present since birth and gradually increased in size.

History showed, the patient was born with facial asymmetry on the left side of the face. The first swellings were on the left upper eyelid and upper lip, for which he underwent a debulking surgery at the age of 5. Two years after surgery he noticed an increase in size, now involving his left cheek, left forehead along with the previously involved regions. Following which he underwent a second debulking surgery at the age of 7. Histopathological diagnosis showed the presence of neurofibromatosis involving his maxillary branch (V2) of trigeminal nerve (CNV). He did not give any history of pain, injury or bleeding from that site. He gave a history of multiple pigmented spots, brownish black in colour all over his body. Motor and sensory function of facial nerve (CNVII) was present on the affected side. There was a history of an open posterior fontanelle since birth.

There were two swellings seen on examination of the left side of the face:

1. A solitary diffuse swelling, on the left supraorbital region involving the glabella extending just 2 cm below the zygomatic arch, involving the nose, ovoid in shape, brownish-black in colour, with irregular margins, soft in consistency and non-tender. No pulsations were felt, no secondary changes were seen and the skin over the swelling was pinchable. The left eyeball was present but without vision and there was complete ptosis of the upper eyelid. The nose appeared raised and deviated to the right. There was obliteration of the nasolabial fold on the left side.
2. A solitary diffuse swelling with ill-defined margins, crossing the midline was present on the upper lip without any deviation of the mouth, brownish-black in colour and of irregular shape; Soft in consistency with no tenderness or secondary changes. Skin was pinchable and no pulsations were felt. There was complete ptosis of the upper and lower lip on the left side.

The mouth was shaped irregularly with considerable drooping of the left angle of the mouth. The mouth opening was adequate. An isolated diffuse swelling was present on the hard palate measuring 7x5 cm. It had ill-defined margins, smooth surface, was non-tender and firm in consistency. It was non-fluctuant and showed no secondary changes. Teeth numbers 21, 22, 27 and 28 were missing and 23 seemed to be displaced.

The patient was advised routine blood and radiographic investigations for pre-operative workup. Computed Tomographic (CT) scan showed thinning of the left calvarium; There was sphenoid wing dysplasia, displacement of left orbit anterolaterally and buckling of optic nerve was noted. Magnetic Resonance Imaging (MRI) brain showed a large, polypoidal soft tissue density seen cranially reaching upto the left supra-orbital ridge and caudally upto the angle of mandible with thickness of 2.5 cm. The left globe appeared deformed and small in size.

With two surgical teams working simultaneously, an excisional biopsy with left partial maxillectomy and orbital enucleation was done under general anaesthesia. Incision was taken along the neck crease on the left side to expose and prepare the internal jugular vein; Optic nerve and ophthalmic artery were ligated postero-medially, followed by infrastructural maxillectomy. Pterygoid plexus haemorrhage was encountered and managed with bipolar cautery and 3-0 figure of 8 silk sutures. 15x15 cm tumour mass was excised and sent for histopathological evaluation. The resulting defect was covered with a left anterior lateral thigh flap and anastomosed to the superior thyroid vessels along with a left saphenous vein graft. Once perfusion to the flap was confirmed, a number 7 endotracheal tube was cut, sized and placed in the left nostril to maintain patency. Inset was done, two negative suction drains were attached followed by closure with 3-0 ethilone sutures. Elective tracheostomy was performed with a number 7 tracheostomy tube. The estimated blood loss was 4.5-5L for the entire surgery. Patient was shifted to the ICU for monitoring until homeostasis was achieved. The patient's recovery was uneventful and discharge was given on post-op day 20. Histopathological impression suggested diffuse neurofibroma.

Correlating the history, clinical, radiographic and histopathological findings, the diagnosis concluded was plexiform neurofibroma in a known case of neurofibromatosis type 1.

Discussion

Our goal of treating this case of plexiform neurofibroma with surgery is to provide information and details which can aid other surgeons in operating such patients.

Studies have shown that partial excisions at an early age is of limited value, which can lead to more aggressive treatment of soft tissue involvement4. Even for our patient, who had undergone two debulking surgeries at a very early stage, resulted in a progressive mass compressing the adjacent vital structures.

Trigeminal nerve neurofibromas account for only 0.26% of all intracranial tumours and 2.9% of the intracranial neurofibromas(Schisano and Olivecrona,1960)3. To our knowledge, a trigeminal nerve neurofibroma of the main trunk has not been reported in the oral and maxillofacial surgery literature7. In our case the ophthalmic and maxillary divisions of CNV were involved.

Hunt and Pughi2 reported skeletal changes in 39% of their cases, whereas Casselman and associates found changes in 71% of their patients.

In majority of the cases, neurofibromas on or adjacent to the cranial nerves also affect motor function of the facial and hypoglossal nerves and sensory function of the trigeminal nerve2, however this was not true in our case.

Even though Baden and Fischer2 found that the majority of intraoral neurofibromas occur in the tongue, we noticed a palatal swelling.

At present, no gold-standard treatment exists for NF. Surgery is the only modality that has been shown to achieve at least temporary control of tumour growth and symptom progression8. We attempted to provide sufficient soft tissue coverage using microvascular reconstruction and noted excellent results with long-term follow-up. From the patient's point of view, they stated that they would undergo further aesthetic improvements via reconstruction of upper left eyebrow and prosthesis for the eye and teeth.

The role of radiotherapy and chemotherapy as adjuvant therapies remains unclear. In one study patients with NF-associated with CNS tumours, most of them(67%)

received stereotactic radiosurgery (range, 1,000–2,400 cGy) and stated that radiation therapy should be considered in NF patients with imaging progression of CNS tumours9. Chemotherapy has been used singly or in combination. In a recent phase 1 trial, pegylated interferon-alfa-2b was assessed in 30 patients with plexiform neurofibromas; Reported effects were pain reduction, decreased tumour mass, and tumour shrinkage or stabilisation. Similarly, in a phase 2 trial of imatinib, 17% of patients with plexiform neurofibromas had a 20% or more reduction in tumour volume10.

We concluded that single stage surgery for the primary tumour—taking care of massive haemorrhage in comparison to piecemeal multiple procedures causing patient physical and psychological anguish—should be the way to treat these tumours with further improvements via facial reanimation to restore the patient's quality of life, boost confidence and exclude the possibility of malignant transformation.

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Legend Figure

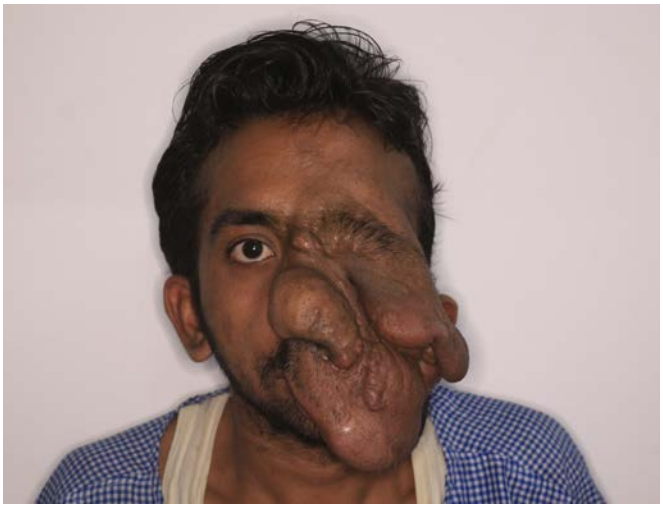

Figure 1: Preoperative Image

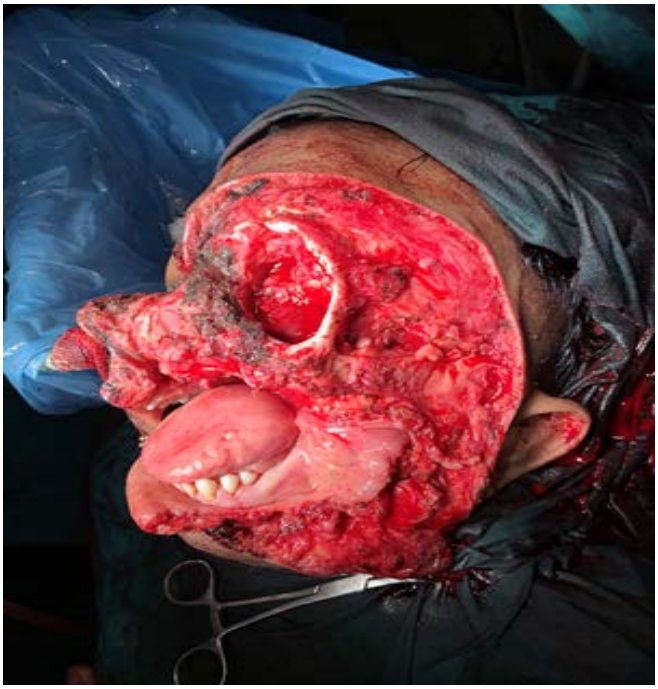

Figure 2: Resulting Defect after Near-Total Excision

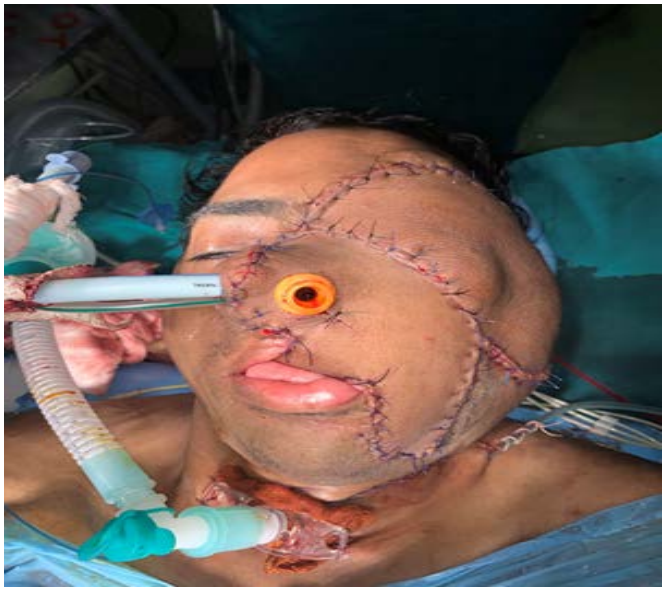

Figure 3: Intraoperative Image after Reconstructing With Free ALT Flap

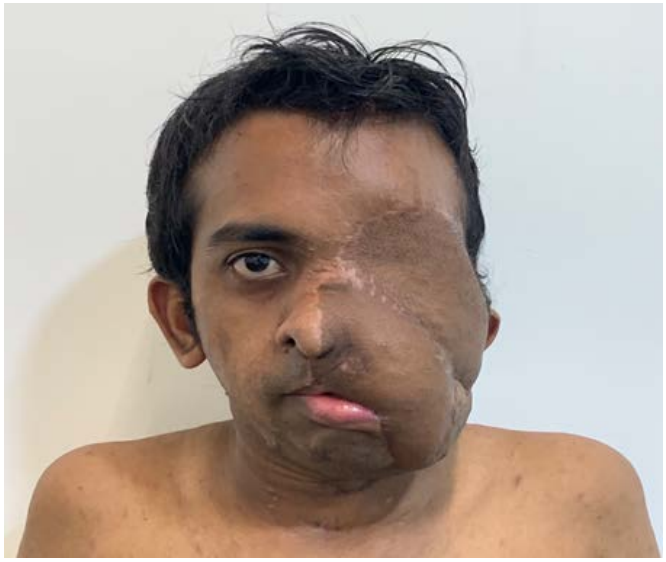

Figure 4: 6 Months Postoperative Photo