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Case Report

Surgery Section

Neurofibroma of Face: A Challenge in Aesthetic Reconstruction

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ABSTRACT

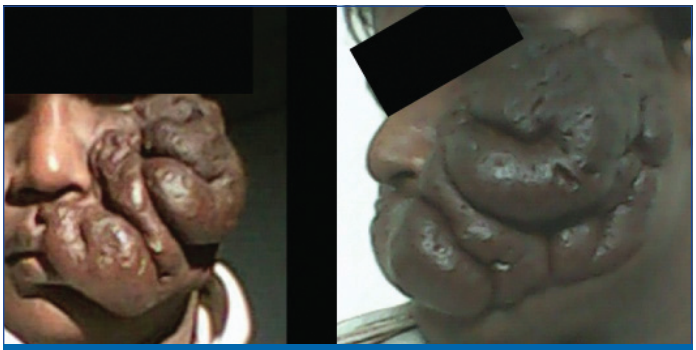
Plexiform Neurofibroma (PNF) presents as a growth of unexplained excess that does not pertain to any other organic causes and is hereditary. The iconic 'bag of worms' appearance is native to its diffuse and pliable character. Region of occurrence being the face adds the challenge of aesthetics to infiltration into vital structures and recurrence rate. The patient presented here is that of serial reconstructions done in order to satisfy the aesthetic demand of the patient. Initial reconstruction done with split thickness skin graft was later augmented with radial forearm flap. No signs of recurrence were seen during the follow-up of six years.

Keywords: Facial neurofibroma, Plexiform neurofibroma, Radial free forearm flap, Split thickness skin graft

CASE REPORT

A 23-year-old male patient has reported with a growth over left side of face since birth. The patient was born with a small, painless growth over left side of face which gradually increased to its present size. It occasionally bleed on manipulation.

On examination, there was a large flappy plexiform growth (16 x 14 cm approximately) with multiple folds present on the left side of face, with ill-defined edges and blackish to reddish brown colour [Table/Fig-1]. No signs of regional involvement were noted on clinical examination. The patient had history of surgical excision of part of lesion at 18 years of age. A differential diagnosis of plexiform neurofibroma and neurovascular hamartomatous lesion was formulated. An ultrasonographic evaluation was done, and a radiological diagnosis of Plexiform Neurofibroma was ascertained.



[Table/Fig-1]: Pre-operative showing the facial growth.

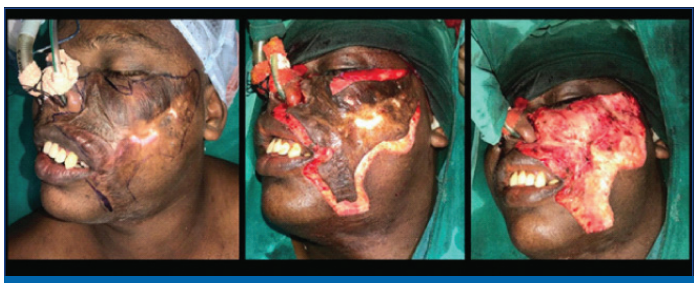
Patient was then planned for surgical intervention after obtaining due written consent for the procedure and its probable outcomes. Keeping in mind the high recurrence rate and aesthetic demand of the involved region, a two stage procedure was planned [1].

Stage I

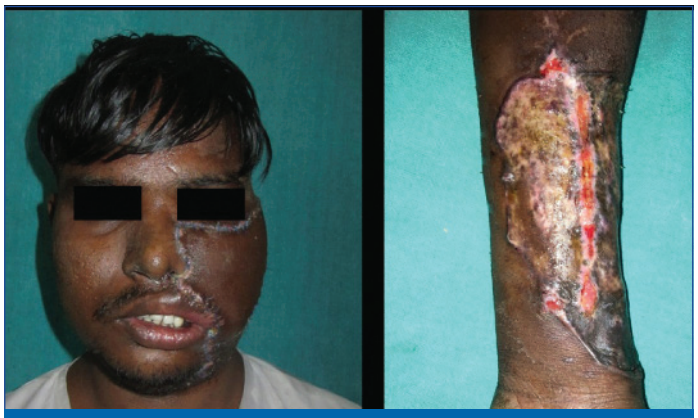
This stage aimed at complete disease removal with minimal postoperative morbidity. Wide local excision of the lesion and reconstruction with split thickness skin graft harvested from left thigh was done [Table/Fig-2]. The patient maintained a regular follow-up.

Stage II

After three years of follow-up, no signs of recurrence were encountered and patient was recalled for second stage of surgery. This stage aimed at fulfilling the aesthetic demand of the site and correction of the loss of bulk due to previous surgery and postoperative fibrosis. Citing these requirements, a vascularised free flap was considered due to its optimum bulk and ease of draping along facial contours. Surgical excision of fibrotic areas of skin and replacement with radial forearm flap was done [Table/Fig-3].



[Table/Fig-2]: Split thickness skin graft placed in 'Stage I' being excised to expose a healthy bed free of recurrence for 'Stage II' surgery (3 years after 'Stage I').

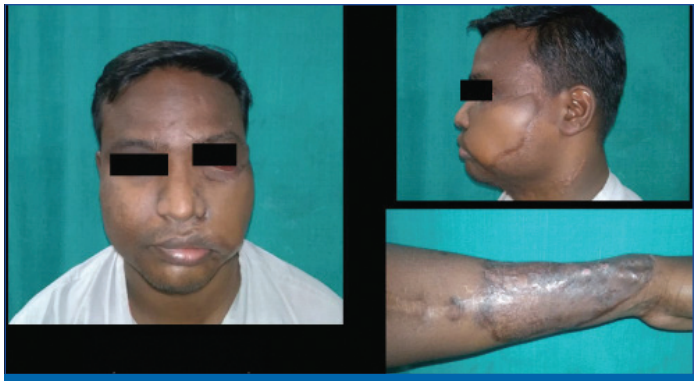


[Table/Fig-3]: Second stage surgery: Postoperative progress after 'Stage II' surgery using radial forearm flap.

After three years, no recurrence of disease was noted but there was contracture of scar around the lower eyelid causing ectropion, for which V-Y Closure of the lower eyelid was performed [Table/Fig-4,5]. The patient was satisfied with the overall outcome with no recurrence in six years and was advised regular follow-up.

DISCUSSION

It is a neuroectodermal abnormality involving skin, nervous system, bones and the eyes, first described in 1882 by Friedrich Daniel Von Recklinghausen. The oldest documented case of Neurofibroma appears in XIII and XVI centuries and was described by Madigan, Schaw, and Masello in 1988 in "Neurofibromatosis in the 13th Century and Report of NF-Like Case-Monstrorum History"- For a hereditary disease spanning



[Table/Fig-4]: Post-operative recovery following 'Stage II' surgery after three years.

[Table/Fig-5]: Ectropion of lower left eyelid due to contracture and bulk of flap; correction by V-Y closure.

so far past into documented medical history, the inquisition into its management is fairly new [2].

Onset being neonatal or early childhood and prevalence of one in every 3000 live births was observed [3]. The mutation of tumor suppressor neurofibromin 1(NF1) gene (17q11.2) and 17q11 microdeletion (only 5%) is the root cause [4]. Isolated plexiform neurofibroma may not be traceable in the familial history if not related to NF1. No family history of similar features could be traced in this case.

Differential diagnosis of such growths can be Schwannoma, Traumatic neuroma , Neurotized nevus, Superficial angiomyxoma, Nerve sheath myxoma , Malignant peripheral nerve sheath tumor (MPNST) , Spindle cell lipoma [5].

Surgical planning of these procedures is a long drawn out process as complete excision is not possible in a single operative sitting leading to the patient requiring numerous procedures over time. Furthermore, tumours involve and infiltrate multiple tissue planes making resection sketchy, causing disfigurement and at times, could be as extensive as hemifacial hypertrophy [5-7].

The gold standard treatment for these tumors remains surgical, but functional disturbances can be worrisome when resecting them involving the craniofacial area [8]. No chemotherapy has successfully reduced the size of such tumors till date [9]. An imminent recurrence rate is associated with the commonest treatment option resection and debulking. A series of reports in paediatric population noted recurrence in 20% of complete resection and 45% in incomplete resections [6].

Vascularity is the limiting factor aiding to its propensity to bleed extensively because of the friability of newer vessels. Adequate arrangements for the blood transfusion must be ensured before planning for resection. Tumescent technique of infiltration was said to be best in decreasing haemorrhage, creating a bloodless field for dissection [10]. Following excision of the excess of tumor mass, redraping of skin flap to be done and few anchoring sutures between the flap and the periosteum underneath should be placed to mitigate the pulling down of the soft tissues due to gravity post operatively [9,11].

In Proprioceptive Neuromuscular Fascilitation (PNF) of the head and neck region, the resection often leads to esthetic impairment and functional debilitation [12]. It is the duty of the head and neck surgeon to reestablish a near normal state in the postoperative period. For this, with the advancement of scientific knowledge and technical development, free tissue transfer has become possible. Various sites in the human body are suited for this technique, and the selection of a donor site depends on the type of tissue to be replaced and the patient factors.

The radial free forearm flap (RFFF) is one of the first free tissue transfer flaps to be described in literature. Yang GF et al. in 1981 described it for the first time and it has since become a workhorse for soft tissue replacement in craniofacial oncosurgery, being mostly used to replace external skin and mucosal linings [13]. It is an extremely versatile flap allowing intricate folding of the skin, using two or more skin paddles/islands, and incorporating vascularised tendon and/or bone (osseocutaneous flap). Due to its lack of bulk and predictable vascularity, the free flap was the best choice for aesthetic reconstruction of the face. The anatomy of the flap allows for it to be draped over uneven bases and to be contoured as per the will of the surgeon following anastomosis [14].

A study conducted by Fang G Q et al compared the reliability of the RFFF with the platysmal flap. Buccal contouring was comparably similar but the RFFF flap group showed less change in interincisal mouth opening as compared to the Platysma Flap (PF) group [15].

Wise J B et al., [16], with 10 patients of trigeminal nerve involvement amongst 39 who underwent cumulative 11 surgical procedures. Friedrich R E et al observed nine cases of type 1 PNF in paediatric patients who had no neurological or functional deficit. They carried out total resection and advocate total resection in smaller tumors as an prophylactic strategy to mitigate future disfigurement and functional deficits [7].

NF has a 5% malignant transformation rate and a recurrence rate as high as 15%. Over the six years follow-up period, no new growth and any sign of recurrence was noted, in the index case. The patient was satisfied with the aesthetic outcome.

CONCLUSION(S)

The challenges of plexiform neurofibroma of the face are its inherent high recurrence rate and the aesthetic demand of the facial region. These challenges were overcome by a perspective treatment plan with serial surgeries spanning over five years, and a keen regular follows up to achieve the desired outcome.

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