Facial Reanimation of Patients with Neurofibromatosis Type 2

Kalpesh Vakharia, M.D.[1], Doug Henstrom, M.D.[1], Scott R. Plotkin, M.D., Ph.D.[2],
Mack Cheney, M.D.[1], and Tessa A. Hadlock, M.D.[1]

[1] Department of Otolaryngology / Head & Neck Surgery, Massachusetts Eye and Ear Infirmary and Harvard Medical School, Boston, MA.
[2] Department of Neurology and Cancer Center, Massachusetts General Hospital and Harvard Medical School, Boston, MA.

Abstract

Objectives/Hypothesis: Neurofibromatosis type 2 (NF2) is a tumor suppressor syndrome defined by bilateral vestibular schwannomas. Facial paralysis--either from tumor growth or from surgical intervention--is a devastating complication of this disorder and can contribute to disfigurement and corneal keratopathy. Historically, physicians have not attempted to treat facial paralysis in these patients. We review our clinical experience with free gracilis muscle transfer for the purpose of facial reanimation in patients with NF2.

Study Design: Retrospective case series.

Methods: Five patients with NF2 and complete unilateral facial paralysis were referred to the facial nerve center at our institution. Charts and operative reports were reviewed; treatment details and functional outcomes are reported.

Results: Patients were treated between 2006 and 2009. 3 patients were men and 2 were women. The age of presentation of debilitating facial paralysis ranged from 13 to 50 years old. All patients were treated with a single stage free gracilis muscle transfer for smile reanimation. Each obturator nerve of the gracilis was coapted to the masseteric branch of the trigeminal nerve. Measurement of oral commissure excursions at rest and with smile, preoperatively and postoperatively revealed improved, and near symmetric smile in all cases.

Conclusions: Management of facial paralysis is often times overlooked when defining a care plan for NF2 patients that typically have multiple brain and spine tumors. The paralyzed smile may be treated successfully with single stage free gracilis muscle transfer in the motivated patient.

Introduction

•Neurofibromatosis type 2 (NF2) is an autosomal dominant, multisystem disease predisposing patients to lesions of the skin, eyes and nervous system.
•Mutations of the NF2 tumor suppressor gene on chromosome 22 result in wide phenotypic variability and nearly 100% penetrance by age 60 years of age [1, 2] .
•The hallmark of NF2 is the development of bilateral vestibular schwannomas; however, other cranial, spinal and peripheral nerves can be affected. Other benign tumors such as meningiomas, ependymomas, astrocytomas and neurofibromas can occur in patients with NF2.
•Unfortunately the disease and its treatment can result in significant morbidity and an earlier mortality for the NF2 patient [3].
•Facial palsy and paralysis can be a manifestation of the disorder or a consequence of its treatment.
•Functionally, these patients can have difficulty with nasal breathing, oral intake and competence, speech, and eye protection. Disfiguring, facial weakness can lead to a decrease in the quality of life.
•Despite the significant impact of facial paralysis on these patients, little attention has been given towards the treatment of this morbidity in patients with NF2.
•The objectives of this study were to review our experience in facial nerve rehabilitation of patients with NF2 and evaluate oral commissure excursion after free gracilis muscle transfer.

Methods

Patient and Data Collection
•A retrospective review was performed of all charts of patients with NF2 who were treated surgically between 2006 and 2009 at the Facial Nerve Center at Massachusetts Eye and Ear Infirmary, Harvard Medical School. The institutional review board at the Massachusetts Eye and Ear Infirmary approved the study.

Operative Technique
•All patients received a single staged procedure via a two teamed approach by the senior authors (MC and TH).
•The gracilis muscle was implanted in the paralyzed side of the face with one end of it secured to the lateral oral commissure and the other end to the true temporalis fascia.
•Muscle revascularization was performed with the facial artery and vein and re-innervation was done by coapting the obturator nerve to the masseteric branch of the trigeminal nerve.

Methods (cont.)

Smile Analysis
•Preoperative and postoperative photographs were used to assess oral commissure movement.
•The SMILE is a validated oral commissure movement measuring tool (publication pending).
•Parameters of oral commissure movement were measured: x, y, z, and angle are presented as mean ± standard deviation.
•A two-sided Student's t-test was used; P value < 0.05 is significant.

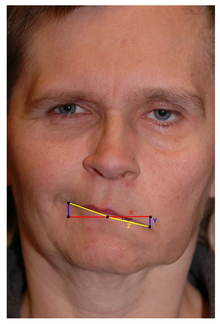

Figure 1. Oral commissure parameters: x,y,z, angle.

Results

•5 patients (2 women, 3 men) with NF2 and unilateral facial paralysis (House-Brackmann 4-6) were identified; they had a age range 13 – 50 years.
•Four of the patients had developed facial paralysis in the immediate or delayed postoperative period after treatment of their tumors. One patient had insidious onset of facial paralysis prior to any treatment of her disease. All had no oral commissure movement.
•There were no significant postoperative complications or surgical site infections.
•Three out of the five patients underwent postoperative physical therapy.
•Figure 2-5 demonstrate oral commissure excursion measurements as defined by changes in parameters x, y, z, and angle as calculated by the SMILE application.
•There is a significant difference between all preoperative and postoperative smile parameters on the rehabilitated, paralyzed side of the face ($p < 0.05$)
•The unaffected side of the face does not show any significant difference between preoperative and postoperative smile parameters ($p > 0.05$).
•The rehabilitated paralyzed side of the face achieved similar values in postoperative smile parameters as the normal side of the face.
•All patients, during their postoperative visits, reported that they were pleased with the resulting smile and facial symmetry.

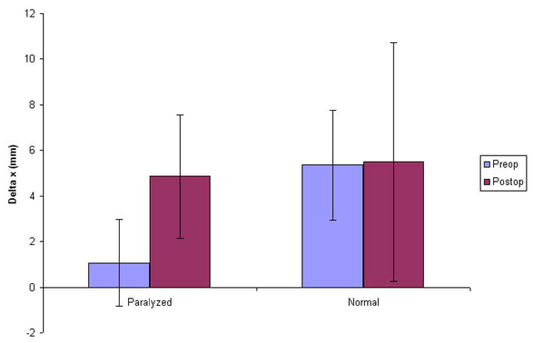

Figure 2. Preoperative and postoperative mean change in smiling parameter x from rest to smile on the paralyzed and normal side of the face.

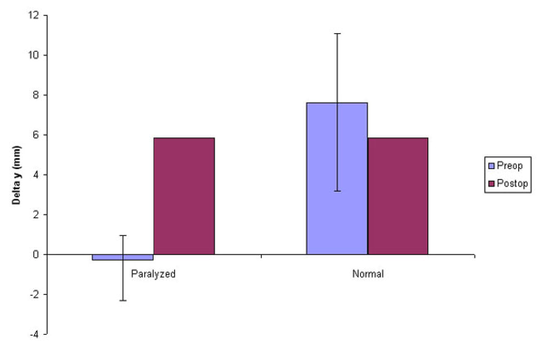

Figure 3. Preoperative and postoperative mean change in smiling parameter y from rest to smile on the paralyzed and normal side of the face.

Results (cont.)

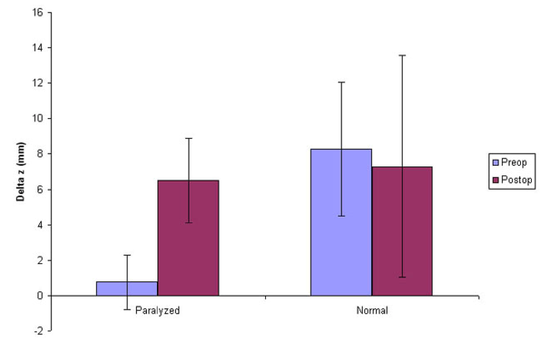

Figure 4. Preoperative and postoperative mean change in smiling parameter z from rest to smile on the paralyzed and normal side of the face.

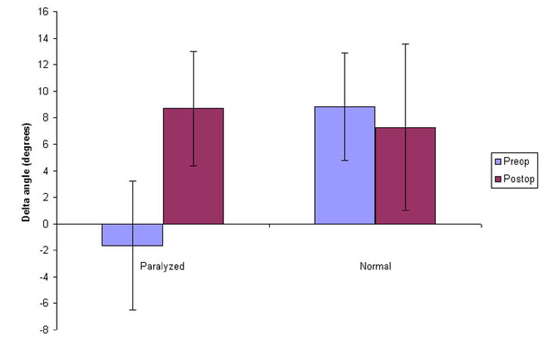

Figure 5. Preoperative and postoperative mean change in smiling parameter angle from rest to smile on the paralyzed and normal side of the face.

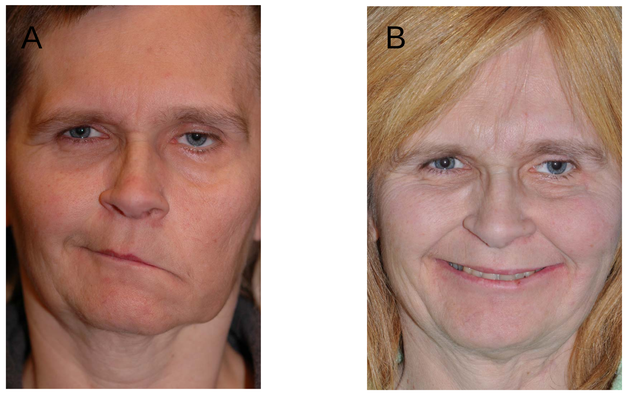

Figure 6. A. Preoperative smile. Notice the left facial paralysis. B. Postoperative smile. Notice the facial symmetry and near normal oral commissure excursion and nasolabial fold of the left, rehabilitated side.

Conclusions

•A single staged free gracilis muscle transfer and re-innervation by the masseteric branch of the trigeminal nerve can be successfully used to treat, motivated NF2 patients with facial paralysis.
•In this small series, NF2 patients and their providers were pleased with the cosmetic and functional outcome.
•Facial rehabilitation after facial paralysis should be part of the complex treatment algorithms that are devised by providers taking care of patients with Neurofibromatosis type 2.

References

1.Evans DG, Huson SM, Donnai D, et al. A genetic study of type 2 neurofibromatosis in the United Kingdom. I. Prevalence, mutation rate, fitness, and confirmation of maternal transmission effect on severity. J Med Genet 1992;29:841-6.
2.Evans DG, Moran A, King A, Saeed S, Gurusinghe N, Ramsden R. Incidence of vestibular schwannoma and neurofibromatosis 2 in the North West of England over a 10-year period: higher incidence than previously thought. Otol Neurotol 2005;26:93-7.
3.Evans DG. Neurofibromatosis type 2 (NF2): a clinical and molecular review. Orphanet J Rare Dis 2009;4:16.