

Hemi-forehead lifting for upper eyelid and brow ptosis in neurofibromatosis type 1

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Abstract

A 63-year-old female patient presented with ptosis in the right eyelid, wrinkles in the right half of the forehead, and narrowing of visual field. She had been diagnosed with giant plexiform neurofibroma. In addition, she had painless, non-tender soft masses on her left shoulder and neck. The patient's ptosis and forehead wrinkles improved in the supine position but were more prevalent in the vertical position. She had undergone blepharoplasty in the previous year. She did not have any disease history other than mild ptosis in the right and a mass in the neck that was present since puberty but had grown within the last decade. Pretrichial hemi-forehead and brow lift for eyebrow ptosis and forehead wrinkles was planned and performed without any complications in the early postoperative period and no recurrence in the 18month follow-up period.

Introduction

Neurofibromatosis type 1 (NF-1), also known as von Recklinghausen disease, is one of the most common phacomatoses and has a worldwide prevalence of approximately 1/3500. Approximately 30-50% of NF-1 cases present with spontaneous mutations. Neurofibromas are present in a wide variety of tissues and organs in the NF-1 patients. There have been cases reported of ptosis due to neurofibroma in the eyelid. In these cases, a marked

downward orientation in the eyelid position is seen with aging. There is a negative correlation between ptosis in the eyelid and eyebrow elevation with aging. Transient wrinkles on the forehead occur due to an increase in frontal muscle activity in individuals with age-related upper eyelid ptosis.⁵ Following blepharoplasty, eyelid ptosis may still be visible.⁶ Limited forehead lifting is recommended to increase the effect of upper blepharoplasty and to increase eyebrow elevation.⁷

Methods

This study is a single patient case report which adhered to the principles of the Declaration of Helsinki. Informed consent was obtained for publication of the study and the photographs.

Case Report

A 63-year-old woman presented with complaints of ptosis in the right eyelid, wrinkles in the right side of her forehead, and constriction of her visual field over several years. The patient had undergone surgery 20 years ago in the left side; however, there was no medical record of the surgery. The patient had undergone blepharoplasty one year before coming to our center. The patient's ptosis was evaluated in the upright position of her head with her eyes closed and in the supine position with complete relaxation of her eyebrows and her forehead while her eyes were closed.

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Clinical examination demonstrated that on the left half of her forehead, her skin was tight and stretched and did not move very easily. When the forehead wrinkles were stretched upward, the levator muscle functioned normally. Frown lines, forehead lines, and eyelid ptosis were prominent in the upright position of her head (Figure 1A). Eyelid and lateral eyebrow ptosis were found to improve in the supine position (Figure 1B).





Figure 1
Clinical examination demonstrated frown lines, forehead lines, eyelid ptosis in the upright position of her head (A). Eyelid and lateral eyebrow ptosis improved in supine position (B).

She had been diagnosed with NF-1 with giant plexiform neurofibromas despite no family history of plexiform neurofibromas. A 38 cm by 21 cm mass with inflamed ulcers in the middle on her left shoulder and a 12 cm by 9 cm painless, non-tender soft mass on her neck were identified (Figure 2A). There were multiple café-au-lait spots on her extremities and back. There was no concomitant systemic disease except neurofibromas and café-au-lait spots.

Ophthalmic examination revealed 20/20 visual acuity in each eye. Anterior segment and fundus examination revealed no pathological findings such as Lisch nodules or optic glioma.

She was scheduled for pretrichial hemi-forehead lifting for eyebrow ptosis under general anesthesia. Surgery was performed using the technique reported by Connell, *et al.*⁵ First, the supraorbital rim was marked medial, middle, and lateral while patient was





Figure 2
Surgical images upon conclusion of the surgery demonstrate the soft masses on the patient's left shoulder and neck (A) and multiple zig-zag incisions in front of the forehead hairline (B). Marked improvement is observed 7 days post-operatively (C).

in the supine position. Then the forehead skin was pulled up until the eyebrow was brought to the desired position. In this new position, the supraorbital rim was re-marked at three points. Skin excision to the vertical distance between the previous and subsequent lines was planned on the forehead. Multiple zig-zag incisions were made 1 mm in front of the forehead hair line to reduce scar formation (Figure 2B). The skin incision was not performed vertically but 30° to the plane of the surface. The incision was extended medially to the mid-forehead and laterally to the preauricular area. Subcutaneous dissection reached the superior orbital rim without damaging the deep branches of the supraorbital nerve. Excess skin was marked by stretching, excised and skin was sutured with metal punch and 4-0 Prolene suture. The patient's sutures were removed on the seventh postoperative day (Figure 2C). The surgical outcome was aesthetically and functionally successful and did not require revision. There were no complications in the early postoperative period and no recurrence was found at 18 months of follow-up.

Discussion

Ptosis usually occurs due to the mechanical effects of neurofibroma in the eyelid in patient with NF-1.^{3,4} This case differs from other giant plexiform neurofibroma cases as it is unilateral and independent from neurofibromas. The wrinkles of the patient were identified as dynamic wrinkles since the forehead



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wrinkles and ptosis were undetectable when the mimic muscles were at rest while they appeared in the vertical position. Aging and exposure to ultraviolet-B rays increases fibroblast-induced elastase expression, leading to a reduction in skin elasticity and wrinkle formation.⁸ In individuals with aging-related eyebrow ptosis, excessive accumulation of skin at the upper orbital edge leads to frontalis muscle hypertonicity, resulting in transient wrinkles on the forehead.^{5, 9, 10} The lateral part of the eyebrow was more ptotic in this case. This may be explained by lesser attachment of the frontal muscle to the lateral parts of the eyebrow and gravitational effects on the lateral superficial temporal fascia.^{10, 11}

Removing excess skin with blepharoplasty removes the tonic reflex stimulus that the frontal muscle creates to lift the ptotic eyebrow; however, blepharoplasty alone may not be enough to achieve the desired cosmetic and functional result. Although blepharoplasty was performed in this case, ptosis in the eyebrow and eyelid could not be sufficiently corrected. Combining blepharoplasty with foreheadplasty has been reported to provide better aesthetic results.^{6,10}

The most effective method to pull the tissues upwards is the facelift to counteract the downward sagging due to gravity. Foreheadplasty, sometimes referred to as forehead lifting, aims to straighten the transverse lines on the forehead, correct ptosis, and resuspend the pseudo-excess skin sagging from the superior orbital area. This approach was critical for the current patient, as she had undergone previous surgeries that were not satisfactory. This also demonstrates that planning before surgery is critical. For instance, lifting the whole eyebrow without proper planning can cause a permanent "surprised" appearance, while excessively lifting the medial part of the eyebrow will cause a sad appearance.

Intervention to the muscles of the brow is more important than the incision site or incision plan in eyebrow lifting surgery. A better and longer-lasting effect can be obtained by increasing the strength of the muscles that lift the eyebrows and decreasing the strength of the muscles that pull the eyebrows

downward.¹² Good surgical planning and selection of the proper surgical method for position-associated unilateral eyebrow and eyelid ptosis is critical to be able to perform satisfactory surgeries on a patient-bypatient basis.

Conclusion

Ptosis due to neurofibroma requires specific intervention. It is generally difficult to identify due to its distinguishability with dynamic wrinkles in the vertical position. It is important to examine patients with NF-1 and forehead wrinkles in the supine position to determine whether the patient has ptosis. Foreheadplasty is a surgery that can be used to correct ptosis in patients.

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