



Blepharoplasty techniques in the management of orbito-temporal neurofibromatosis

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Summary We aimed to present blepharoplasty techniques we used for severe orbito-temporal neurofibromatosis (NF). A retrospective noncomparative single-center case study was undertaken on patients with orbito-temporal NF. Twenty-two patients with orbito-temporal NF treated at the Department of Ophthalmology of Shanghai Ninth People's Hospital between 2007 and 2011 participated in the study. They underwent a standard ophthalmologic assessment for orbito-temporal NF involving both the orbito-temporal soft tissue and bony orbits. The orbits were examined with three-dimensional computed tomography (CT) and all 22 patients underwent tumor debulking, blepharoplasty, and orbital reconstruction. We modified the conventional procedures. Our reconstructive techniques included eyelid reduction; lateral canthal reattachment; for patients with collapse of the lateral orbital margin, reconstruction of the orbital margin to be performed before reattaching the lateral canthus to the implanted titanium mesh; anterior levator resection; and frontalis suspension according to preoperative levator muscle function. Visual acuity, tumor recurrence, and postoperative palpebral fissure and orbital appearance were evaluated to assess outcomes. Acceptable cosmetic results were obtained in 22 patients after debulking of the orbito-temporal NF and surgical reconstruction. There was no loss of vision or visual impairment postoperatively. All patients did not display recrudescence after a follow-up period of >1 year. Three patients with residual ptosis were successfully treated with a second ptosis repair. We believe that the blepharoplasty techniques described in the treatment of orbito-palpebral NF may provide both functional and esthetic benefits.

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Introduction

Neurofibromatosis (NF) type 1 is an autosomal-dominant inherited disorder with an incidence of approximately one in 3000 live births.¹ Orbito-temporal deformities, which develop in utero, can cause severe disfigurement and affect visual development.^{2,3} The typical clinical findings of orbito-temporal NF include neoplasms involving the orbital,⁴ eyelid, and temporal regions with ptosis. In addition, bony malformations involving the orbit and sphenoid dysplasia are also observed. NF is not sensitive to radiotherapy and chemotherapy; thus, surgical excision is the first line of treatment. Because NF-infiltrated tissues are diffusely infiltrated and rich in blood vessels, treatment is frustrating to both patients and surgeons.^{5–7}

The eyelid deformity of the orbito-temporal NF primarily demonstrates eyelid hypertrophy due to tumor invasion; the hypertrophic eyelid can induce lateral canthal displacement and palpebral fissure extension.⁸ At the same time, blepharoptosis occurs because of the gravity of the tumor itself as well as the implication of the levator muscle. The involved eye appears as an “S” shape, and the soft tissue can even swell and prolapse at the orbital region of the affected side. The transverse oblique approach was formerly used for upper eyelid reduction and levator resection was often adopted as the corrective method, which, however, improved this approach.⁹ Horizontal resection of the skin orbicular muscle and vertical resection of the full-thickness eyelid were performed on the upper eyelid; additionally, vertical resection of the lateral full-thickness eyelid on the lower eyelid was also performed, as it has the same redundancy as the upper eyelid. The cut ends of the upper and lower eyelids were reshaped and fixed at the lateral canthus angle. Blepharoptosis correction was chosen on the basis of preoperative eyelid muscle strength. In this article, we report our experience on orbital–temporal NF and evaluate the efficacy of improved blepharoplasty techniques used for correction.

Patients and methods

A retrospective interventional case series review was performed of patients with orbito-temporal NF treated by one of the authors (XF). All consecutive patients with both orbito-temporal soft tissue and/or bony involvement who underwent tumor debulking and the aforementioned blepharoplasty techniques between 2007 and 2011 were included in this study. The following data were extracted from patient records: age and gender, previous lid and/or orbital surgery, dates of the initial examination, best corrected visual acuity, type of upper/lower lid infiltration, and surgical procedures (Table 1).

Every patient underwent surgery during a stationary phase of the disease, in which disease progression had slowed for >1 year. The surgical procedures included in this study were as follows: debulking of the eyelid and temporal neurofibroma, eyelid reduction, lateral canthal reattachment, and ptosis repair. For patients with severe bony orbit deformity with dysplasia of the zygomatic arch and inferior orbital margin, tension-free sutures could not reattach to the lateral tendon. Because no obvious pulsating proptosis

appeared, we did not repair the sphenoid wing. We first implanted a titanium mesh to remodel the lateral and inferior orbital margin. The local tension of the surrounding soft tissue significantly dropped; then, the lateral canthal tendon was reattached to the periosteum with sutures at a relatively anatomical level. Six patients underwent titanium mesh placement and all had satisfactory correction of the detached lateral canthus. To our knowledge, surgical intervention of the detached lateral canthus with titanium mesh has not been reported.

We first carefully dissected the infiltrative tumor, which can adhere to the soft tissue in the eyelid and temporalis, and then removed it partially with impunity. Orbital neurofibroma debulking was performed via an anterior, lateral, or anterolateral orbitotomy.

To guide the complex reconstruction, the patient's defect was modeled on the basis of computed tomography (CT) data, previously collected (Figure 1A). The implant that matched the defect precisely was created with a three-dimensional plastic model, with a material similar to the titanium mesh (Figure 1B). Utilizing the orbital implant, the orbital floor was elevated and the inferolateral orbital rim was repositioned, both of which reduced the enlarged orbit in size and volume and counteracted the downward globe displacement.¹⁰ The patient with a blind eye underwent enucleation, and an orbital prosthesis was fitted in the course of his surgical management.

To correct upper eyelid laxity, we resected the excess skin and orbicularis muscle in a fusiform shape and performed a wedge resection of the lateral one third of the upper lid. This resection shortened the horizontal width and also improved the unpleasant “S” shape with a natural curvature of the lid. A lower eyelid-shortening procedure was performed as necessary to match the upper eyelid length in the reconstructed palpebral aperture. The lateral canthal angle was reformed by carefully bringing the upper and lower eyelid margins together to approximate the grey line and edge of the eyelid margin and was then sutured with 7/0 polyglactin. The lateral canthal tendon was reattached to the periosteum of the lateral orbital wall with a nonabsorbable tension-free suture. For patients with severe bony orbit enlargement, zygomatic arch dysplasia, and inferior orbital margin, the individually designed titanium mesh (extended orbital rim implant) was implanted under the periosteum and the orbital aperture was remodeled. Then, the free end of the lateral canthal tendon was reattached to the periosteum with sutures at a relatively normal level (Figure 1C). Anterior levator resection was undertaken in patients with upper eyelid mechanical ptosis accompanied by > 1 mm levator function. A frontalis suspension procedure with autogenous fascia lata was undertaken in patients whose preoperative evaluation of levator function was 0 mm.

All patients were followed up for a minimum of 1 year after surgery. Postoperative visual acuity, neurofibroma recurrence, and palpebral fissure and orbital appearance were assessed in all patients.

Results

Twenty-two patients (14 males and eight females) with ages ranging from 16 to 43 years were included in the study.

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