OPEN

PRS GLOBAL OPEN

CASE REPORT
Reconstructive

Frontalis Suspension in Muscular Dystrophy: 16-years Follow-up

Badr M. I. Abdulrauf, MD, FRCSC

Summary: Acquired eyelid ptosis in adults, with complete loss of levator palpebrae superiosis function, can be a challenging problem to diagnose and treat. A 48-year-old woman with chronic bilateral severe blepharoptosis of 10 years duration is presented, whose neurological investigations excluded myasthenia gravis. The patient was preliminarily diagnosed with chronic progressive external ophthalmoplegia. The levator excursion was negligible, and a frontalis suspension procedure was considered using a conventional autogenous fascia lata graft. An optimal outcome was achieved with over 16-years follow-up. Although the patient was healthy otherwise upon first presentation, 10 years later, she developed other neurologic manifestations, including dysphagia and oral dryness. The fact that blepharoptosis did not recur over the years in this case differentiates an oculopharyngeal type of muscular dystrophy in this patient from other types and from the more frequent condition of chronic progressive external ophthalmoplegia.
(Plast Reconstr Surg Glob Open 2022;10:e4225; doi: 10.1097/GOX.0000000000004225; Published online 22 March 2022.)

Apart from congenital ptosis where the muscle would not develop, few acquired myogenic pathologies have been recognized in adults. Senile ptosis does have a myogenic component; however, it usually responds well to a Levator shortening procedure. Myasthenia gravis is a disease in which the neuromuscular transmission is interrupted with typical worsening of ptosis diurnally.

Clinical testing of levator excursion is the single most important method of differentiating whether the problem exists in the aponeurotic component, a much more common factor, or otherwise in the muscle. In patients with neurogenic cause such as third nerve palsy or in a primarily myogenic disease, they would demonstrate minimal or no levator excursion while the frontalis is passively resisted. In such a situation, or with poor levator excursion (less than 4 mm), there would be a need for a donor motor.

CASE PRESENTATION

Here, we present a 48-year-old female patient referred by a neurologist. She reported a history of gradually worsening eyelid ptosis of 10 years duration. There was no diurnal variation and the patient was healthy otherwise. The blepharoptosis was bilateral, symmetric, and severe (Fig. 1). The orbicularis oculi muscles (OOM) were working fine and Bell's phenomenon was intact. The patient was thoroughly investigated by neurologists for myasthenia gravis and was excluded. After an ophthalmology consultation, the patient was primarily diagnosed with chronic progressive external ophthalmoplegia (CPEO), and was suggested for an oculoplastic intervention. (**See Video 1 [online],** which shows patient's initial presentation with muscular dystrophy and complete loss of levator function.)

A frontalis suspension procedure was considered, under local anesthetic and IV sedation. Autogenous fascia lata was used as a sling; two vertical strips were used per side. With a supra-tarsal incision, a tunnel was created superficial to the orbital septum. A counterincision made within the upper limit of eyebrow and the frontalis fibers were identified. The fascial strips were initially secured at tarsal plate (Fig. 2). With some cooperation from the patient's side, the amount of tension to be considered was judged. Cephalic ends of the fascial slings were then retrieved and tied to the frontalis fibers. Initially and after resolution of edema, partial and relative improvement in the ability of spontaneous upper eyelid opening was noted, but was thought to be suboptimal. Three weeks later, we readjusted the tension on the upper end. The patient did

From the Section of Plastic Surgery, King Faisal Specialist Hospital and Research Center (General Organization), Jeddah, Saudi Arabia.

Received for publication January 1, 2022; accepted February 7, 2022.

DOI: 10.1097/GOX.0000000000004225

Disclosure: *The author has no financial interest to declare in relation to the content of this article.*

Related Digital Media are available in the full-text version of the article on www.PRSGlobalOpen.com.

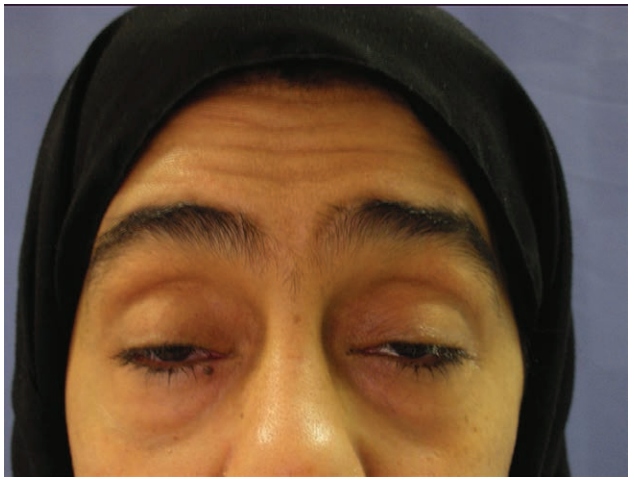

Fig. 1. A 48-year-old woman with a 10-year history of progressively worsening eyelid ptosis (without diurnal variation). The patient is attempting to maximally open her eyes.

very well and was eventually able to fully open her eyes without lagophthalmos, but used lubricants on the long term (Fig. 3). Ten years later, the patient began complaining of dysphagia and increasing oral dryness, for which she has been treated and followed by a gastroenterologist. Regarding the eyelids, however, it has been over 16 years since the procedure and she has been doing extremely well without the need for any further intervention or revision (Fig. 4). (**See Video 2 [online],** which shows the patient 16 years post-frontalis suspension.)

DISCUSSION

The three better known and specific myogenic entities associated with acquired eyelid ptosis include:

1. CPEO, a relatively more frequent entity among others, caused by genome mutations in the mitochondria and nuclei, making it transmissible maternally as well as autosomal dominant and recessive. Patients would have significant limitation in their extraocular muscle function as well for the OOM. Cardiac conduction defects and cerebellar ataxia might coexist. Muscle biopsy, if considered, would reveal mitochondrial abnormality. As the name implies, this condition is progressive, and blepharoptosis correction would need to be revised within a few years of the initial reconstruction.[1–3]
2. Myotonic dystrophy is an autosomal dominant disease. The extraocular muscle dysfunction is not as severe, but the OOM is more significantly affected.
3. Oculo-pharyngeal muscular dystrophy (OPMD) is known to affect certain geographical clusters in Quebec (French descent), Northern New Mexico (Hispanic descent), and Israel/ Palestine (Bukhara Jewish descent). Patients with OPMD would have intact OOM strength and Bell's phenomenon. Dysphagia is commonly associated and may precede the ocular manifestation. Dysphonia is also a common manifestation. Proximal muscle weakness might follow.[4–7]

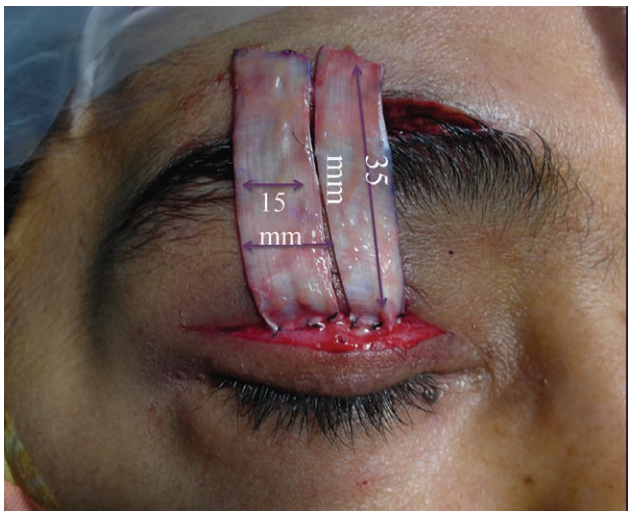

Fig. 2. Intraoperatively, fascia lata graft, two curtain shaped strips are secured side by side to the upper margin of the tarsal plate (the patient's right side is shown), and would be tunneled under the eyelid flap to be attached to frontalis fibers. We believe this pattern and vector of the sling best simulates the levator.

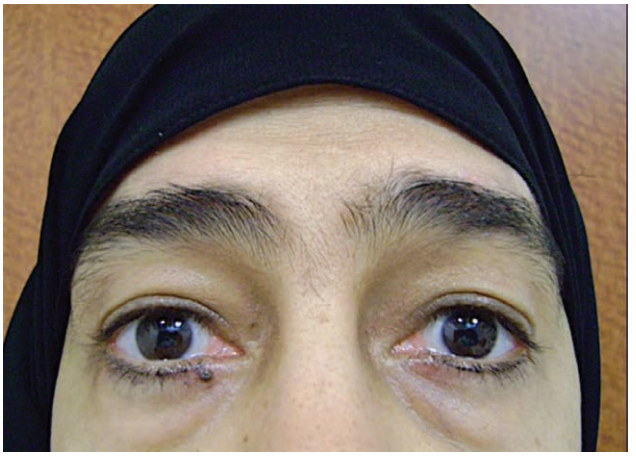

Fig. 3. Five months postoperative, the patient could actively and satisfactorily open her eyes.

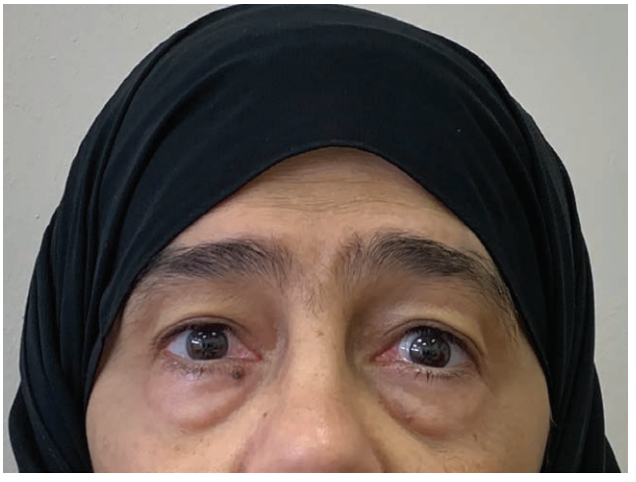

Fig. 4. Sixteen-year follow-up, without any further intervention.

Our patient, who first presented with almost complete obliteration of visual field, was initially inadequately co-diagnosed with CPEO, simply due to the absence of other neurologic or systemic manifestations, as well as on the basis of "common things are common." Myasthenia gravis was further excluded by the appropriate neurologic tests. A muscle biopsy was not considered; however, retrospectively speaking, it would have been mostly of academic significance but not as much in terms of technical treatment plan.

In order to transfer the force of a functioning frontalis to the tarsus, various materials could be used. Monofilament nylon, poly-filament sutures, silicone rods, allogenic and autogenous fascia lata, temporal fascia and palmaris longus tendons have been used.[8–11] It is our belief, from our experience, that autogenous materials are far more reliable due to various reasons. In fact, the recurrence rate and complication of infections and granulomas were found to be quite high in pediatric blepharotosis correction, due to the fact many are being done using nonautogenous materials.[12,13] Various configurations of fascial strips such as a straight fan shape, triangular, pentagon, and W or inverted Y have been described.[14,15]

However, in our practice, we find it ideal to use "curtain" shaped strips. It must be realized that a normal levator muscle truly works as a curtain with its vector of pull, cranially and directly. Furthermore, this simple uninterrupted sling reduces operative time by avoiding sewing various strips together.

The idea of using an OOM–frontalis muscle local flap in treating blepharoptosis was first introduced in the period of 2005–2009. The concept is based on the presence of interdigitations between the frontalis and orbital portion of the OOM; the cross linkage between vertical fibers of the former and horizontal ones of the latter would enable dynamically substituting the levator function. Such a method would avoid the need for a grafting material and a distant donor site issue.[16] Earlier experiences and reports were associated with subsequent imbalance due to the antagonistic effects upon eyelid closure.

In 2021 Huang et al[17] described the function preserving frontalis orbicularis oculi muscle flap for treatment of severe blepharoptosis with poor levator function. This refinement stresses on sub-muscular dissection from the lower portion of the OOM, thereby leaving most of it in situ without disturbing its integrity and function.[17]

The three most common sequelae that must be discussed preoperatively include under-correction, over-correction, and asymmetry. Apart from routine minor ptosis correction during esthetic blepharoplasty, it is always advisable to educate the patient about such issues and the possibility of requiring a small revisionary surgical step.

An important piece of advice to be considered for all patients with blepharotosis, whether untreated or those who have undergone a levator sling or other surgical correction, is to abstain from Botox or other injectable toxins to the forehead.

Badr M. I. Abdulrauf, MD, FRCSC
Section of Plastic Surgery, Department of Surgery
King Faisal Specialist Hospital and Research Center
PO Box 40047, Jeddah 21499
Saudi Arabia
E-mail: babdulrauf@kfshrc.edu.sa

PATIENT CONSENT

The patient provided written consent for the use of her image.

REFERENCES

1. Piccolo G, Cosi V, Poloni M, et al. Chronic progressive external ophthalmoplegia. Clinical, electrophysiological, histochemical and ultrastructural studies of 14 cases. *Schweiz Arch Neurol Neurochir Psychiatr.* 1982;131:161–174.
2. Jiménez-Caballero PE, Serviá M, Cabeza CI, et al. Oftalmoplejía externa progresiva crónica: manifestaciones clínicas y electromiográficas en una serie de casos [Chronic progressive external ophthalmoplegia: clinical and electromyographic manifestations in a series of cases]. *Rev Neurol.* 2006;43:724–728.
3. McClelland C, Manousakis G, Lee MS. Progressive external ophthalmoplegia. *Curr Neurol Neurosci Rep.* 2016;16:53.
4. Allen RC. Genetic diseases affecting the eyelids: what should a clinician know? *Curr Opin Ophthalmol.* 2013;24:463–477.
5. Allen RC, Jaramillo J, Black R, et al. Clinical characterization and blepharoptosis surgery outcomes in Hispanic New Mexicans with oculopharyngeal muscular dystrophy. *Ophthalmic Plast Reconstr Surg.* 2009;25:103–108.
6. Yotam R, Vered R. Oculopharyngeal muscular dystrophy as a paradigm for muscle aging. *Front Aging Neurosci* 2014; 6:317.
7. Trollet C, Boulinguiez A, Roth F, Stojkovic T, et al. Oculopharyngeal muscular dystrophy. In: Adam MP, Ardinger HH, Pagon RA, et al, eds. *GeneReviews.* Seattle, Wa.: University of Washington; 2001:1993–2021.
8. Elsamkary MA, Roshdy MM. Clinical trial comparing autogenous fascia lata sling and Gore-Tex suspension in bilateral congenital ptosis. *Clin Ophthalmol.* 2016;10:405–409.
9. Katowitz JA. Frontalis suspension in congenital ptosis using a polyfilament, cable-type suture. *Arch Ophthalmol.* 1979;97:1659–1663.
10. Ahn J, Kim NJ, Choung HK, et al. Frontalis sling operation using silicone rod for the correction of ptosis in chronic progressive external ophthalmoplegia. *Br J Ophthalmol.* 2008;92:1685–1688.
11. Wagner RS, Mauriello JA Jr, Nelson LB, et al. Treatment of congenital ptosis with frontalis suspension: a comparison of suspensory materials. *Ophthalmology.* 1984;91:245–248.
12. Philandrianos C, Galinier P, Salazard B, et al. Congenital ptosis: long-term outcome of frontalis suspension using autogenous temporal fascia or fascia lata in children. *J Plast Reconstr Aesthet Surg.* 2010;63:782–786.
13. Wasserman BN, Sprunger DT, Helveston EM. Comparison of materials used in frontalis suspension. *Arch Ophthalmol.* 2001;119:687–691.
14. Shimizu Y, Nagasao T, Shido H, et al. Intra-eyebrow frontalis suspension using inverted Y-shaped short autogenous fascia lata for blepharoptosis with poor levator function. *J Plast Reconstr Aesthet Surg.* 2015;68:49–55.
15. Helveston EM, Ellis FD. *Pediatric Ophthalmology Practice.* 2nd ed. St Louis, Mo.: CV Mosby; 1984:204–205.
16. Lai CS, Chang KP, Lai CH, et al. A dynamic technique for the treatment of severe or recurrent blepharoptosis: frontalis-orbicularis oculi muscle flap shortening. *Ophthalmologica.* 2009;223:376–382.
17. Huang SH, Lee CC, Lai HT, et al. The function-preserving frontalis orbicularis oculi muscle flap for the correction of severe blepharoptosis with poor levator function. *Aesthet Surg J.* 2021;41:NP260–NP266.