

Case Report

Surgical Treatment of Ptosis in Chronic Progressive External Ophthalmoplegia Patient

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Abstract

Chronic progressive external ophthalmoplegia (CPEO) is a rare mitochondrial disease which affects the ocular muscles and levator muscles, and subsequently result in ophthalmoplegia and ptosis. However, surprisingly, there are few papers about CPEO and its ptosis surgery in plastic surgery fields. In various aspects, ptosis correction in CPEO patients needs special cautions. As CPEO is usually associated with systemic symptoms such as cardiac arrhythmias or retinopathy, thorough preoperative examination is important. Besides, due to weak Bell's phenomenon and lagophthalmos, CPEO patients are in high risk of postoperative exposure keratopathy, so, for these reasons, overcorrection must be avoided. In here, a case of 76 years old female ptosis patients who presented our department with visually severely obstructed condition is presented. We believe this paper help plastic surgeon to understand clinical spectrum of CPEO ptosis and broaden his/her understanding of operative procedures in CPEO patients.

Keywords: Chronic progressive external ophthalmoplegia (CPEO); Ptosis; Frontalis sling; Autologous fascia lata

Introduction

During ptosis surgery, it is easy to surgeons to focus on raising the droopy eyelids and making bigger looking eyes. However, there are some conditions require special considerations in operation. Chronic Progressive External Ophthalmoplegia (CPEO) is a rare mitochondrial myopathy most often involving the levator palpebrae superioris muscle, orbicularis oculi muscle and extra ocular muscle [1]. CPEO is clinically characterized by slowly progressing bilateral ptosis, extraocular movement impairment, poor Bell's phenomenon and poor lid closure. Due to weak Bell's phenomenon and lagophthalmos, CPEO patients are in high risk of exposure keratopathy, and subsequently, ptosis surgery in CPEO patients need specialized cautions [2,3]. However, surprisingly, still, there are few papers about its surgical treatment in plastic surgery fields. Nearly all papers are written in ophthalmologic point of view and the mainstay of treatment is frontalis suspension using silicone rod. In here, we describe our experience of a ptosis patient with CPEO and some surgical tips for plastic surgeons.

Case Presentation

A 76 years old woman presented our department with bilateral ptosis developing at age 30 years. Due to severe vision impairing ptosis, she even could not walk without support and she always adopted a compensatory backward head tilt and chin up posture. In the physical examination, palpebral fissure was just 1.5 mm with maximal frontalis

contraction and levator function was almost zero (Figure 1). Her eye movements were restricted in all direction. There was no sign of diplopia despite the ocular immobility. In her family history, her little sister showed the same symptoms which raising the suspicion of a genetically inherited disease. And in orbital MRI, atrophy of the extraocular muscles was detected (Figure 2). Suspect CPEO ptosis, we consulted ophthalmologist and internist and evaluated systemic involvements, such as pigmentary retinopathy, optic atrophy, cardiac arrhythmia, skeletal muscle weakness, ataxia, dementia, dysphagia or hearing loss. Luckily, there were no systemic symptoms other than oculomotor immobility. Serum lactate and pyruvate level were normal range, too. Considering these results, suspicious of ptosis in CPEO isolate type, we explained the patient and her family member about the CPEO as a mitochondrial myopathy, progressive nature of ptosis, possibility of systemic involvements, as well as frontalis sling surgery for ptosis correction. Few days later, frontalis suspension surgery using autogenous fascia lata was performed under local anesthesia. At first, a 4-cm-long and 3-cm-wide piece of the fascia lata from the right thigh was harvested as an autologous sling material and separated into four 4-cm-long and 0.75-cm-wide strips. After fascia lata harvesting, eyelid skin incision line was designed (Figure 3). Excision line for excess upper eyelid skin, 2-mm-width, was designed in eyelid crease and removed. Two 8-mm-long incision lines were made symmetrically at superior border of eye brows. From tarsal plate to eyebrow incision line, beneath orbicularis muscle, the tunnels for fascia strips were made. Guiding the fascia strips through the tunnel, the lower part of the strips was anchored at the tarsus with 6-0 monofilament polypropylene suture. The eyelid height was meticulously adjusted as eyelid margin was 2 mm above the pupil in frontalis muscle contraction and as about 1 mm above the lower eyelid margin in frontalis muscle relaxed condition. After adjustment, the other two ends of fascia strips were sutured to frontalis muscle and subcutaneous fibroadipose tissue under the eyebrow, with 6-0 monofilament polypropylene suture, with a manner of V. Finally the eye lid skin was closed with continuous 6-0 nylon sutures. For diagnosis of CPEO, muscle biopsy sample was obtained from vastus lateralis muscle of thigh and orbicularis oculi muscle during operation, and

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subsequently it was consistent with mitochondrial myopathy, CPEO (Figure 4). Eight months post-operative follow up showed excellent results with eyelid excursion of about 10 mm on both eye without keratopathy nor lagophthalmos. No revisional surgery was required and cosmetically and functionally, satisfactory results were acquired (Figure 5).

Discussion

Ptosis is a very common symptom for plastic surgeon, but, still yet, with unknown etiology, there are some cases that we plastic surgeon need to special cautions in treatment. Chronic Progressive External Ophthalmoplegia (CPEO) is a very rare mitochondrial myopathy disorder characterized by slowly progressive paralysis of



Figure 1: Preoperative appearance of the patient. Resting state (a) and with maximal frontalis contraction state (b). Palpebral fissure was just 1.5 mm with maximal frontalis contraction and levator function was almost zero.

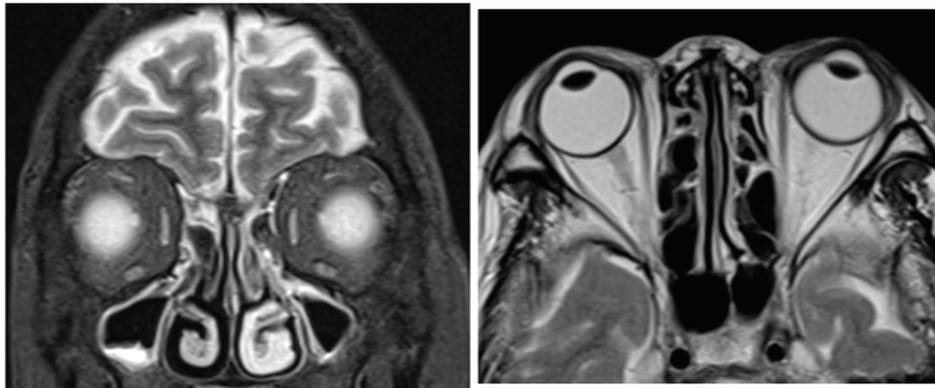


Figure 2: Preoperative MRI examination showed diffuse extra ocular muscle atrophy on coronal view (a) and axial view (b).



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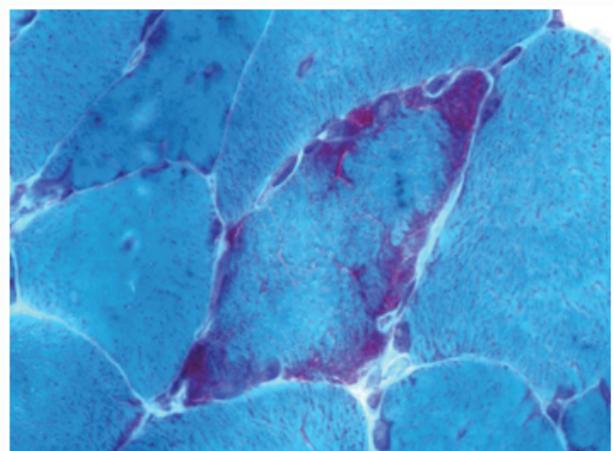


Figure 4: Biopsy of the extraocular muscle showed ragged red fiber in Gomori-trichrome stain.



Figure 5: Postoperative 8 months after surgery. Resting state (a) and with maximal frontalis contraction state (b).

the extraocular muscle and levator muscle [1,4]. It was described in 1868 by Von Grafe and as the name implies, CPEO takes a very slowly progressive course, over a course of years. Clinically, CPEO patients, manifests bilateral ptosis, ophthalmoplegia. As a mitochondrial cytopathy, ophthalmoplegia symptom can occur in isolation, often referred to as "isolated CPEO", or often co-occurs with other symptoms of mitochondrial dysfunction ("CPEO plus syndrome") [1]. Among these, CPEO plus syndrome includes such as Kearns-Sayre syndrome, Stephens syndrome, oculopharyngeal muscular dystrophy or myotonic dystrophy. And Kearns-Sayre syndrome, the most severe form, is a multisystem disorder characterized by an early onset (< 20 years old) of external ophthalmoplegia, pigmentary retinopathy, ataxia and cardiac conduction [5,6]. Due to the possibility of systemic involvement, whenever suspected CPEO, meticulous and accurate evaluation is required. Moreover, in surgical treatment of ptosis in CPEO patients, there is a significant risk of postoperative corneal exposure and exposure keratitis due to lagophthalmos and a poor Bell's phenomenon, therefore, surgical correction should be conservative. And surgical treatment for ptosis is only performed when the visual axis is extremely obscured or when the ptosis is cosmetically unacceptable [7,8]. And in surgical repair, as frontalis muscle is relatively spared, levator function usually become a decisive factor. Johnson and Kuwabara mentioned a surgical guideline that if levator function was less than 8mm, frontalis suspension will be the surgery of choice [9]. Whereas, if the levator function is 8mm or greater, than levator resection is indicated. Lane and Collins [8] proposed a treatment of protocol based on a threshold of 4mm as opposed to 8 mm. Additionally, intraoperative adjustment of the frontalis sling also plays a pivotal role in ptosis correction. Shorr et al. [10] explained that the frontalis sling has to be adjusted to provide a firm linkage between the eyebrow and eyelid, but it is loose enough to allow eyelid closure when the frontalis is relaxed. In our case, we also continuously checked the patient that she was able to close the eyes when the frontalis muscle was relaxed, both in supine and sitting position. Even though, recurrence rate using autogenous fascia lata is varied from 0%-22% [11,12] and there is no large population study of recurrence rate of CPEO patients using frontalis sling, this sling can be tightened for recurrent CPEO ptosis. Besides, CPEO ptosis patients frequently have an ectropion or large ophthalmos symptoms, and thereby, upper eyelid surgery in conjunction with lower eyelid elevation with an appropriate spacer may be more effective than upper eyelid surgery alone [3,10] to protect corneal damage. Furthermore, in terms of frontalis sling material, we used autologous fascia lata due to

its low infection (e.g. formation of granuloma, foreign body reaction), low allergic reaction and long-lasting effect, but, other materials such as banked fascia lata, palmaris longus tendon, silicone, polyester fiber (Mersilene Mesh®), polytetrafluoroethylene (Gore-Tex®) also may be used for frontalis suspension [13-15]. As each material has own unique advantage and disadvantage, we believe that judicious use sling material is a safe and efficient way. Finally, perioperative period use of lubricating eye drops or ointment and close clinical follow up are also critical for preventing postoperative keratopathy in CPEO patients.

Conclusion

Here, we described ptosis surgery of 76 years old CPEO patient. Ptosis is a common symptom in the plastic surgery field, but most plastic surgeons are unfamiliar with CPEO. Since CPEO is a mitochondrial myopathy, which is usually associated with systemic features, understanding of its hereditary and progressive nature is imperative. And if patients appear to have ptosis with CPEO, including isolated CPEO, consulting professional internist or neurologist and undergoing a thorough examinations also cannot be neglected. Besides, as a plastic surgeon, surgical considerations not doing ptosis surgery as a usual manner, but striving to prevent postoperative corneal exposure or recurrence of ptosis is required.

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