

Frontalis Suspension Sling Using Silicone Rods in Chronic Progressive External Ophthalmoplegia

Ayşe Dolar Bilge², Dilcan Kotan¹, Cem Mesci³

¹Sakarya Üniversitesi Tıp Fakültesi, Nöroloji, Sakarya, Türkiye

²Erzincan Üniversitesi Tıp Fakültesi, Göz Hastalıkları, Erzincan, Türkiye

³İstanbul Medeniyet Üniversitesi Tıp Fakültesi, Göz Hastalıkları, İstanbul, Türkiye

ABSTRACT

A 72 year old patient with bilateral ptosis and progressive restricted eye movements was diagnosed as chronic progressive external ophthalmoplegia (CPEO). We performed frontalis suspension surgery with silicone rod as a sling material. The result after twelve month follow up was satisfactory, the visual axis remained open without any complications. Ptosis surgery, is one of the most common operations performed by oculoplastic surgeons. A proper neurologic examination is necessary to diagnose the underlying cause of ptosis. CPEO is a type of mitochondrial myopathy which affects extraocular muscles. Frontalis sling operation is generally performed in patients with severe ptosis and poor levator muscle function. Many autogenous and nonautogenous suspension materials are available with numerous reports of successful correction. Here presented a case of blepharoptosis in CPEO and the surgical result of silicone rod sling of this case.

Key words: ptosis , chronic progressive external ophthalmoplegia, frontalis sling

KRONİK PROGRESİF EKSTERNAL OFTALMOPEJİDE SİLİKON ÇUBUKLARLA FRONTAL ASKILAMA CERRAHİSİNİN KULLANIMI

ÖZET

Bilateral pitozu ve progresif göz hareket kısıtlılığı olan 72 yaşındaki hasta, kronik progresif eksternal oftalmopleji (KPEO) tanısı aldı. Hastaya, askı malzemesi olarak silikon çubuk ile frontal askılama ameliyatı uygulandı. Oniki aylık takip sonrası sonuç tatmin ediciydi, görme aksı herhangi bir komplikasyon olmaksızın açık kalmıştı. Pitoz ameliyatı oküloplastik cerrahlar tarafından sık yapılan operasyonlardan bir tanesidir. Düzgün bir nörolojik muayene, pitozun altında yatan nedenini teşhis etmek için gereklidir. KPEO, ekstraoküler kasların etkilendiği bir mitokondriyal miyopati tipidir. Frontal askılama operasyonu genellikle ağır pitozu ve zayıf levator kas fonksiyonu olan hastalarda uygulanmaktadır. Otojen ve otojen olmayan materyaller ve bunların başarılı sonuçlarını bildiren pek çok yayın mevcuttur. Burada, blefaropitozisi olan KPEO tanılı bir olgu ve bu olgunun silikon çubuk askılama cerrahisi sonucu sunulmuştur.

Anahtar sözcükler: pitoz, kronik progresif eksternal oftalmopleji, frontal askılama

Pptosis correction in CPEO problematic because of poor Bell's phenomenon and possible exposure keratitis. Although fascia lata is the gold standard as sling material, silicone rod sling result is as satisfactory as autogenous materials.

Case report

A 72 year old man presented with progressive bilateral ptosis (Figure 1). His visual acuity, fundoscopic and biomicroscopic examinations were normal. He had chin-up posture and restricted eye movements and significant ptosis (Table).

Diagnosis of CPEO was confirmed on muscle biopsy. Operation was performed under local anesthesia. The upper eyelid, brow and lower forehead were injected with %1 lidocaine containing 1: 100,000 epinephrine. Crawford type ¹ bilateral frontalis brow suspension surgery was performed using silicone rods.

Because of poor Bell's phenomenon and possible exposure keratitis, surgical goal was just opening the visual axis. Eyelid height was adjusted as the eyelid margin was above the pupil. Following surgery, palpebral aperture was bilaterally 7 mm. and patient was happy with the result. No corneal exposure was observed after surgery. After 12 months follow up result was satisfactory, eyelid height was symmetrical and eyelid margins were above the pupil (Figure 2).



Figure 1. Preoperative image of patient with chronic progressive external ophthalmoplegia.



Figure 2. Postoperative image of patient which has satisfactory result.

Table 1. Ptosis measurements

Right eye	Measurements	Left eye
4 mm.	Palpebral aperture	4 mm.
0 mm.	Margin reflex distance 1	0 mm.
6 mm.	Margin reflex distance 2	6 mm
absent	Levator function	Absent
absent	Skin crease	Absent
weak	Orbicular function	Weak
nil	Lagophthalmos	Nil
present	Corneal sensation	Present

Discussion

Chronic progressive external ophthalmoplegia is a mitochondrial disease which affects extraocular muscles, first described in 1867 by Albert von Graefe, as a clinical syndrome and disease entity.² CPEO is characterized by extraocular motility impairment, generalized muscle weakness and blepharoptosis. Disease usually causes total ophthalmoplegia and loss of levator function.³ The most common presenting symptom in CPEO is ophthalmoplegia and ptosis which develop by years. The diagnosis of CPEO is based on the results of a muscle biopsy or Southern blot analysis of muscle DNA to test deletions.² The findings on muscle biopsy in CPEO include the typical ragged-red fibers described by Cesen et al (4) in 1972 and a mosaic pattern of cytochrome C-oxidase (COX-negative fibers).

Frontalis sling procedure is the treatment of choice in the patients with CPEO because patients usually have severe ptosis and poor levator function (5). Many materials are available for frontalis sling suspension. Using autogenous fascia lata as a sling material in general ptosis surgery is the gold standard. This technique has lower complication rate but sometimes it can be problematic because of postoperative pain, infection, hematoma and surgical scar. Stored fascia lata on the other hand has a higher rate of recurrence (5). Other choices of sling materials include nonautogenous materials like silicone rods. Elastic nature of silicone rods allows good eyelid approximation, minimizes lagophthalmos and corneal exposure, and it is easy to remove it in case of exposure keratopathy which usually occurs in CPEO patients (6). Palmaris longus tendon can be also used as sling material like other tendons. Salvi et al. has reported successful results of using palmaris longus tendon at CPEO (5). Although autogenous fascia lata is the gold standard sling material and using autogenous palmaris longus has satisfactory results (6), harvesting fascia lata and palmaris longus tendon is not always possible because of technical impossibilities and experienced staff deficiency. Palmaris longus tendon is routinely used for orthopedic, traumatic and plastic surgery, if we had a chance to use synthetic materials, we can keep autogenous materials for more important situations. Frontalis sling operation using silicone rod is safe and effective and easy to remove in case of exposure keratopathy which is not rare in CPEO patients.

References

- Crawford JS. Repair of ptosis using frontalis muscle and fascia lata. *Trans Am Acad Ophthalmol Otolaryngol* 1956;60:672–8.
- Benedikt GHS, Dieter P. Extraocular mitochondrial myopathies and their differential diagnoses. *Strabismus* 2006;14:107–113.
- Daut PM, Steinemann TL, Westfall CT. Chronic exposure keratopathy complicating surgical correction of ptosis in patients with chronic progressive external ophthalmoplegia. *Am J Ophthalmol* 2000;130:519–21.
- Cesen W, Engel WK, Walsh GO, et al. Oculocraneosomatic neuromuscular disease with ragged-red fibers. *Arch Neurol* 1972;26:193–211.
- Salvi SM, Currie ZI. Frontalis suspension sling using palmaris longus tendon in chronic progressive external ophthalmoplegia. *Ophthalm Plast Reconstr Surg*. 2009;25: 140-141.
- Ahn J, Kim NJ, Choung HK, Hwang SW, 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.