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Case Report

Ptosis correction with frontalis sling surgery in a patient with bilateral chronic progressive external ophthalmoplegia: A case report

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Abstract

Chronic progressive external ophthalmoplegia commonly manifests as bilateral ptosis and external ophthalmoplegia which is quite disabling for the patient. The complication with conventional ptosis surgery is exposure keratitis due to induced lagophthalmos. This is a case report of 50 year old male who presented with complain of drooping of both upper lids since 10 years. On examination he had bilateral severe ptosis with absent levator function and poor Bell`s phenomenon. A diagnosis of chronic progressive external ophthalmoplegia was made after a detailed history and various ocular and systemic investigations. Bilateral frontalis sling surgery using silicone slings was performed at a gap of two weeks. The results of surgery were satisfactory. Frontalis sling surgery can be a safe and effective option for ptosis correction in patients with chronic progressive external ophthalmoplegia. The complication of exposure keratitis can be largely prevented by slight undercorrection.

Keywords: Ptosis, Ophthalmoplegia, Frontalis, Exposure keratitis, Bell`s phenomenon.

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1. Introduction

Chronic progressive external ophthalmoplegia (CPEO) is a collection of hereditary myopathies affecting extraocular muscles (EOMs), which commonly manifests as ophthalmoplegia and bilateral ptosis.[1] It is a chronic, bilateral, progressive, typically symmetric, and there is external (i.e., pupil sparing) ophthalmoplegia.[2] Ciliary and iris muscles are typically spared. It affects both males and females equally. The prevalence of mitochondrial disease is 11.5 cases per 100,000 population, worldwide.[3] CPEO can occur as isolated entity involving only eye (isolated CPEO) or in association with other systemic findings ("CPEO-plus"). CPEO plus can be evaluated as part of a neurologic syndrome or systemic myopathies.[1] Ptosis associated with chronic progressive external ophthalmoplegia (CPEO) poses difficult management problems due to poor levator function, limited upgaze and poor Bell`s phenomenon. All these factors lead to increased risk of exposure keratopathy post ptosis correction in these patients. Medical management include use of adhesive tapes or crutch glasses but these provide temporary relief. Surgical management with frontalis sling surgery has shown promising results with or without minor complications.[4] In this case report, we describe a case of severe bilateral ptosis with poor levator function and good orbicularis function and absent bell`s phenomenon. This patient did quite well after ptosis correction with frontalis sling and did not develop exposure keratopathy.

2. Case Presentation

We are presenting a case of 50 years old male who presented Ophthalmology OPD with complaint of drooping of both upper eyelids since 10 years. Ptosis was gradual, progressive and was followed by restriction of extraocular movements. There was no history of diurnal variation of ptosis or any aggravating or relieving factors. No history of any trauma, snake bite or any ocular surgery. No history of proximal muscular weakness or dysphagia. There was no family history of ptosis.

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Ocular examination revealed visual acuity (UCVA) - R/E 6/24 and L/E was 6/9. BCVA was 6/24 in right eye due to presence of cataract and in left eye it was 6/6. Visual axis was orthophoric. His ocular movements (ductions/versions) were restricted in all gazes. There was excessive wrinkling of forehead, eye brows were raised due to frontalis overaction. He assumed a chin up position. The rest of his neurological assessment was unremarkable. His upper eyelids showed bilateral severe ptosis (>4mm) and poor levator function (<4mm) with absent lid crease (**Figure 1**). Bell's phenomenon was absent. Corneal surface and sensations were normal. Pupils were round, regular, normal in shape and size with intact pupillary reaction. Lens in R/E showed cataract and L/E pseudophakia. Fundus examination was normal.

Laboratory investigations were done for CBC, ESR, RBS which were normal. To rule out Ocular Myasthenia Gravis, Tensilon test, Sleep test, ICE test and electromyography (**Figure 2**) were done, all of which were normal. To rule out Graves disease, thyroid function tests were done which were normal. Oculopharyngeal muscular dystrophy was ruled out by a negative family history and absence of dysphagia. The likelihood of Kearns Sayre syndrome was ruled out by age of presentation, normal fundus and normal electrocardiogram and Echocardiography. Muscle Biopsy was advised but patient did not get it done due to unavailability of the test in the area. On the basis of history, clinical assessment and investigations diagnosis of Chronic Progressive external ophthalmoplegia was made.

Patient was advised ptosis correction surgery after explaining all risks and complications. Frontalis sling surgery using silicone slings was performed, first for left eye (**Figure 3**) and after two weeks for the right eye (**Figure 4**). Slight under correction of approximately 1mm was done after explaining to the patient to avoid complication of exposure keratitis. The patient did quite well after surgery and there was no sign of exposure keratitis. Patient was followed up for a period of two years and there were no complications.

Figure 1:

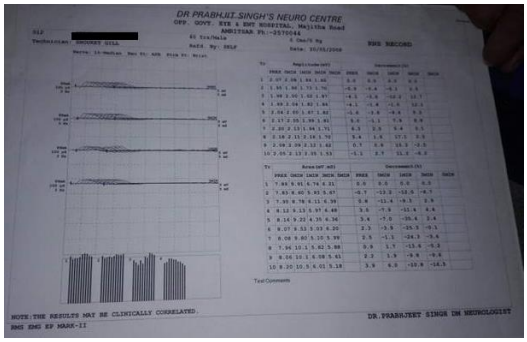

Figure 2:

Figure 3:

Figure 4:

3. Discussion

Mitochondrial myopathies most frequently manifest as CPEO.[5,6] CPEO that occurs in association with mutations in mitochondrial DNA (mtDNA) is usually associated with skeletal muscle weakness. However, mitochondrial defects may differ in individuals with a similar clinical presentation.[3] Most common presentation of patients with chronic progressive external ophthalmoplegia is ptosis and proximal limb weakness.[7] The fact that extraocular muscles are affected preferentially is because their fraction of mitochondrial volume is several times greater than that of other skeletal muscle.[8,9] CPEO usually begins in young adulthood. The first clinical sign of CPEO is usually ptosis, which is usually bilateral and symmetrical. Ophthalmoplegia associated with CPEO may not become apparent for months to years. With the ptosis progression, the patient uses the

frontalis muscle to elevate the eyelids and adopts a chin-up head position. As ptosis often becomes complete, the patient eventually resorts to manual elevation of the eyelids.[4] There is documentation of cases where patients have developed ophthalmoplegia but not ptosis. The ptosis that develops may be unilateral or asymmetric. Patients with CPEO often does not complain of diplopia because of the symmetric nature of this disorder. Patients become aware of their decreased motility when it becomes severe. The downward gaze is usually preserved to a greater extent than horizontal movement or upgaze.[4] The course of CPEO shows constant progression without periods of remission or exacerbation. Exposure keratopathy due to ophthalmoplegia leads to patients complaining dryness of eyes. Adhesive tape and lid crutches are given as a part of medical management of ptosis to assist patients with advanced chronic progressive external ophthalmoplegia (CPEO). A case study discusses successful treatment with a combination spectacle-mounted lid crutch and moisture chamber in a patient with exposure keratopathy after previously unsuccessful lid surgery.[10]

4. Conclusion

Ptosis associated with CPEO is difficult to manage because of increased risk of exposure keratopathy post ptosis correction in these patients due to poor levator function, poor bell`s phenomenon and limited upgaze. All these result in poor corneal protective mechanisms, therefore ptosis surgery often is contraindicated. The complication of exposure keratitis can be prevented by slight undercorrection. The frontalis sling should be adjusted to provide a firm linkage between the eyebrow and the eyelid but should be loose enough to allow eyelid closure when the frontalis muscle is relaxed. This provides lessening of ptosis and relief from visual obstruction while avoiding complication of exposure keratitis. Because a silicone sling is reversible, it could be a possibility for patients with chronic progressive external ophthalmoplegia.

5. Source of Funding

None.

6. Conflict of Interest

None.

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