

Case Report

Frontalis sling surgery: A treatment modality for cyclic oculomotor nerve palsy



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Abstract

Cyclic oculomotor nerve paresis and spasm are a rare disorder characterized by alternating spastic and paretic phase of the muscles supplied by the oculomotor nerve. A 26-year-old male presented with abnormal right eyelid movements since early childhood. Examination findings were consistent with diagnosis of right eye cyclic oculomotor nerve palsy. A paretic phase with complete upper eyelid ptosis, exotropia, hypotropia and dilatation of the pupil was followed by a phase of orthotropia, the absence of ptosis and normal pupil size. The whole cycle was of about 100 s duration with patient having no voluntary control over the eyelid movements and ocular alignment. Systemic evaluation and neuroimaging were normal. A tarsofrontal silicone sling surgery done for the ptosis correction decreased the eyelid movements during different phases of the cycle.

Keywords: Cyclic paralysis, Oculomotor nerve, Ptosis, Tarsofrontal sling

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Introduction

Cyclic oculomotor nerve paresis is a rare condition which is characterized by episodic contraction and relaxation of the muscles supplied by third cranial nerve. Rampoldi first described this condition in 1884 where he reported two cases and termed it as an oculo-palpebral imbalance.¹ The characteristic findings are an alternating paretic phase wherein the eyelid becomes ptotic, eyeball rotates downward and outward, pupil is dilated and a spastic phase when ptotic eyelid elevates, eyeball returns to midline and pupil gets constricted. Various medical and surgical treatment modalities have been described to control the cyclic phases. Baclofen which is a central muscle relaxant was tried and was found to be ineffective.² Levator transposition procedure was effective in abolishing the eyelid movements in 3 patients.³ We report a case of cyclic oculomotor paresis and spasms (COPS) where tarsofrontal silicone sling was performed for correction

of ptosis and was effective in reducing the cyclic eyelid movements and provided good cosmetic outcome.

Case report

A 26-year-old male presented with complaints of abnormal movement of the right upper eyelid since childhood. The abnormal movements developed following an episode of fever at 1.5 years of age. His antenatal and birth history were uneventful. The movements decreased during sleep as noticed by his family members.

On examination, his best corrected visual acuity was 1/60 in the right eye and 6/6 in the left eye. Cyclic movements of the right eyelid and eyeball were noted consistent with COPS. In paretic phase the eyelid became completely ptotic with poor levator action (Fig. 1a). The eyeball was abducted and depressed and extraocular movements were restricted in superior, medial and inferior gaze (Fig. 1b). The pupil

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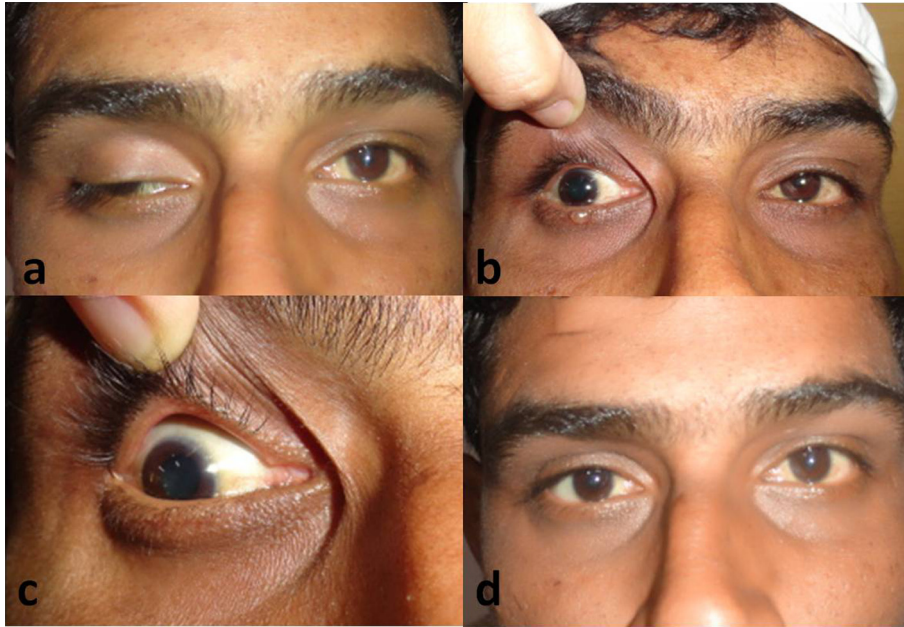


Figure 1. Right eye cyclic oculomotor nerve paresis. Note the complete ptosis (a), exotropia and hypotropia (b) and dilated pupil (c) during the paretic phase and complete disappearance of ptosis, orthotropia and normal pupil size during the spastic phase (d).

diameter was 6 mm during this phase (Fig. 1d). The paretic phase lasted for 55 s and was followed by the spastic phase when the eyelid lifted up with complete correction of ptosis, eyeball was orthotropic, extraocular motility was normal and pupil constricted to normal size (Fig. 1d). The spastic phase lasted for 45 s. These cycles of paresis and spasm were repeated continuously. Anterior and posterior segment examinations were normal in both eyes. There was no other neurological deficit and his MRI brain was normal.

Patient was cosmetically concerned and Bells phenomenon was fair in the right eye. A tarsofrontal sling surgery was performed in the right eye using silicone sling material and Fox pentagon technique. Intraoperatively, the sling was tightened and eyelid height was adjusted to align the upper eyelid margin with the upper limbus to allow for the slight postoperative eyelid droop which happens due to elastic nature of the silicone sling material. At 12 months follow-up, the palpebral fissure heights in primary gaze were 11 mm in both eyes. There was 1 mm lagophthalmos in the right eye and no exposure keratopathy. The eyelid position remained constant during the paretic and spastic phases of the cycles (Fig. 2). Extraocular movements varied during the different phases.

Discussion

COPS is a non-progressive disorder involving rhythmic contractions and relaxations of the muscle supplied by third cranial nerve. The pathophysiology behind this phenomenon remains debatable. Loewenfeld and Thompson suggested that the primary lesion involves the oculomotor nerve nucleus.⁴ Bateman et al. suggested involvement of peripheral part of the nerve as depicted by electromyography changes.^{5,6} The most accepted theory is a combination of central and peripheral nerve damage which produces this cyclical phenomenon as suggested by Lowenfeld et al.⁴

COPS is an uncommon entity and in majority of patients (92.5%) this condition is noted before 1 year of age.⁴ The etiology could be either birth trauma or infections such as measles as was reported in half of the cases.⁴ Our patient also had history of fever at 1.5 years of age following which the abnormal movements developed.

The regular cyclical pattern of paresis and spasm are the hallmark of COPS with the paretic phase reported to last from 1 to 3 min and the spastic phase lasting for 30–100 s.⁷ Isolated involvement of the iris sphincter and ciliary muscles have also been reported and levator palpebrae is the third most

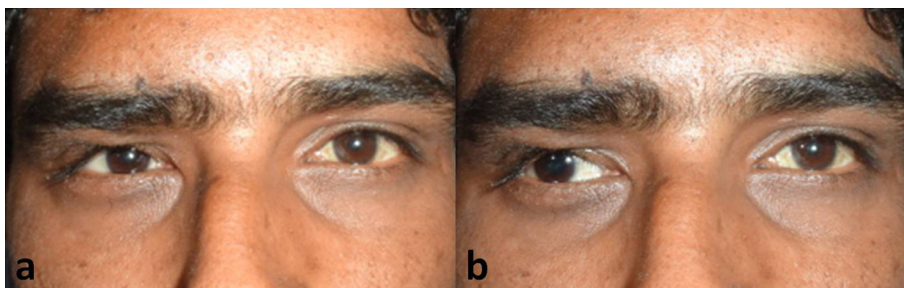


Figure 2. Appearance of the right eye following tarsofrontal silicone sling surgery showing no change in the eyelid position during the paretic and spastic phases. Note the position of the eyelid during the spastic phase (a) and the position during the paretic phase (b).

frequently involved muscle in COPS.⁷ The right eye is more commonly involved than left eye (2:1) and there is no significant gender predominance.⁴ Neuroimaging is normal in most patients and a supraclinoid lesion has been reported in one patient.⁵ Several medical and surgical treatment techniques have been tried for this condition with variable results. Central muscle relaxant Baclofen was ineffective in controlling the cyclic movements.² Levator muscle transposition was found effective in abolishing the cyclic eyelid movements in 3 patients.³ Our patient was cosmetically concerned about the cyclic movements of the eyelid as the eyelid movements are the most noticeable and most bothersome component of this disease. A tarsofrontal sling surgery was done using silicone material for correction of the ptosis. This simple procedure was able to correct the ptosis and abolish the eyelid movements during the paretic and spastic phases of the cycle. A sling procedure was preferred over levator resection as measurement of the levator function was unpredictable in this patient. Also, the cyclic spasms and paresis are likely to be persistent after a levator resection. In tarsofrontal sling procedure the action of the levator muscle is bypassed and the eyelid is suspended at the desired position by the sling material and eyelid height is maintained by the frontalis muscle action which is transmitted to the eyelid via the silicone sling. This mechanical suspension of the eyelid will prevent the eyelid droop which happens during the paretic phase of the COPS.

In conclusion, this report describes a case of COPS managed by tarsofrontal silicone sling surgery. Tarsofrontal sling surgery using silicone material is a simple technique and is effective in controlling the cyclic movements of the eyelid during the different phases of COPS.

Conflict of interest

The authors declared that there is no conflict of interest.

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