

A Case of Complex Congenital Ptosis

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Abstract:

AIM: To report a case of a 6 year old girl with severe congenital ptosis associated with chin elevation, bilateral esotropia with high myopia.

METHODS: A 6 year old girl whose informant was the mother, presented with drooping of both upper lids and inward deviation of both eyes since childhood. On examination, she was found to have chin elevation, 40 PD bilateral esotropia, bilateral myopia and bilateral severe ptosis with poor LPS function.

RESULT: She underwent bilateral recession resection surgery for esotropia with frontalis sling using silicon rod for ptosis. She was provided spectacle correction for myopia and occlusion therapy. Her vision has improved and the abnormal head posture is absent.

CONCLUSION: It is essential for all patients with congenital ptosis to have a comprehensive ophthalmic evaluation to look for any associated ocular conditions which should also be treated promptly to provide both visual and cosmetic benefit to the patient.

Keywords- Amblyopia, Bells Phenomenon, Frontalis Sling, Ptosis, Strabismus

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I. Introduction

Congenital ptosis is a relatively common condition. It accounts for approximately one per cent of all ophthalmic surgery performed. Congenital ptosis is due to a dysgenesis of the levator complex, with the muscle fibres being replaced by fibrous and areolar tissue. ^[1]There is correlation between the degree of ptosis, and the amount of abnormal tissue and hence levator function,. This leads to the classical clinical picture of ptosis in the primary position and in upgaze, lagophthalmos in downgaze, decreased levator function, poor eyelid crease and poor eyelash position. Surgical correction is required if the ptosis occludes the visual axis causing amblyopia, for an abnormal head posture or for purely cosmetic reasons. Simple unilateral congenital ptosis rarely occludes the pupil to produce amblyopia. ^[2,3,4]

Early detection and treatment of amblyogenic factors such as refractive errors and strabismus is recommended and, when the pupillary axis is occluded with severe ptotic eyelids, surgery should be performed as soon as possible to prevent stimulus deprivation amblyopia. Most cases of congenital ptosis are idiopathic. However, congenital ptosis may occur through autosomal dominant inheritance. Common familial occurrences suggest that genetic or chromosomal defects are likely. There is no known racial or gender preference, and roughly 75% of cases are unilateral. ^[5]

II. Materials And Methods

A 6 year old girl (informant – Mother), presented with drooping of both upper eyelids since birth and inward deviation of both eyes noticed at the age of three years. There was a progressive increase in the drooping of eyelids with a gradual adoption of a chin elevated head posture. There was no history of consanguinity with normal antenatal history. She was a preterm hospital delivery at 28 weeks with birth weight 1.5kgs. There was no history of birth trauma and social and motor developmental milestones were normal. Immunization status was normal for age and there was no family history of similar complaints.

On examination, there was chin elevation. (Fig.1)The palpebral aperture height was 6mm in the right eye and 5mm in the left eye and the width was 26mm in each eye. The lid crease was absent. (Fig.1) The margin reflex distance 1(MRD 1) was minus 1mm in both eyes with zero LPS action and Bells phenomenon. Phenylephrine test and Ice pack test were negative in both eyes. Corneal sensations were present and Marcus Gunn jaw winking phenomenon was absent. Ocular movements showed bilateral absence of elevation. Forced duction test done under short GA was negative. She had a visual acuity of 4/60 in right eye and 2/60 in the left eye. Refraction OD: - 6.0x90°; -6.0x180° and OS: -7.0x90°;-7.0x180°. BCVA OD: 6/24 with - 8Ds; OS: 6/36 with -9Ds. There was an alternate esotropia of 40 and 45 PD for distance and near respectively with no change after patching of one eye for 45 minutes. Worth Four dot test showed alternate suppression. Fundus was within

normal limits. Axial lengths of the right and left eye were 23.5mm and 23.1mm respectively. Keratometry values were 44.5D in the right eye and 43.75D in the left eye. B scan was within normal limits.



Fig.1 Chin elevated posture of the head



Fig.2 Bilateral congenital ptosis (note absence of lid crease)

III. Results

Bilateral Lateral Rectus Resection (8mm) under GA resulted in residual esotropia of 30PD. This was followed by Bilateral Medial Rectus Recession and frontalis sling surgery with silicone sling. As there was no Bells phenomenon, the aim of the surgery was to counteract the chin elevated head posture only and a full cosmetic correction was not attempted. She was prescribed spectacle correction for myopia, and was advised occlusion therapy for amblyopia.

The patient was on regular follow-up and at the end of six months, she was orthophoric with a normal head posture. (Fig.3) The palpebral aperture height of 9mm and BCVA 6/12 with -5.0Ds-1Dc@180° OD and 8mm along with BCVA 6/12 with -5.5Ds-1Dc@180° OS. (Fig.4)However, restriction of elevation persisted



Fig.3 Normal head posture after sling surgery for the ptosis



Fig.4 Increase in the size of the palpebral aperture after sling surgery

IV. Discussion

The overall incidence of amblyopia varies between 1.6 to 3.6% whereas the overall incidence of amblyopia in strabismic population is 59.9%.^[6] In the study by Hornblass A et al, amblyopia was detected in 7 of 36 (19%) patients with congenital ptosis.^[7] Harrad RA et al (1988) in his study of 216 cases found 17% of patients with simple congenital ptosis had amblyopia of which 14% was attributable to the ptotic occlusion of the pupillary axis resulting in stimulus deprivation, 21% patients had anisometropic amblyopia and 51% had strabismic amblyopia. Early refraction, orthoptic assessment and treatment and, where the pupillary axis is occluded, surgery to prevent stimulus deprivation amblyopia were recommended.^[8] In the study by Anderson RL (1980), the overall incidence of strabismus in patients of congenital ptosis was found to be 32%.^[9] In the study conducted by Gusek-Schneider (2000) on unilateral ptosis, the frequency of myopia was 14%.^[10] In the study conducted by Thapa R (2010), among the 78 cases (95 eyes) of congenital ptosis, refractive error was present in 13 cases (16.7%) with astigmatism as the commonest refractive error (8.9%).^[11]

In the study conducted by Srinagesh V et al (2011) on patients with congenital ptosis, amblyopia was seen in 23.9%, and in majority of these cases, there was coexisting anisometropia or strabismus in the eye with the more severe ptosis. Children with congenital ptosis are at risk of anisometropic and strabismic amblyopia, which may progress during early development. Patients should be examined at regular intervals to evaluate and treat these potential complications.^[12]

Amblyopia in congenital ptosis may result from anisometropia, induced astigmatism, associated strabismus or occlusion of the pupillary axis. The primary factor in the production of amblyopia is a modification of normal visual experience during a period when the visual system is susceptible to such alterations. In spite of the aetiological difference in the nature of the underlying clinical conditions, common amblyogenic factors appear to be operative in strabismic, anisometropic, and pupillary occlusion amblyopia. These causative mechanisms are stimulus deprivation by inadequate image formation on the fovea, abnormal binocular interaction by incongruity of the visual input received by the two eyes, and perhaps a combination of both factors. Amblyopia in complex congenital ptosis should be prevented by a combination of early refraction, orthoptic assessment and surgical treatment for ptosis and squint.^[13]

V. Conclusion

Amblyopia remains a leading cause of treatable monocular vision loss in children and adults. Given that the associated factors that place a child at risk for amblyopia are identifiable, ideally there would be an inexpensive, accurate, and fast test to detect these factors at an age when amblyopia is most responsive to treatment. An apparently obvious ocular condition such as strabismus or ptosis alerts the family and pediatrician that referral to an ophthalmologist is in order. Effective treatment must be initiated during the ages that the visual system is immature. In general, the younger the child, the more effective is the prevention and treatment of amblyopia.^[14] Strabismus and amblyopia have a much greater prevalence in patients with congenital ptosis than in patients with normal eyelid function. Both strabismus and squint are amblyogenic. Therefore, it is essential for all patients with congenital ptosis to have a comprehensive ophthalmic evaluation.^[15]

Management of a patient with congenital ptosis extends beyond the surgical correction and includes evaluation for associated ophthalmic disorders, appropriate referral if underlying systemic disorders are suspected, and close monitoring of the visual status until the patient has passed the amblyogenic stage of development. If amblyopia is present and is related to the eyelid malposition, early surgical intervention is required to maximize the potential for normal visual development.^[16] We have reported on a case of bilateral complex congenital ptosis with double elevator palsy and poor Bells phenomenon and hence probably of nuclear origin associated with infantile esotropia, high myopia and amblyopia. Amblyopia was due to a combination of anisometropic, stimulus deprivation and strabismic amblyogenic factors.

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