Primary Infantile Exotropia with Lambda Pattern- A Case Report

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ABSTRACT

Infantile exotropia is a relatively rare strabismus disorder characterized by outward deviation of one or both eyes. Because of its rarity, there are few population-based studies to identify its true incidence. Here is a 12years old child presented with abnormal outward eye deviation since birth. Ocular examination revealed infantile exotropia with lambda pattern which is among the rarest case ever found of which squint surgery was done and the alignment was brought back to normal. Definitive treatment is surgery before age of two years to have best binocular single vision outcome, wherein this patient lost his binocular single vision due to the delay.

Keywords: infantile exotropia, lambda pattern, strabismus, binocular single vision, recession, resection, transposition.

INTRODUCTION

Infantile exotropia is a relatively rare strabismus disorder characterized by outward deviation of one or both eyes. Its onset is almost always at birth or sometimes before age of 6 months and persists beyond this age[1]. Infantile exotropia commonly presents without any pattern though some patterns like A, V and X has been reported. Among the rarest case is lambda pattern that is alignment is ortho in up gaze and primary gaze, with an exodeviation in downgaze.

In primary infantile exotropia; patients are usually healthy with no evidence of systemic or ocular disease as seen in this case while secondary infantile exotropia they usually have either ocular diseases like retinoblastoma, albinism, Duane syndrome, Brown syndrome or systemic diseases such as cerebral palsy, hydrocephalus, craniofacial syndrome or Prader- Willi syndrome[1,2]. Because of its rarity, there are few population-based studies to identify its true incidence. Researchers estimated this at 1 in 30,000 births in the general population[3,4].

Infantile exotropia causes have been reported in many theories such as neurophysiologic cause, this includes either deficient development of the convergence system during the vulnerable early period between two to four months of age. Sometimes it may be due to monocular defocusing and total deprivation since birth. Disruption of binocular vision shortly after birth that alters the development of normal vergence reflexes until three months of age or disruption of normal binocular processing in the visual cortex due to neuro-anatomic causes can lead to infantile exotropia. Other theories suggest asymmetry in muscle structure between the lateral rectus and medial rectus or increased in diameter of the lateral rectus compared to the medial rectus. However, in the above theory, whether the deviation caused the changes in the muscle or the muscle anatomy led to the deviation is unclear[3].

CASE PRESENTATION

A 12 years old child attended eye clinic with the complaint of abnormal outward eye deviation since birth. Mother reported her child having abnormal outward deviation since birth, which is present all the time and alternates from one eye to other. There is no history of abnormal head posture, fever, excessive vomiting, convulsion, loss of consciousness or trauma.

There was no any history of blurry vision, double vision, thus patient had no complaint during his studies at school but was been bullied with several bad names due to cosmetic appearance at school and home / neighbours. He is the only son, currently in form one. No history of squint in the family in both paternal and maternal side.

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On examination vital signs were normal, he had manifest squint without any facial asymmetry and the systemic examination for all systems were normal.

Visual acuity was 6/6 in both eyes, intraocular pressure was 12mmHg right eye and 13mmHg in left eye, cornea and lens were transparent and fundus of the eye was normal without any pathology in both eyes.

Orthoptic assessment: Hirschberg corneal reflex test showed 30^0 nasally, cover and alternate cover test revealed alternating exotropia deviation, ocular motility was full in all gazes with lambda pattern. Prism cover test showed 60PD BI (prism dioptre base in) for distant and 50PD BI for near. Up gaze was 50PD BI, primary gaze 50PD BI and downgaze 60PD BI which states he has clinically significant lambda pattern. Worth 4 dot test both for distant and near showed alternate suppression. He had no fusion and TNO test revealed no stereopsis.

The diagnosis of primary infantile exotropia with lambda pattern was made because of exodeviation since birth, corneal reflex test revealed nasally, cover and alternate cover test had alternate exodeviation, prism cover test was more than 10PD BI and ocular motility was full in all gazes with clinical significant lambda pattern.

Patient had undergone cycloplegic refraction which showed plano in both eyes. Full blood picture was normal. He was scheduled two visits of three months apart for follow up of stability of deviation angle and thereafter planned for strabismus surgery for cosmetic reason since the angle remained stable for all his visits.

Under general anesthesia, aseptic technique, force duction test was negative both eyes, peritomy at 2-5 o'clock done in left eye, lateral rectus muscle identified, 8.5mm recession with half tendon inferior transposition was done using 6-0 spatulated vicryl suture. Peritomy from 7-11 o'clock done in left eye, medial rectus muscle identified, 5.5mm resection with half tendon superior transposition was done using 6-0 spatulated vicryl suture. Conjunctival was sutured using 9-0 vicryl. Subconjunctival medication given; eye padded. Post-operative analgesics and antibiotics given.

Figure 1: Preoperative findings of the patient in all nine gazes.

Post-operative findings were visual acuity 6/6 in both eyes, corneal reflex < 10^0 nasally, cover test revealed exodeviation and alternate cover test had exotropia. Ocular motility was full in all gazes with no pattern. Prism cover test for distant was 8PD BI and near was 5PD BI. Up gaze, primary gaze and down gaze all showed 5PD BI hence, no pattern observed.

Figure 2: Postoperative findings of the patient in all nine gazes.

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Patient and his mother thanked millions time for making her child no more bullied by friends and relatives with different names and socially acceptable appearance of the eyes; since he has normal gaze now.

DISCUSSION

Primary infantile exotropia presents with eye outward deviation since birth as seen in this case. Parents and other persons who have regular contact with patient usually see abnormal eye because of its large degree of deviation; it usually exceeds 30PD and may be a large as 90PD[4,5], similarly this patient had 60PD. Refractive errors are like that of normal infant population, since this case had plano on cycloplegic refraction. Strabismic amblyopia is seen in a minority of patients, contrarily this patient did not have it. Sometimes pattern deviations may also occur such as V patterns are much more commonly observed than A patterns, and X patterns seen in tight lateral rectus syndrome. Lambda pattern is very rare[5], as occurred in this case, hence a rare case reported.

Surgery is required almost universally in cases of true infantile exotropia which should be performed within the first 6 months of life to maximize potential for attaining binocular single vision with stereopsis and sensorimotor results are obtained in healthy infants aligned before the age of two years[6]; due to the delay of surgery this child lost his stereopsis. Once diagnosis is made, after only a few weeks of follow-up to establish repeatable, reproducible angles of deviation and amblyopia treatment has commenced, surgery should be performed with minimal delay in contrast to this, the patient delayed for more than 10years and lost his binocular single vision. The goal of surgery is to align the patient's eye to within 8PD of orthotropia of which our patient attained 5PD. Its usually recommended to recess lateral rectus muscle and resect medial rectus muscle based on established surgical tables as done with this patient[1,2,3].

Sometimes bilateral lateral rectus muscle recessions can also suffice. Knapp advocated the vertical displacement of horizontal recti to correct for pattern strabismus. When there is a pattern in the absence of oblique muscle overaction, upshifts or downshifts of the horizontal recti can be used to collapse the pattern of which similarly done to this patient and the results were promising[1]. It has also been observed that postoperative surgical outcomes appear to be influenced more by the duration of the misalignment, rather than the age at surgery. This would suggest that the timing of early surgery is of equal importance in the management of infantile exotropia[7].

CONCLUSION

Early detection of infantile exotropia and timely squint surgery before the age of two years retains binocular single vision and good alignment of the eye.

Consent

Written informed consent was obtained from the patient and his mother for the publication of this case report and accompanying images.

Conflict of interests

The author declares that there is no conflict of interests.

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