Case Report

Convergent Acquired Strabismus Fixus in High Myopia Patient- an Overview

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Abstract

Purpose: To identify and report such rare entity and keep special concern to manage such case. **Method:** It is a prospective study. We hereby reviewed six cases of this rare entity selected from pediatric outpatient department of Ispahani Islamia Eye Institute & hospital since January '2019- December'2019. Selected patients underwent detailed history taking, cyclo refraction, slit lamp examination, orthoptic evaluation, fundoscopy and nystagmus evaluation. **Result:** Average age of presentation was 8+/-2 years. Aided visual acuity of two patients was (3/60-6/60), three (6/60-6/24), one (6/24-6/12). Four patients had positive family history of high myopia. On slit lamp examination revealed deep anterior chamber. One case showed normal fundus where remaining fives had characteristic myopic fundus. Four were associated with jerky nystagmus. Three of them had right beating and one left beating nystagmus. Head tilt was present in one patient. Ocular motility was free for two cases whereas restricted in variable degree in four cases may be due to herniation of progressive enlarging myopic globe supero temporally and retro equatorially through muscle cone. **Conclusion:** Understanding the different spectrum of presentation of high myopic patient facilitates early diagnosis, rehabilitation and management of such patient.

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Introduction

Myopic strabismus fixus is an acquired restrictive strabismus occurred in patients with pathologic myopia. This is a form of acquired strabismus fixus which is either monocular or binocular. Another term for the high myopia esotropia syndrome is heavy eye syndrome. These patients may be manifest with extreme esotropia, hypotropia and restrictive ocular motility. This disease may progress over several years, from a small degree of esotropia with free ocular movement to the end stage of large angle fixed esotropia.

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This rare entity of combined esotropia and hypotropia is due to massive expansion of posterior globe against a tight medial rectus and displaced lateral and superior rectus muscle which change the normal vector force. Slippage of the lateral rectus muscle is towards the superior temporal quadrant probably due to the laxity of its pulley system which weakens the abduction vector and turns the globe downwards. Normally, the orbital connective tissue acts like a pulley supporting the extra ocular muscles. The elongated eyeball mechanically compresses the pulley consequently it fails to maintain the normal position of extra ocular muscles and prolapse of the elongated posterior eyeball where's the lateral rectus (LR) muscle is displaced inferiorly and superior rectus (SR) muscle is displaced nasally. In case of late presentation, it often results in significant cosmetic concerns and technical difficulties for ophthalmological examinations.

Sometimes this rare entity is misdiagnosed and mal treated due to lacking of proper evaluation. Purpose of the study is to create attention, identify and keep special concern in management of such case.

Method:

It was a prospective observational study; we hereby reviewed six cases of the rare entity since January 2018-December 2019 in pediatric department of Ispahani islamia Eye Institute & Hospital.

Children between three to 16 years old who had high myopia with eso deviation were included and same criteria with developmental delay, previous strabismus surgery, associated ocular pathology like nerve palsy were excluded from this study. After selection detailed history was reported like family history of refractive error and strabismus, previous ocular surgery, history of trauma, age of onset of strabismus, age of presentation.

Visual acuity was measured by using Kay picture chart and snellen's test type. Cycloplegic refraction was done by Cyclopentolate 1% eye drop or atropine eye drop according to age. Slit lamp examination was done for anterior segment evaluation; intraocular pressure was measured by tonopen. Indirect ophthalmoscope was used for background fundus evaluation.

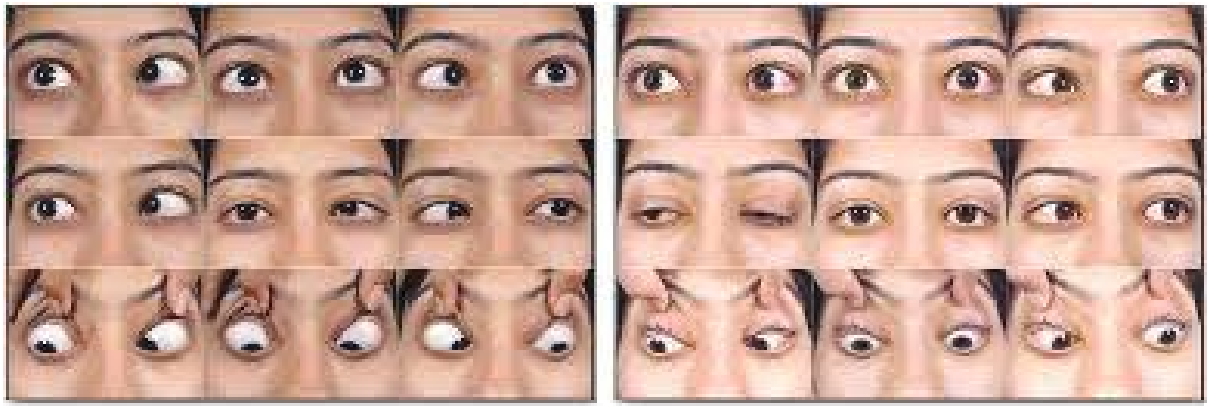

Fig: Ocular motility

Orthoptic evaluation was carried out which included Hirschberg's reflex, prism cover test for cooperative child, Krimsky test for very young and non-cooperative child. Ocular motility was tested. Binocular single vision test was conducted by using stereo fly test to see the level of stereopsis. Nystagmus was evaluated which included type, direction, amplitude and frequency of nystagmus. Orbital imaging was done to see the size of globe and displacement of any muscle like- lateral rectus and superior rectus muscle.

The degree of restriction of extraocular movement was graded into 4 grades of impairment. Grade 1 is small restriction; Grade 2 is moderate restriction; Grade 3 is severe restriction in which eye could not cross midline; Grade 4 is extreme restriction with eye fixed in adduction with minimal movement.

Fig: Orthoptic evaluation

Result:

Average age of presentation was 8 ± 2 years. Aided visual acuity of two (33.3%) patients was (snellen3/60-6/60 logMAR1.3-0.1), three (50%) patients was (snellen6/ 60-6/24 logMAR0.1-0.6) and one (16.7%) patient (snellen6/24-6/12 logMAR0.6-0.3).

Deviation: Three (50%) patients had eso deviation within25-35 prism diopter (pd) and two (33.3%) patients had 35-45pd whereas one (16.7%) had 40- 50 pd.

Binocular single vision was absent for four (66.7%) cases, 100 degree of arc for two (33.3%) cases, 80 degree of arc for one (16.7%) case.

Ocular motility: -1 restriction in abduction for two (33.3%) cases whereas-2 to -4 restrictions in four (66.7%) cases.

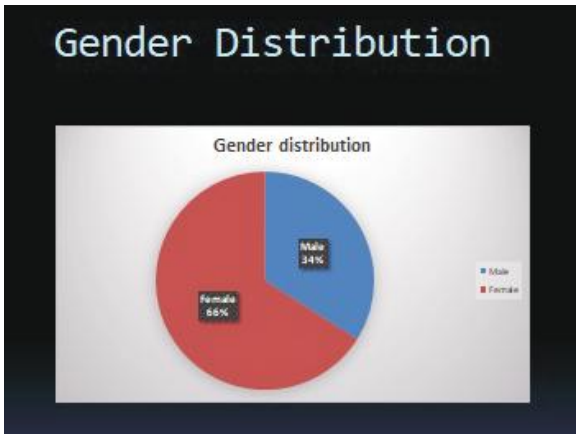

Fig: 1 Gender distribution

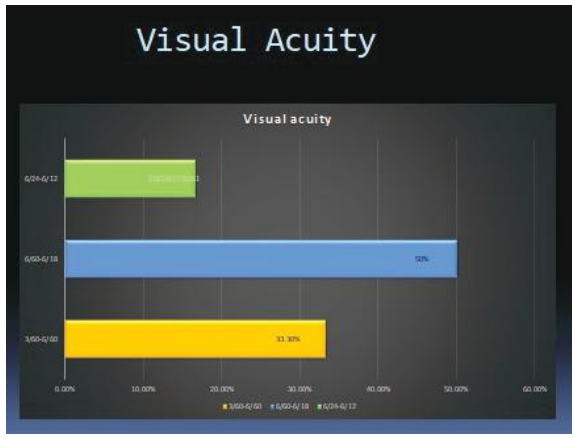

Fig: 2 Visual Acuity

Fig: 3 Ocular motility

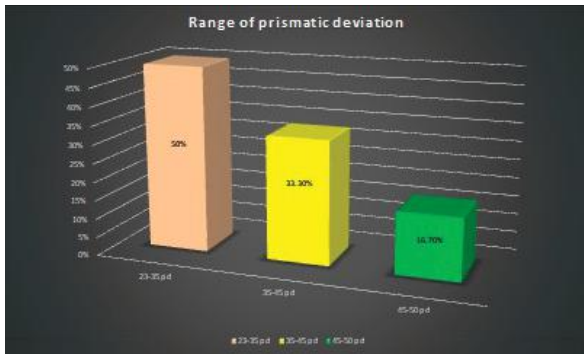

Fig:4 Prismatic deviation

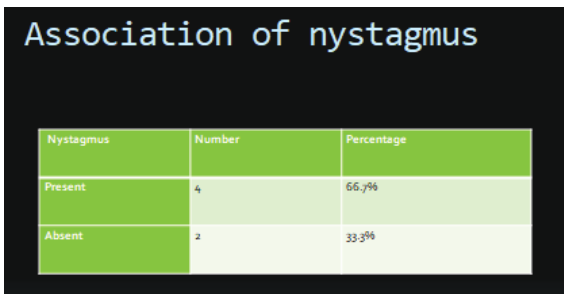

Fig:5 Association of nystagmus

Discussion

Convergent acquired strabismus fixus in high myopia patient is not too much common in our country aspect. Presentation pattern is different from usual myopic patient. Management is also different. It is characterized by not only tight muscle but also extreme shortening of conjunctiva, tenon and basically all orbital tissues. It was initially thought to be caused by the increased weight of elongated myopic eye and subsequent prolapse of its anterior portion. Over the years, many theories have been presented. Researchers have attributed this progressive esoptropia to compression of lateral rectus muscle against the lateral orbital wall [5,6] whereas Demer and von Noorden showed mechanical restriction from contact between the posterior globe and bones of the orbital apex.

Strabismus fixus was first described as a type of retraction syndrome – congenital structural anomaly (fibrosis of medial rectus). Villaseca described this condition as different from retraction syndromes and documented this condition as an acquired condition rather than a congenital anomaly.[1] He suggested that fibrosis of the medial rectus is a consequence following lateral rectus paralysis rather than a primary anomaly.

Krzizok et al.3 described its pathophysiology by using MRI scans on 37 patients with high myopia, they showed that 13 were demonstrated as esotropia and hypotropia with lateral rectus was 3.4 mm inferiorly displaced. In the two patients with exotropia and hypotropia, the medial rectus was downwardly displaced. MRI studies also showed superotemporal prolapse of the elongated posterior portion of eyeball leading to an inferior shift of the lateral rectus and nasal shift of the superior rectus. This was previously described by Yokoyama et al.4 The supero temporal quadrant is particularly susceptible to globe prolapse, as ligament that joins lateral rectus and superior rectus band degenerate over time.

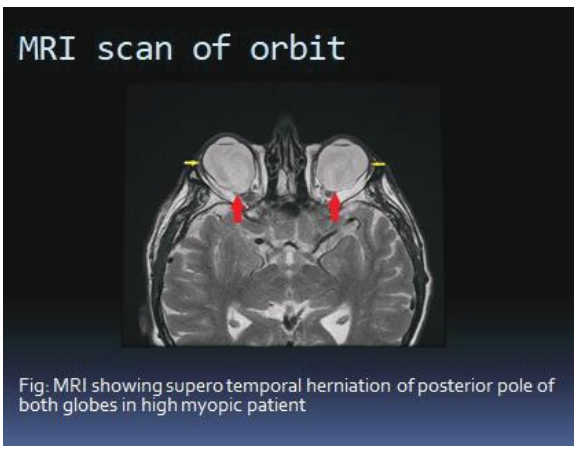

Fig: MRI showing supero temporal herniation of posterior pole of both globes in high myopic patient

Krzizok et al.3 noted a change in the muscle path of the lateral rectus muscle on MRI 3. They noticed an inferior displacement of the lateral rectus and nasal displacement of superior rectus muscle which leads to limitation of abduction and elevation, respectively. The inferior displacement of lateral rectus causes it to act as a depressor more than as an abductor, whereas nasalization of the superior rectus causes it to act as an adductor than as an elevating muscle. 3 These changes lead to the patient developing esotropia and hypotropia.

Our study is not without limitation. This is a prospective study with relatively small sample size. However, because of the rarity of this disease, our case series is one of the largest series reported in our country.

Conclusion:

To avoid complication and providing better visual future; it is critical to timely diagnosis, appropriate work up and proper intervention .To understand the different presentation pattern of this rare entity facilitates early diagnosis, rehabilitation and reducing ocular morbidity.

Financial disclosure:

The authors declare that they have no competing interests.

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