Case Report

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A RARE CASE OF BILATERAL DUANE'S RETRACTION SYNDROME WITH MUTE AND DEAFNESS.

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ABSTRACT **Background:** Duane's retraction syndrome (DRS) is a congenital disease characterized by limitation in adduction and/or abduction eye movements and shortening of the interpalpebral fissure during adducting, and could also involve eyeball retraction, upshoot or downshoot. Various systemic anomalies, syndromes, and other ocular variations may occur in Duane's Retraction Syndrome. **Case Report:** A 12-year-old female was brought to the hospital for a routine ocular examination without symptoms. The examination demonstrated left eye esotropia in primary gaze, an abduction deficit O.U., and O.S. globe retraction with palpebral fissure narrowing on dextroversion and vice versa. The history and examination are suggestive of a bilateral congenital type I DRS. **Conclusion:** Most cases of DRS are observed as isolated conditions. However, various ocular and systemic abnormalities are associated with DRS. Therefore, a complete ocular examination and detailed case study are crucial in the diagnosis of cases with DRS.

KEYWORDS : Duane's retraction syndrome, Bilateral strabismus, Stilling-Turk-Duane syndrome, globe retraction, palpebral fissure narrowing

INTRODUCTION

Duane's retraction syndrome (DRS), also known as Stilling-Turk-Duane syndrome, is defined as a congenital ocular movement disorder of the horizontal recti muscles, resulting in an impaired ocular motility syndrome that includes palpebral fissure narrowing. [1] The incidence of DRS is approximately 1% of the total cases of strabismus. [1] Eighty percent of cases are unilateral and characterized by either limited abduction, limited adduction, or both. Three varieties can be seen. [1]

Type 1: This is the most common variety presenting with - (a) A marked restriction or complete abolition of abduction of one, or rarely of both eyes. (b) A mild reduction of adduction in the involved eye. (c)When attempting adduction, an apparent retraction frequently accompanied by an upshoot or a down shoot in the affected eye occurs. (d) Slight ptosis and shortening of the interpalpebral aperture during adduction (as a result of retraction of the eye. (e) When trying to abduct, there is a slight widening of the interpalpebral aperture and retraction of the upper eyelid (due to minimal protrusion of the eye). (f) Convergence insufficiency is where the affected eye fails to fully converge while the other eye converges correctly. [2]

Type 2: This is rarer. In this case, convergence and normal adduction are present alongside faulty abduction.[2]

Type 3: Both adduction and abduction are faulty in this variation, although the former is far more affected.[2]

CASE REPORT

A 12year old female was brought to our Ophthalmology OPD by her mother for a regular ophthalmology workup. The mother also gave a history of restriction of movements in both eyes. The child was the only child in the family, born out of a non-consanguineous marriage. There is no family history of strabismus, her strabismus is also associated with muteness and deafness.

Examination

On examination, the child had no anomalous head posture and no facial asymmetry but she had left eye esotropia. Anterior segment and dilated fundus examination of both eyes were normal. Her uncorrected visual acuity was 6/12 in the right eye, Finger Counting 3M in the left eye, and N6 in both eyes. Her best-corrected vision is 6/6 and 6/9 in right and left eye respectively and N6 in both eyes. She exhibited fusion which was tested with Worth's four dot test for distance.

Extra ocular motility examination demonstrated limitation of abduction in both eyes with narrowing of palpebral fissure on adduction and widening of palpebral fissure on attempted abduction. Globe retraction was noted in both eyes but more significant in the right eye. The lacrimal apparatus in both eyes was functionally and anatomically normal. In addition to the ocular findings, there was an associated mute and bilateral deafness as well. A conclusive diagnosis of bilateral Type-I Duane's Retraction Syndrome (DRS) was hence made.

Treatment

Since the patient had no complaints of diplopia and abnormal head posture, also the patient did not have cosmetically significant globe retraction, the patient was advised regular follow-up and no surgical intervention.

DISCUSSION

Duane's retraction syndrome is more common in females. It usually occurs as an isolated sporadic entity although familial patterns have been observed in 5 to 10 % of cases. [3] Even though involvement is most frequent on the left side it is bilateral in 18% of cases. A number of associated findings have been reported with the retraction syndrome. This has led some authors to propose that a teratogenic insult at about eight weeks gestation may be the cause. The associated anomalies include spinal deformities, ear malformations and hearing defects, palatal changes, epibulbar dermoid or lipodermoids, naevus of Ota, Brown tendon sheath syndrome, micro cornea, Smith-Lemli-Opitz syndrome, Wildervanck syndrome. [4] Perceptive deafness is associated with 18% of cases. [4]

Bilateral DRS is a rare syndrome. In this case, this syndrome is associated with other congenital anomalies like muteness and bilateral deafness as well, making it an even rarer occurrence.

CONCLUSION

DRS is a rare syndrome and bilateral DRS is even more so. Any child with bilateral DRS should be completely evaluated to rule out other syndromic associations. Immediate and early ophthalmological interventions with refractory error correction, the use of prisms, and surgical corrections, will be helpful in making life more normal for these children.

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