Doi: 10.32677/IJCR.2017.v03.i02.013 Case Report

Rare association of Keratoconus in the contralateral eye in a case of Duanes retraction syndrome

Raman Yenugandula, Rekha Priya Kalluri Bharat

From, the Department of Pediatric Ophthalmology and Strabismus, Sarojini Devi Eye Hospital, Hyderabad. TS India.

Correspondence to: Dr Raman Yenugandula, Sarojini Devi Eye Hospital, Hyderabad. Email: drraman33@gmail.com

Received: 10 Mar 2017 *Initial Review: 01 Apr 2017* *Accepted: 09 Apr 2017* *Published Online: 21 Apr 2017*

ABSTRACT

Duanes retraction syndrome is characterised by restriction of ocular motility, changes in palpebral fissure width and globe retraction. Keratoconus is a bilateral, progressive non-inflammatory corneal ectatic disease characterised by central or paracentral thinning and steepening causing irregular astigmatism. We report a case of rare association of DRS with keratoconus in the contralateral eye in a thirteen year old male child. The ocular motility revealed left DRS with exotropia. Corneal topography, based on new Scheimpflug and placido-disk analyser, revealed the findings consistent with features of keratoconus in the right eye. There were no associated systemic abnormalities. This case gives evidence of the importance of cycloplegic refraction which provided a clue towards suspecting keratoconus. It also highlightsthe fact that a patient with DRS should be examined thoroughly to rule out any associated ocular anomalies.

Keywords: Duanes retraction syndrome, Heischberg corneal reflex test.

Duanes retraction syndrome (DRS) is a special type of strabismus characterised by limitation of horizontal eye movements, narrowing of palpebral fissure on adduction and retraction of globe [1]. There may be an upshoot or downshoot on adduction. It is usually unilateral with 18% bilateral incidence. It is caused by an innervational disturbance originating in the brainstem [2]. Numerous ocular and systemic anomalies have been reported with DRS such as dysplasia of the iris stroma, pupillary anomalies, cataracts, heterochromia, persistent hyaloid arteries, choroidal colobomas, distichiasis, crocodile tears, and microphthalmos. Numerous systemic anomalies [3,4] include Goldenhar's syndrome [4,5], facial hemiatrophy [6], dystrophic defects such as the Klippel-Feil syndrome, arthrogryposis multiplex congenital [7], cervical spina bifida, cleft palate, sensorineural hearing deficits [8], Chiari's type I malformation, deformities of the external ear, and anomalies of the limbs, feet, and hands.

Keratoconus is an ectatic disorder characterised by central or paracentral corneal thinning and steepening that leads to corneal surface distortion. Visual loss occurs primarily from irregular astigmatism and myopia and anisometrtropic amblyopia [9]. We report a case of unilateral DRS with keratoconus in contralateral eye. To our knowledge, no such association of DRS with keratoconus has been reported.

CASE REPORT

A 13 year old male child presented to the squint department with the chief complaint of outward deviation of left eye since birth which was constant and non-progressive. The parents also noticed that the child adopted a head turn to the right while viewing objects. He was the product of full term normal vaginal delivery with no prenatal and perinatal complications, born out of a non-consanguineous marriage with history of similar complaints in paternal grandfather.


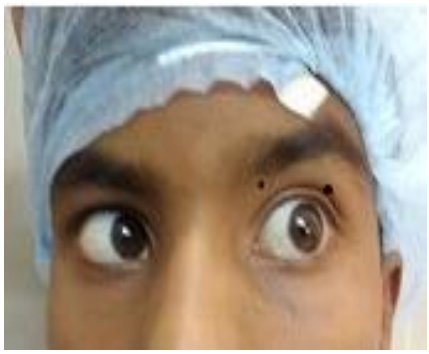
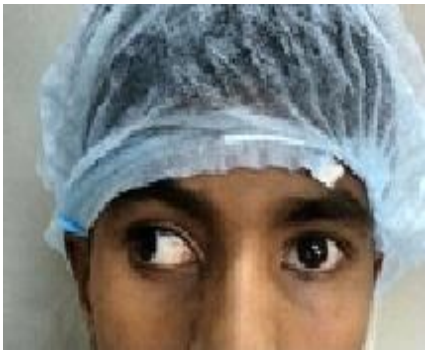

Figure 1 - Exotropic DRS, Figure 2 - Limitation of abduction, Figure 3 - Limitation of adduction

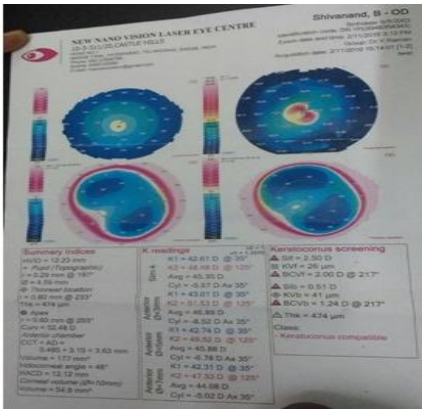
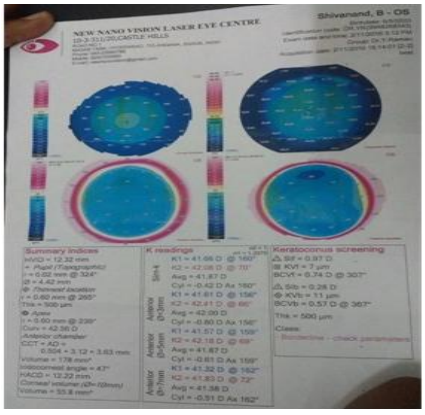


Figure 4 - Keratoconus in OD, Figure 5 - Normal OS, Figure 6 - Post operative day 1

The best corrected visual acuity was 6/12 in right eye and 6/6 in the left eye. The refraction in the right eye was -8.00*35 D cylinder and -0.25 sphere in the left eye. The anterior segment examination on slit lamp bio microscopy and fundus examination with 90D lens was normal. There was compensatory head turn to the right side by which he could maintain binocular single vision. Heischberg's corneal reflex test was central in right eye and 15 nasal papillary border in left eye (**Fig. 1**). The primary deviation measured was 30 prism diopters and secondary deviation 40 prism diopters. Ocular motility revealed limitation of abduction (**Fig. 2**) and adduction in left eye (**Fig. 3**) with minimal narrowing of palpebral fissure on adduction. Ocular movements in the right eye were normal.

Based on the cycloplegic refraction using short acting mydriatic eye drops which showed high cylindrical value (-8.00 D cyl at 35^0) in right eye, the auto refractometry values are closely correlating with retinoscopy findings, the child was subjected to corneal topography using New scheimpflug and placido-disk analyzer to rule out keratoconus. Corneal topography revealed findings suggestive of keratoconus in right eye (sim k1-42.61D at 35^0, K2-48.48D at 125^0, thickness-474 microns). It was normal in the left eye (sim k1-41.66D at 160^0, K2-42.08D at 70^0, thickness-500 microns).Above findings shown in (**Fig. 4**). The left eye having DRS, topography shows within normal limits (**Fig. 5**).

As the patient was not orthotropic in the primary gaze and had a compensatory head turn to the right, an 11 mm lateral rectus recession was done under general anaesthesia in the left eye. Informed consent was taken from the parents regarding the surgery as well as for clinical pictures, taken for the future scientific use. Subsequently, the patient was orthotropic in primary gaze and there was reduction in the compensatory head posture (**Fig. 6**). The patient is on regular follow up in view of keratoconus in the right eye.

DISCUSSION

The nuclei of 3^{rd}, 4^{th} and 6^{th} cranial nerves develop by the 4^{th} or 5^{th} week of gestation and their neural connection

with corresponding muscles is complete by 8th week. During the same period, there is differentiation of extra-ocular muscles from primitive mesodermal tissue.It is hypothesised that there is abducens hypoplasia with anomalous innervation of lateral rectus muscle from Oculo motor nuclei. Duanes retraction syndrome is associated 10-20 times more frequently with other congenital systemic anomalies.It is postulated that it is due to an insult during the early weeks (8-10wks) of pregnancy. It has been associated with congenital malformations of the musculoskeletal, cardiac, ocular, genitourinary, neural and auricular systems.

Shrestha et al [10] reported a case of Goldenhar syndrome with DRS in a 8month old child. Miller et al [11] presented the association of DRS with craniofacial malformations, Bosley et al [12] reported a case of bilateral DRS with Duchenne muscular dystrophy. Jethani et al reported a case of DRS with Bardet-Biedel syndrome in a 7yr old child.Kawano et al [13] reported a case of DRS with Morning glory syndrome in a 9yr old child. Mittal et al [14] presented a 7yr old girl with unilateral DRS with bilateral posterior microphthalmos. Kekunnyaya R et al found partial accommodative esotropia superimposed in a case of esotropic DRS with the help of cycloplegic refraction [15]

Keratoconus has a common association with atopy. It is also associated with Down's syndrome, Ehler-Danlos syndrome, connective tissue disorder, and mitral valve prolapse [9]**.** In our case, the child was diagnosed as a case of left sided DRS with keratoconus in contralateral eye. It was not associated with vernal kerato conjunctivitis. This case provides the importance of good cycloplegic refraction and clinical examination which guides towards correct diagnosis.Thus with cycloplegic refraction and confirmation with topography, the rare association of DRS and keratoconus could be diagnosed. The parents and child was not aware of deteriorating vision in the right eye.

CONCULSION

The present case report enlightens the need for cycloplegic refraction which helped us in comprehensive ophthalmic evaluation as well as recognition of a rare association of keratoconus, DRS and overall management of case.

references

1. DeRespinis PA, Caputo AR, Wagner RS, Guo S. Duane's retraction syndrome. Surv Ophthalmol. 1993; 38: 257–88.
2. Gurwood AS, Terrigno CA. Duane's retraction syndrome: Literature review. Optometry. 2000; 71: 722–6.
3. Duke-Elder S: System of Ophthalmology, vol 3, pt 2: Congenital Deformities. St Louis, Mosby–Year Book, 1963, p 991
4. Pfaffenbach DD, Cross HH, Kearns TP: Congenital anomalies in Duane's retraction syndrome. Arch Ophthalmol. 1972; 88: 635.
5. Pieroni D: Goldenhar's syndrome associated with bilateral Duane's retraction syndrome. Am J Ophthalmol. 1970; 70: 945.
6. Sharma DC, Parihar PS, Kumawat DC, et al: Duane's retraction syndrome with facial hemiatrophy (a case report). Postgrad Med J. 1990; 36: 51.
7. Miller BA, Pollard ZF. Duane's retraction syndrome and arthrogryposis multiplex congenita. Surv Ophthalmol. 1994; 38: 395.
8. Kirkham TH: Duane's syndrome and familial perceptive deafness. Br J Ophthalmol. 1969; 53: 335.
9. Rabinowitz YS. Keratoconus. Survey of Ophthalmology. 1998; 42(4): 297–319.
10. Shrestha UD, Adhikari S. Goldenhar syndrome in association with Duane syndrome. J Nepal Med Assoc. 2012; 52(185): 33-5.
11. Miller MT. Association of Duane retraction syndrome with craniofacial malformations. J Craniofac Genet Dev Biol. 1985; 1: 273-82.
12. Bosley TM, Salih MA, Alkhalidi H, et al. Duane retraction syndrome in a patient with Duchenne muscular dystrophy. J Ophthalmic Gen. 2016; 37 (3): 276-280.
13. Kawano K, Fujita S. Duane's retraction syndrome associated with morning glory syndrome. J Pediatr Ophthalmol Strabismus. 1981; 18(1): 51-4.
14. Mittal V, Gupta A, Sachdeva V, Kekunnaya R. Duane retraction syndrome with posterior microphthalmos: a rare association. J Pediatr Ophthalmol Strabismus. 2012;7; 49.
15. Kekunnaya R, Velez FG, Pineles SL. Outcomes in patients with esotropic Duane retraction syndrome and a partially accommodative component. Indian J Ophthalmol. 2013; 61: 701-704.

How to cite this article: Yenugandula R, Bharat RPK, Rare association of Keratoconus in the contralateral eye in a case of Duanes retraction syndrome. Indian J Case Reports. 2017; 3(2): 91-93.

Conflict of interest: None stated, Funding: Nil