

Case Report

Bilateral Type-I Duane's Retraction Syndrome with bilateral Crocodile Tears: A Case Report

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Abstract

Background: Duane's retraction Syndrome is a congenital form of strabismus characterized by horizontal eye movement limitation and globe retraction with palpebral fissure narrowing in attempted adduction. It may be associated with co-existing ocular and systemic pathologies. Crocodile tears, or a paradoxical gustatory lacrimal reflex can be either congenital or acquired. The congenital cases typically are associated with Duane's syndrome in most of the cases. **Case:** We present in our case report, uncommon case of bilateral Duane's syndrome with bilateral crocodile tears - both of which can be assumed to be of probably central origin. Though several cases have been reported, there are very few from the Indian subcontinent. Here, we also review the literature of the syndrome with crocodile tears. **Conclusion:** All cases of Duane's retraction syndrome warrant a thorough screening for coexisting ocular and systemic abnormalities. Ours is a classic report of bilateral Duane's syndrome with bilateral congenital crocodile tears, the coexistence of these two anomalies emphasising the aetiology to be anomalous innervation occurring at central level i.e. is brain stem

Key words: Duane's syndrome, congenital tears, paradoxical innervation, central origin

Case report

A 6 year old girl was brought to our ophthalmology clinic by her mother with history of watering from both the eyes while eating food. This was noted from a very early age. The mother also gave a history of eyes appearing small and some limitation of eye movement. The child was the only

offspring of her parents and was born out of non-consanguineous marriage. Antenatal and postnatal history were unremarkable. There was no family history of squinting and no associated systemic complaints.

On examination, the child had no anomalous head posture and no facial asymmetry. Anterior segment and dilated fundus examination of both eyes were normal. Her uncorrected visual acuity was 6/6 N6 in both eyes. She exhibited fusion which was examined using the Worth four dot test for distance. Her stereoacuity measured with Randot stereoacuity chart showed 50 arc/seconds. She was orthophoric for near and distance on alternate cover test examination (Figure 1).

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Figure 1: Orthophoric in primary gaze

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 left eye with minimal upshoot in left eye (Figure 2 and 3).

In order to assess specific complaints of mother, the child was asked to eat biscuits in the presence of the examining doctor and profuse lacrimation from both eyes was noted (Figure 4).

The lacrimal apparatus in both eyes was functionally and anatomically normal and a thorough systemic examination did not reveal any other anomalies. A conclusive diagnosis of bilateral Type-I Duane's Retraction Syndrome (DRS) with congenital aberrant tearing (crocodile tears) was made.



Figure 2: Limitation of abduction in right eye with widening of palpebral fissure and narrowing of palpebral fissure in left eye



Figure 3: Limitation of abduction in left eye with widening of palpebral fissure and narrowing of palpebral fissure with globe retraction right eye



Figure 4: Hyper lacrimation following ingestion of biscuits

Owing to the fact that patient had no anomalous head posture and globe retraction which was not cosmetically disfiguring, she was advised a regular follow up and no surgical intervention. The option of lacrimal gland botulinum toxin injection for the aberrant tearing was offered to the parents after due counselling, however, they refused any surgical intervention.

Discussion

DRS may be seen as an isolated finding or in association with both ocular and systemic anomalies (Sachsenweger et al,1966; Kirkham TH,1970). Numerous concomitant ocular and systemic anomalies have been reported with DRS. Some ophthalmic manifestations include iris dysplasia, ptosis, keratoconus, nystagmus,

optic disc anomalies, colobomata, congenital cataracts, microphthalmia, and Marcus-Gunn jaw winking syndrome (Gutowski NJ, 2000 ; Jafari AK et al ; 2010). Systemic anomalies include Goldenhar's syndrome, Klippel-Feil syndrome, Noonan's syndrome, Rubinstein-Taybi syndrome, spinal and rib malformations, sensory hearing loss, facial nerve palsy, genitourinary dysfunction, malformations of the limbs, and facial deformities (Gutowski NJ, 2000). The aetiology of DRS was thought to be replacement of lateral rectus muscle by fibrous tissue but recent studies point to co-contraction of medial and lateral rectus due to paradoxical innervation as to the main reason responsible for the features of this syndrome. The cause of this anomalous innervation is varied, but the evidence has pointed to a supranuclear lesion presumably in the brainstem (Pfaffenbach DD et al, 1972). Duane's syndrome warrants surgical management only if there's unacceptable head posture, large primary gaze squint, upshoots and downshoots and severe globe retraction which is cosmetically unacceptable being some of the primary indications.

Aberrant tearing or gustatory lacrimation (Crocodile tears) can be congenital or acquired. The congenital variant is very rare as compared to acquired type which is seen typically after facial nerve paresis secondary to lesions at or proximal to the geniculate ganglion. Congenital crocodile tears is usually associated with cases of Duane's retraction syndrome (Awan KJ, 1975). Various studies done till date have tried to determine the cause of simultaneous occurrence of these two anomalies of Duane's and crocodile tears. Many attempts have been made to abolish the annoying manifestations of this reflex. These include intraorbital injection of alcohol or cocaine to destroy postganglionic fibres of the sphenopalatine ganglion, subtotal resection of the lacrimal gland, sectioning of the glossopharyngeal and facial nerves at

different levels and the use of anticholinergic drugs (Axelsson A et al, 1962; McCoy FJ et al, 1979). Recent reports (Boroojerdi B et al, 1998; Hofmann RJ, 2000 ; Reimann R et al, 1999) suggest that injection of botulinum toxin type A (BTX-A) into the lacrimal gland of patients with CTS may effectively and safely abolish the reflex by chemodenervating the cholinergic neurons of the parasympathetic nervous system to the affected gland.

If we assume the presence of paradoxical innervation as the etiology of our retraction syndrome, a finding proved by electromyography in a similar case reported by Regenbogen and Stein, then we have the simultaneous presence of two syndromes of paradoxical innervation (Regenbogen T et al, 1968). This points to a central location as a possible anatomic site to explain both anomalies. Such a location in the pons was first suggested by Lutman (Lutman FC, 1947). The obvious explanation is an abnormality in the brain stem involving the embryonic development of the superior salivary nucleus and the supranuclear center for lateral gaze.

The other explanation of the lacrimal and eye movement disorders in Duane's syndrome would be a lesion causing a nuclear degeneration or dysgenesis in the immediate vicinity of the abducens nucleus, the paradoxical aspects of Duane's syndrome and the lacrimal disorder being the result of a substitute innervation of the lateral rectus by fibres from the oculomotor nerve and the lacrimal gland by fibres subserving salivation (Ramsay J et al, 1980).

To summarise, the rare association of two syndromes of paradoxical innervation, bilateral Duane's retraction syndrome and congenital crocodile tears, is presented here. This is probably due to a central anomaly at nuclear level in the pons. Very few cases have been reported so far from the Indian subcontinent.

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