Clinical Case Report

Duane retraction syndrome: A rare cause of strabismus

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ABSTRACT

Duane retraction syndrome (DRS) is a rare cause of strabismus in children. It has a characteristic ocular motility disorder and constitutes approximately 1% of all cases of strabismus. We describe a 9-year-old girl with absent or restricted horizontal eye movement, globe retraction and narrowing of the palpebral fissure on attempted adduction. DRS is classified into three types based on the different restrictions of ocular motility and can be associated with various ocular and systemic anomalies.

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INTRODUCTION

Duane retraction syndrome (DRS) is a congenital form of strabismus, first described by Sinclair, Turk and Stilling in 1895. In 1905, Duane described the various presentations and postulated a theory for its pathogenesis. We describe a patient with strabismus who was diagnosed to have DRS type I.

THE CASE

A 9-year-old girl was brought to our outpatients department with abnormal posture of the head while reading and writing, observed by her school teacher about a year ago. There was no history of diplopia, trauma or family history of similar complaints. The child was born as a full term normal delivery.

Evaluation by a paediatrician revealed a normal systemic examination with normal routine investigations. Ocular examination showed that unaided visual acuity in both eyes was 20/40. The best corrected visual acuity was 20/20 in both eyes. Her near vision was N6 and she did not have any amblyopia. Anterior segment examination was normal. On primary gaze, the left eye showed mild exotropia (Fig. 1). The left eye showed defective abduction, minimal defective adduction, narrowing of the palpebral fissure on attempted adduction along with globe retraction. She had a compensatory head turn to the left to maintain binocular vision. However, this adaptation was obvious only during reading and writing (near work). The Leash phenomenon, upshoot and downshoot were also observed (Fig. 1:1 and Fig. 1:3). MRI of the brain and orbit showed an absent left abducens nerve (Fig. 2).

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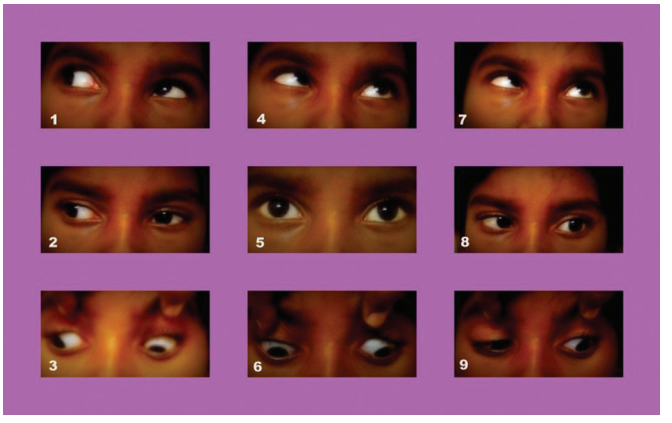

FIG 1. Ocular movements in different gazes. 1. Dextroelevation: overshoot of the left eye; 2. Dextroversion: narrowing of palpebral fissure with globe retraction and minimal restriction of adduction of the left eye; 3. Dextrodepression: downshoot of the left eye; 4. Sursumversion; 5. Primary gaze: mild exotropia of the left eye; 6. Deosursumversion; 7. Levoelevation; 8. Levoversion: restriction of abduction, lid retraction with widening of palpebral fissure in the left eye; 9. Levodepression

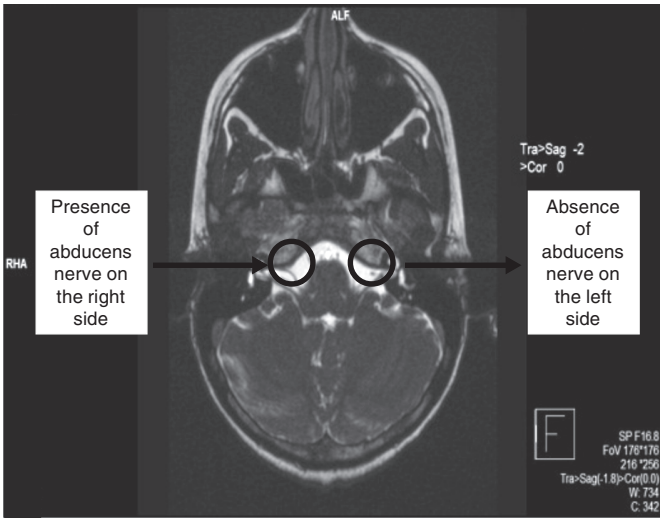

FIG 2. MRI scan showing absence of abducens nerve on the left side

Electromagnetic studies of the extraocular muscles were not done.

Based on Huber's classification of DRS[1–3] (Table I), the child was diagnosed as type I DRS. The associated ocular features of DRS[1,2] (Table II) were not seen. Systemic abnormalities known to be associated with DRS (Table II) were also not present.

There was minimal exotropia in primary gaze and the compensatory head posture was noticed only during near work, and did not cause any cosmetic problem. Appropriate spectacle correction was prescribed and she was asked to follow-up every 6 months.

TABLE I. Huber's classification of Duane retraction syndrome

Type I (70%)	Type II (7%)	Type III (15%)
Inability to abduct	Inability to adduct	Inability to abduct or adduct
Normal or minimal defect in adduction	Normal or minimal defect in abduction	—
Esotropia with straight head	Exotropia of the affected eye	Straight or nearly straight head position
A or V pattern	—	—
Usually upshoot or downshoot of the affected eye on adduction or attempted abduction	—	Possible upshoot and downshoot on adduction
Globe retraction and palpebral fissure narrowing on adduction	Globe retraction and palpebral fissure narrowing on adduction	Globe retraction and palpebral fissure narrowing on attempted adduction
Usually face turn to the affected side	Face turn to the normal side	Straight or nearly straight head position
Electromyography shows the absence of electrical activity in the lateral rectus muscle on abduction but paradoxical electrical activity on adduction	On electromyography the lateral rectus muscle shows peak impulse on abduction and a second paradoxical peak on attempted adduction. There is normal behaviour of the medial rectus	Electromyogram demonstrates co-contraction of the horizontal rectus muscles on both adduction and abduction

TABLE II. Ocular and systemic abnormalities seen in Duane retraction syndrome

Ocular abnormalities	Systemic abnormalities
Microphthalmos	Goldenhar syndrome
Ptosis	Klippel–Feil syndrome
Distichiasis	Sensory neural hearing loss
Crocodile tears	Foetal alcohol syndrome
Epibulbar dermoid	Oculocutaneous albinism
Nystagmus	Maternal thalidomide ingestion
Heterochromia	Holt Oram syndrome
Iris dysplasias	Wildervanck syndrome
Pupillary abnormalities	Facial hemiatrophy
Cataracts	Cervical spina bifida
Persistent hyaloid artery	Cleft palate
Optic nerve hypoplasia	Deformity of limbs and hands
Morning glory syndrome	
Marcus Gunn phenomenon	

DISCUSSION

DRS is known to affect young girls in their first decade of life. The left eye is involved more often than the right and there is a female preponderance.[1–6] Unilateral DRS is more common than bilateral DRS.[1,5] Most cases are sporadic but of familial occurrence, and autosomal dominant inheritance has been reported.

The prevalence of amblyopia in patients with DRS ranges from 3% to 48%. Our patient did not have amblyopia. Amblyopia, when present, is due to anisometropia rather than strabismus.[7] Esotropia is the most common strabismus reported with DRS type I, followed by exotropia and orthophoria.[1]

DRS is believed to be due to congenital absence or poor development of the abducens nucleus due to which the lateral rectus muscle receives abnormal innervation from branches of the oculomotor nerve.[1,2,4] This is likely to occur during the 4th to 8th week of intrauterine life during which the development of nerves and extraocular muscles occurs. This period coincides with other defects during embryogenesis which may present in DRS.

The characteristic presentation of DRS includes absent or restricted horizontal eye movement, globe retraction and narrowing of the palpebral fissure on attempted adduction.[1–4] Retraction of the globe occurs due to co-contraction of the lateral and medial rectus muscles,[1,4] which occurs due to misdirected nerve fibres from the oculomotor nerve to the lateral rectus muscle.

The Leash phenomenon, seen in our patient, has been explained on the basis of secondary fibrosis of the non-innervated fibres of the lateral rectus muscle. A part of the muscle is innervated by branches of the oculomotor nerve which also supplies the superior rectus muscle.[1]

Electromagnetic studies of the extraocular muscles are useful in detecting synergistic innervation of the medial rectus and the lateral rectus muscles.[2,6] These play a role in the classification of DRS.

Treatment options include correction of the refractive error, treatment of amblyopia and surgical correction. Surgical correction is required for patients with significant head turn, strabismus in primary gaze, and significant upshoot and downshoot on adduction.[2,4] Surgical treatment has its limitations as it does not assure complete clinical recovery.

The differential diagnosis of DRS includes[1,3,4] (i) sixth nerve palsy, (ii) congenital esotropia/familial esotropia, (iii) Moebius syndrome, and (iv) congenital oculomotor apraxia. Our case highlights the importance of MRI of the brain as an important diagnostic tool in correctly identifying the cause of strabismus in children.[8]

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