Inverse Duane's retraction syndrome – A case report

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Inverse Duane retraction syndrome (IDRS) is a rare and atypical form of DRS. Duane's retraction syndrome (DRS) is characterized by abnormal ocular movements such as pseudoptosis, globe retraction, and upshoots or downshoots on attempted adduction. Cases of IDRS are "inverse" in the sense that these abnormal movements are seen on attempted abduction than on adduction.

Most of the cases of IDRS are acquired with only four cases of presumed congenital etiology reported to date.[1-4] All reported cases have been characterized by globe retraction with narrowing of palpebral fissure on attempted abduction, but with a variable amount of abduction limitation, inverse of type I DRS.

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A 13-year-old boy presented to us with narrowing the left eye when looking toward the left, noticed in the last few years. There was no history of previous trauma or ocular surgery.

On examination, BCVA was 6\6; N6, in both eyes. Stereopsis of 40 arc-seconds on Titmus fly test was noted in compensated head posture. He had a left face turn of 15°.

Alternate Prism cover tests showed exophoria in the primary position which increased to 12 PD exotropia with 7-PD hypertropia in left gaze and orthophoria in right gaze. He had exophoria at near as well. The compensatory head posture was constant and not just during inattentiveness.

Ocular movements showed limitation on adduction of − 2 grade in the right eye, while the left eye was normal. Narrowing of the palpebral fissure was noted (palpebral fissure height: 9.5 mm on abduction and 13.5 mm on adduction) along with globe retraction in the right eye [Fig. 1]. Anterior and posterior segment examination was within normal limits. Cycloplegic refraction revealed +0.50 DS in right eye and +0.50 DS with −0.50 DC in left eye.

A clinical diagnosis of right IDRS of possible congenital innervational etiology was made.

MRI of the brain and orbit was ordered in view of the short history and to rule out possible acquired causes, which was reported to be normal. He was offered strabismus surgery (lateral rectus Faden/Scotts procedure) and is yet to report for the same.

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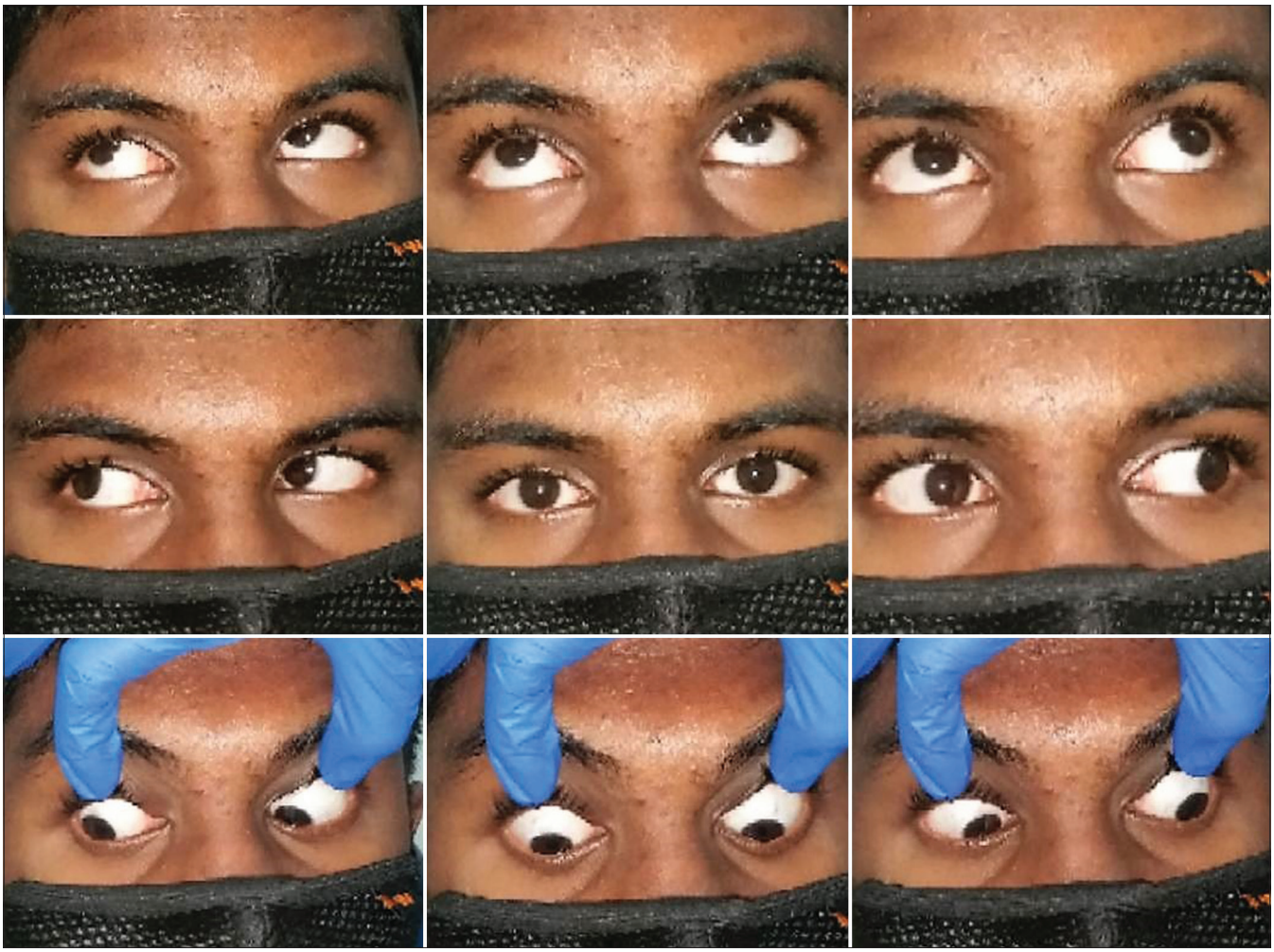

Figure 1: Nine gaze photo of a case showing limitation of adduction in the right eye with palpebral fissure widening in adduction and narrowing on abduction

Discussion

DRS is an innervational abnormality of the lateral and medial rectus muscle, which causes variable limitation of abduction and adduction, respectively. It is associated with globe retraction with narrowing of the palpebral fissure on attempted adduction. DRS type I are cases with predominant abduction limitation, type II are cases with predominant adduction limitation, and type III are cases with both components of movement restriction.[5] Cases of IDRS have a distinct etiology and pathogenesis ranging from medial rectus abnormality, innervational misdirection, or secondary to restriction from trauma and surgery.[1,5-8]

They are "inverse" in the sense that abnormal movements of globe retraction and palpebral fissure narrowing occur on abduction and not adduction as seen in classical DRS.[6]

The first case series describing IDRS was reported in 1977 by Duane *et al*.[6] and were termed as pseudo DRS. They described five cases of medial rectus entrapment resulting in an abduction limitation, and associated globe retraction, narrowing of the palpebral fissure, and pseudoptosis on attempted abduction were reported. All five cases had a medial rectus involvement, four of which were due to previous history of trauma, and one case was secondary to orbital metastasis from primary breast carcinoma. Similar presentations due to myocysticercosis[7] and restrictive effect due to recurrent pterygium[8] causing medial rectus involvement have been reported. All these cases were of acquired etiology and showed limitation of abduction.

Only four cases of IRDS of congenital etiology have been reported to date. Chatterjee *et al*.[1] first described a case with left-sided deafness and bilateral abduction limitation, alternating esotropia with globe retraction, and narrowing of palpebral fissure on attempted abduction bilaterally. Intraoperatively, extensive fibrous bands surrounding the medial rectus extending to the medial orbital wall were noted. Subsequently, Lew *et al*.[3] reported another case of inverse retraction syndrome in a 6-year-old girl with contracture of the medial rectus muscle noted intraoperatively. A medial rectus tenotomy was performed instead of the originally planned medial rectus recession. Another case of bilateral inverse globe retraction syndrome reported by Khan A.O. in 2007[2] presented with an exotropia and hypertropia in the left eye, but there was a prior history of esotropia at birth which was corrected with bilateral medial rectus recession and left-eye lateral rectus resection. The most recent report of this condition is by Agarkar *et al*.[4] in 2017 with left esotropia of 10 PD and left face turn.

All four cases showed narrowing of palpebral aperture and globe retraction on attempted abduction. The extraocular motility limitation was a limitation of abduction similar to the extraocular motility restriction seen in type I DRS.

Our case presented with features of IDRS with limitations of adduction and normal abduction. Among the spectrum of cases reported on IDRS, this is the first case showing extraocular movement restriction inverse of type II DRS. Our case being of congenital etiology and with features of IRDS except normal abduction makes it a rare presentation of inverse DRS.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Chatterjee PK, Bhunia J, Bhattacharyya I. Bilateral inverse Duane's retraction syndrome--A case report. Indian J Ophthalmol 1991;39:183-5.
2. Khan AO. Bilateral inverse globe retraction (Duane's) syndrome. Indian J Ophthalmol 2007;55:388-9.
3. Lew H, Lee JB, Kim HS, Han SH. A case of congenital inverse Duane's retraction syndrome. Yonsei Med J 2000;41:155-8.
4. Agarkar S, Kaduskar A. Congenital inverse Duane's retraction syndrome: A rare presentation. Indian J Ophthalmol 2017;65:422-3.
5. DeRespinis PA, Caputo AR, Wagner RS, Guo S. Duane's retraction syndrome. Surv Ophthalmol 1993;38:257-88. Erratum in: Surv Ophthalmol 1996;40:423.
6. Duane TD, Schatz NJ, Caputo AR. Pseudo-Duane's retraction syndrome. Trans Am Ophthalmol Soc 1976;74:122-32.
7. Murthy R. Inverse Duane's retraction syndrome following myocysticercosis. Indian J Ophthalmol 2008;56:89-90.
8. Khan AO. Inverse globe retraction syndrome complicating recurrent pterygium. Br J Ophthalmol 2005;89:640-1.