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Case Report

Type III Duane's Retraction Syndrome with Severe Upshoot: a Rare Presentation with Ipsilateral Superior Rectus Contracture

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Abstract: Type III Duane's retraction syndrome (DRS) is a rare condition and represents only 1% of all cases of Duane's syndrome. It consists of limited/absent abduction as well as adduction of the affected eye. A characteristic upshoot/downshoot/both, in adduction, and globe retraction may occur in DRS patients. Secondary muscle changes like medial rectus contracture or superior rectus contracture may rarely be associated with DRS. Superior rectus contracture syndromehas been reported in patients with unilateral superior oblique palsy and dissociated vertical deviation(DVD).However, no case report of association of SRC with DRS has been reported. We report a rare case of type III DRS with severe upshoot and ipsilateral superior rectus contracture. It is hypothesized that a long standing severe upshoot could be the reason for development of secondary superior rectus contracture. The upshoot was treated with Y-split of lateral rectus and hypertropia was corrected by superior rectus recession.
Keywords: Superior rectus contracture, Duane's retraction syndrome, Upshoot

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INTRODUCTION

Duane'sretraction syndrome (DRS) is an unusual congenitalform of strabismus characterized by limitation ofhorizontal eye movements and globe retraction with palpebral fissure narrowing on attempted adduction [1]. Type III DRSis rare; representing only 1% of all cases of DRS and consists of limited/absent abduction with variable limitation of adduction. A characteristic upshoot / downshoot or both may occur in adduction**.** Secondary muscle changes like medial rectus or superior rectus contracture may rarely be associated with DRS [2].

Superior rectus contracture syndrome (SRC) has been described in patients with unilateral superior oblique palsy.

Its' association with dissociated vertical deviation (DVD) has been reported [3], but no study in literature has reported superior rectus contracture in DRS. We report a rare case of type III DRS with superior rectus contracture.

CASE REPORT

A 23 year old male presented with the deviation of left eye since birth. There was history of adopting an abnormal head posture while concentrating over distant objects. There were no preceding illnesses and the patient's birth history, family history and past medical history were non-contributory. There was no history suggestive of any surgical intervention in the past.

On examination, his best corrected visual acuity was 20/20 in both the eyes with no significant refractive error, with a near vision of N6 in both eyes. He preferred his right eye to fixate and adopted a face turn to right. Ocular motility testing was notable for limitation of adduction (-2) as well as abduction (-1) in the left eye with severe upshoot of the left eye in adduction, consistent with the clinical picture of type III DRS in left eye.Prism alternate cover test showed incomitant lefthypertropia (LHT)in various gaze positions. LHT measured40 PD with 25 PD exotropia (XT) in the primary position, 20 PD LHT with 30 PD XT in right gaze and increased to 45 PD with no horizontal deviationon the left gaze (Figure 1). His LHT was worse on left head tilt (45 PD) than on right head tilt (30 PD) (Table1). He had evidence of fusion with Worth 4 dot test in left gaze position (habitual right face turn).However, in other gaze positions, left eye suppression was noted. Stereopsis was not detected with Titmus fly and Wirt circles in any gaze position.

Table 1: Measurements of binocular alignment (PD) in various gaze positions and head tilts with right eye fixation

Gaze direction	Primary position	Dextroversion	Levoversion	Right tilt	Left tilt
Before surgery	LHT 40, XT 25	LHT 20, XT 35	LHT 45	LHT 30 ,XT 25	LHT 45
After surgery (3 months)	LHT 14	LHT 16	LHT 16	LHT 14	LHT 14

LHT: Left hypertropia, XT: Exotropia

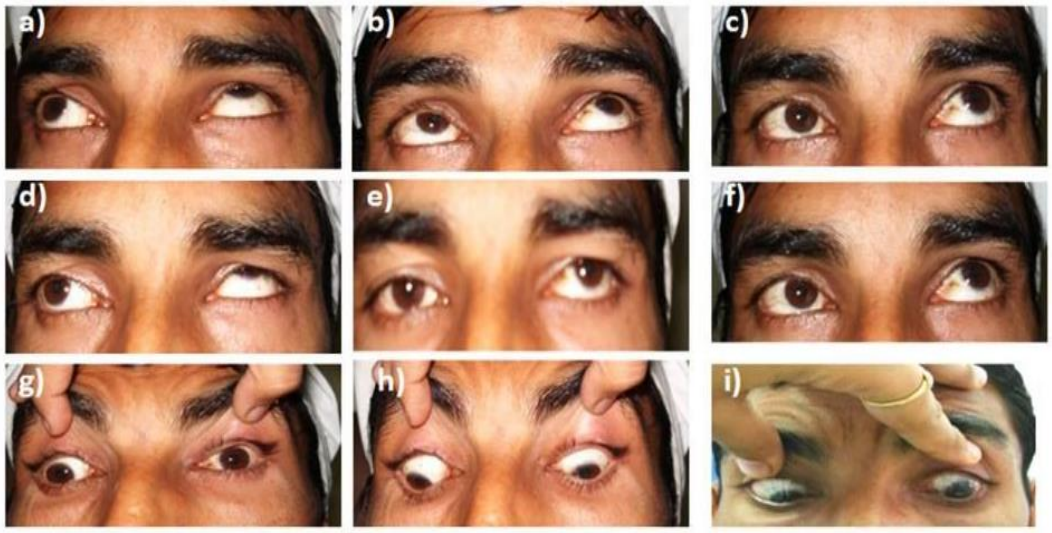

Fig. 1: Preoperative (a-i): Ocular alignment in nine cardinal gaze postions.In primary gaze, left eye showed hypertropia and exotropia (e), which increased in left gaze (f). When the left eye was adducted (d), the left eye abruptly moved upwards, crossed the horizontal line, and the cornea disappeared under the upper lid (severe left eye upshoot).

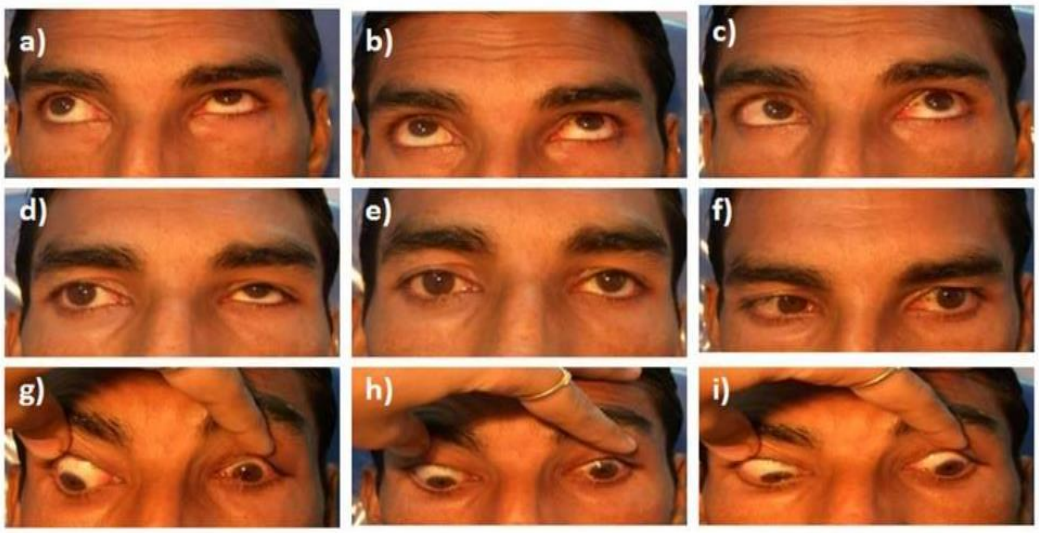

Fig. 2: Postoperative (3months)(a-i): Ocular alignment in nine cardinal gaze postions. Horizontal realignment was complete with marked reduction of left hypertropiain primary position (e), and similar comitant deviation in other gazes (d,f). The upshoot in adduction was markedly reduced (d).

Forced duction test was performed which revealed a tight superior rectus of the left eye but was negative for right eye in all directions. Fundus evaluation was within normal limits. To summarize, the presence of large left hypertropia in primary position which increased on ipsilateral gaze and ipsilateral head tilt, suggested the possibility of superior rectus contracture, which was further confirmed by a positive forced duction test. The presence of upshoot, palpebral fissure changes and limited ocular movements confirmed the diagnosis of Type III DRS.

The patient underwent surgery in the form of both eyes lateral rectus recession of 6mm with Y- split of left lateral rectus and left superior rectus recession of 6mm. Postoperatively, the face turn improved significantly, horizontal realignment was complete with marked reduction of left hypertropia in primary position and with similar comitant deviation in all other gazes. The upshoot in adduction was markedly reduced (Table 1, Figure 2). Fusion was noted in primary position. In his latest follow up 1 year postoperatively, there was no evidence of abnormal head posturing and the alignment was stable and cosmetically acceptable.

DISCUSSION

Superior rectus contracture syndrome was first reported in 1964 in patients with unilateral superior

oblique palsy[3]. SRCshould be suspected when there is hypertropia of > 15 PD in primary position which increases in ipsilateral version (difference > 5PD) and on head tilt to the affected side [4]. Tight superior rectus muscle on forced duction test is another clue towards the diagnosis [4, 5]. Jampolsky [6], however suggested that this syndrome is not seen exclusively in superior oblique palsy but also in several other entities such as dissociated vertical deviation, thyroid myopathy, orbital floor fracture, sensory exotropia, and monocular elevation deficit

An upshoot/downshoot is reported to be more frequent in type III DRS [7]. An upshoot may occur due to co-innervation of superior rectus muscle with the lateral rectus [8]. So, it can be hypothesized that a chronically innervated superior rectus muscle may result in contracture in patients with long standing upshoot, and thus seems plausible that upshoot could cause superior rectus contracture. Khawam et al explained Jampolsky's findings, that because the fixing (non-Duane's) eye constantly works against the imbalance of innervational forces in the Duane's eye, contracture of the fellow eye (Duane's eye) yoke muscle may result [3]. So, in our patient the superior rectus contracture could have been the result of long-standing upshoot caused by chronically innervated superior rectus muscle.

Y-split of the lateral rectus has been accepted as a standard procedure for management of upshoot and downshoot in DRS [9]. The management of superior rectus contracture includes large recession of the superior rectus muscle. Our patient had type III Duane's retraction syndrome with severe upshoot and superior rectus contracture. The condition was successfully managed by superior rectus recession along with lateral rectus recession and Y-split surgery.

In conclusion, superior rectus contracture can occur in association with Duane's retraction syndrome, due to long standing upshoot. The clinicians should be aware of the possible combination and plan their management accordingly.

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