Type II Duane’s retraction syndrome with severe upshoot with ipsilateral superior oblique muscle palsy: a rare presentation

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Abstract
Type II Duane’s retraction syndrome (DRS) is the least common strabismus characterized by limitation of adduction with the presence of upshoot, downshoot or both. Abduction can be normal or slightly altered. Secondary muscle changes like fibrosis, and anomalous insertions have been associated with DRS. Its associations with dissociated vertical deviations have been reported. We report a rare case of DRS with superior oblique palsy presenting post trauma in a tertiary care centre.

Introduction
Duane's retraction syndrome (DRS) is an unusual form of strabismus characterized by limitation of horizontal movements and globe retraction with palpebral fissure narrowing on attempted adduction of the affected eye. In general, 1-4% of strabismic patients have Duane’s syndrome. Type II DRS is least common and presents as limitation of adduction with exotropia of the affected eye. Abduction can be normal or slightly affected. A characteristic upshoot, downshoot or both may occur in adduction.

Its association with dissociated vertical deviation has been reported previously. Superior rectus overaction/contracture syndrome (SRSy) was described by Jampolsky in 1964. Superior rectus contracture has been previously reported in a patient with unilateral superior oblique palsy (SO). Here, we report a rare case of type II DRS with ipsilateral SO.

Case Report
A 50-year-old male presented with complaints of blurring of vision and double vision in downgaze following head trauma 6 days back, which was associated with black eye. There were no preceding systemic illnesses, and patient’s birth history, family history and medical history were not significant. There was no history of surgical intervention in the past. Previous CT scan showed thickening of right superior oblique muscle.

On examination, best corrected visual acuity was 6/6 with no significant refractive error. The patient had small left head tilt with left face turn. Ocular motility showed global limitation in adduction with upshoot on adduction with palpebral fissure narrowing in the right eye. The alternate prism cover test showed 20PD right hypertropia (RHT) with 6PD exotropia in primary gaze and 12 PD RHT with 6PD exotropia in downgaze. Deviation of near showed 14RHT with 6PD exotropia. Rest movements in both eyes were full. The patient had diplopia in a distance Worth 4-dot test. Double Maddox rod showed 10° exotorsion in primary gaze and 15° in downgaze (Figs 1 and 2). Diplopia and torsion were noted in diplopia charting and superior oblique underaction on Hess charting. Fundus examination showed mild exotorsion. Review of the old photographs prior to trauma did not reveal any face turn. There was a diagnostic dilemma regarding the co-existence of SO with DRS. Although right hypertropia pointed towards the severe upshoot in adduction, the appearance of left face turn after trauma, and

Figure 1: Left face turn.

Figure 2: Extraocular movements in all gazes showing global limitation in adduction with upshoot on adduction with palpebral fissure narrowing in the right eye.
large vertical and torsional diplopia in the field of action of superior oblique muscle and underaction of the same in Hess charting, suggested the presence of right SO. The presence of exotropia, adduction limitation with upshoot and palpebral fissure changes, on the other hand, confirmed diagnosis of Type II DRS. Subsequently, surgery was planned to correct his face turn and vertical squint. On table, inferior oblique muscle was found to have an abnormal insertion. Forced traction test showed laxity in the right superior oblique muscle. A superior oblique tuck of 6 mm with inferior oblique recession in the right eye was performed. A repeat traction test at the conclusion showed no evidence of iatrogenic Brown’s. On the first postoperative day, his face turn and small head tilt improved significantly with collapse of the vertical squint to a large extent. He was kept under close follow-up and the next plan of action is to take care of the exotropia and the residual upshoot by lateral rectus recession with or without a Y-split (Figs 3 and 4).

**Discussion**

Heuck described patients with severe limitation of ocular motility and retraction of the globe in nineteenth century. Type II DRS is the least common type (<10%) characterized by a marked limitation of adduction with exotropia of the affected eye, abduction normal or slightly limited, retraction of the globe and narrowing of the fissure on attempted adduction. On electromyography, the lateral rectus showed peak impulses on abduction and a second paradoxical peak on attempted adduction. There was normal behaviour of the medial rectus.

**Figure 3:** Diplopia chart showing increased separation of the vertical images in the field of action of superior oblique muscle and the presence of torsional diplopia more in downgaze.

**Figure 4:** HESS chart showing severe underaction of superior oblique muscle and mild overaction of inferior oblique muscle in the right eye and overaction of superior oblique and inferior rectus along with underaction of superior rectus in the left eye.
The pathogenesis of DRS has a wide spectrum, which can be classified into mechanical, anatomical and innervational disorder. Anatomical changes most commonly seen are fibrotic changes and anomalous insertions.7 Mechanical disturbances can occur due to the presence of facial bands found in some cases of DRS. These band causes the limitation of eye movement.

An upshoot may occur due to co-innervation of superior rectus muscle with the lateral rectus or could be because of the mechanical factors as the bridle or leash effect due to tight lateral rectus.3

The innervational type of upshoot is characterized by the presence of hypertropia in the primary position.8 Another characteristic feature is that, in innervational type, there is a gradually increasing upshoot of the eye as it moves in adduction. Our case had the obvious features of exotropic DRS with a limitation of adduction, upshoot and palpebral fissure narrowing in adduction.

In our case, the exact pathogenesis of SO is not known. The laxity of the superior oblique tendon on forced traction testing hint towards a possibility of congenital SO. But the absence of significant head tilt even prior to trauma and sudden onset vertical and torsional diplopia in the field of action of superior oblique muscle following trauma points more towards an acquired SO with DRS.

**Conclusion**

DRS and SO have been shown to co-exist in our case although the exact pathogenesis still remains unraveled. Identification of each of these components requires a meticulous history taking, review of old photographs and thorough clinical examination and supportive investigations. The plan of surgery in such cases is a staged procedure: first, correcting the SO by superior oblique tucking with inferior oblique recession and, second, DRS surgery, as recession of lateral rectus muscle on the involved side in proportion to the size of exotropia9 with or without Y split for upshoot.

**References**


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