Congenital inverse Duane's retraction syndrome: A rare presentation

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A 12-year-old girl presented with esotropia and face turn since birth. Ocular motility examination showed restricted abduction associated with down shoot and retraction on attempted abduction characteristic of inverse Duane’s retraction syndrome. To the best of our knowledge, this is one of the very few reported cases of congenital inverse Duane’s retraction syndrome.

Key words: Abnormal head posture, congenital inverse Duane’s retraction syndrome, strabismus

Inverse Duane’s retraction syndrome is a rare condition characterized by limited abduction which may be accompanied by retraction of the eyeball, narrowing of the palpebral fissure, and pseudoptosis. This was first reported by Duane et al. in 1976 as a series of five patients. Here, we report a case of a 12-year-old girl with classical features of inverse Duane’s retraction syndrome.

Case Report

A 12-year-old girl presented to a tertiary referral center with left esotropia noticed since birth. There was no history of trauma or any previous surgery. There was no family history of strabismus. There was no history of diplopia on examination; she adopted a left face turn of 15° while fixing. There was no facial asymmetry or any facial abnormality [Fig. 1]. Visual acuity in both the eyes was 6/6 (20/20), N6. The cover test showed left esotropia of ten prism diopters for both distance and near in primary gaze with corrected head posture. Ocular movements were full in the right eye while in the left eye, there was a limitation of abduction beyond midline. In addition, she also had significant down shoot and globe retraction on attempted abduction. There was narrowing of the palpebral...
Limited abduction and globe retraction on attempted abduction of inverse globe retraction syndrome due to the restrictive pathology rather than the innervational abnormality. Interestingly, Khan, in another report, describe a case of bilateral inverse globe retraction syndrome due to abnormal innervation; however, this case had previous bilateral medial rectus recession surgery. It is difficult to comment if previous surgical scarring in anyway contributed to inverse Duane’s retraction syndrome. We considered oculomotor nerve paresis with aberrant regeneration as a possible differential diagnosis, but the patient neither had ptosis nor any pupillary changes suggestive of the same. Even though our patient refused imaging, it would have been interesting to look for any abnormal innervation or any localized medial rectus pathology which could explain her spectrum of motility abnormalities. To the best of our knowledge, this is one of the very few reported cases of congenital inverse Duane’s retraction syndrome.

Discussion

Inverse Duane’s retraction syndrome is a rare musculofacial anomaly. The etiology of the syndrome is complex, and there is a paucity of information in the literature on this clinical entity. Duane et al. in 1976 first reported five cases of inverse Duane’s retraction syndrome. In his series, four cases were following trauma, and one patient had orbital metastasis. The cause of inverse Duane’s retraction syndrome has been identified to be an agenesis of the sixth nerve and nucleus, with the inferior division of the oculomotor nerve (nerve to the medial rectus muscle) splitting to innervate the medial and lateral rectus muscles.

Inverse Duane’s retraction syndrome is a rare condition characterized by limited abduction. Motility abnormalities in inverse Duane’s retraction syndrome are thought to be driven by abnormal medial rectus muscle as opposed to lateral rectus muscle in Duane’s retraction syndrome. The basic pathology of palpebral fissure narrowing in Duane’s retraction syndrome is aberrant innervation, while in inverse Duane’s retraction syndrome, it is postulated as restrictive in nature. Literature search in English language publications for inverse Duane’s retraction syndrome reveals only 2 reports of congenital inverse Duane’s retraction syndrome. Chatterjee et al. have reported a case of bilateral inverse Duane’s retraction syndrome with perceptive deafness. Lew et al. have reported a case of congenital inverse Duane’s retraction syndrome with medial rectus shortening. However, our patient did not have any history of deafness. Among other reports of acquired inverse Duane retraction syndrome, Khan has reported a case of inverse globe retraction syndrome due to the restrictive effect of a recurrent pterygium. There is also a report of inverse Duane’s retraction syndrome due to myocysticercosis involving the medial rectus muscle. Psuedo Duane retraction syndrome has been reported secondary to orbital trauma due to entrapment of the medial rectus muscle. All these reports, indicate restrictive pathology rather than the innervational abnormality.

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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Conflicts of interest

There are no conflicts of interest.

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