Isolated Unilateral Congenital Sixth Nerve Palsy A Case Report and Review of Literature

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Introduction

A congenital sixth nerve palsy is rare and may be related to birth trauma. The deficit often is transient, has a very good prognosis and usually resolves in the first month of life. The title of a Souza-Dias publication stated:"Congenital VIth nerve is Duane's Syndrome until disproven", and it also reflects the rarity of congenital sixth nerve paresis. A long standing VIth nerve palsy leads to secondary contracture of Medial Rectus (MR). The tight MR would cause retractionof the globe on adduction with consequent narrowing of palpebral fissure (PF), adding complexities to the clinical findings by mimicking Duane retraction syndrome (DRS) type I. Very few cases of long standing congenital VIthNerve palsy have been reported in the literature. We report a case of unilateral congenital VIth nerve palsy presenting to us with face turn and esotropia(ET)successfully managed by medial rectus(MR)recession.

Case summary

A 3-year-old girl presented to us with face turn to right side since birth. Ante, peri and postnatal history werenormal. There was no history of trauma or prior treatment forthe strabismus.Family album tomography (FAT) showed right esotropia (RET). The general and systemic examinations were essentiallynormal. The child was not cooperative for the vision assessment. Refraction under cylcoplegia revealed no significant refractive error. No abnormality detected in the fundus evaluation. Ocular examination revealed orthophoria with face turn to right side. On eliminating the head posture the Prism Bar Cover Test (PBCT) revealed esodeviation of right eye(RE) of 15 prism dioptre(PD). Fixing with RE (FRE) the esodeviation was more (20PD) suggesting an incomitant deviation.There wasmarked limitation of abductionof RE with widening of the PF onabductionwith mild limitation in dextro-elevation and dextro-depression [Figure 1]. Binocular functions on Worth Four Dot and RandotStereoacuity Tests could not be assessed.Forced duction testing revealed a tight medial rectusRE.A high resolution T1-weighted MRI (including fast imaging enhancing state acquisition [FIESTA]) at the level of

brain stem showed intact abducens nerve. The patient underwent 5 mm recession of right medial rectus(MR) by

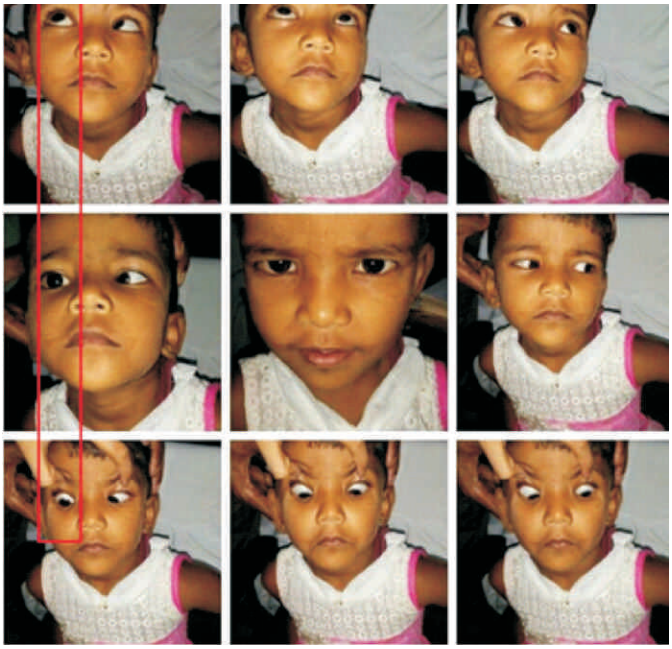

Figure 1: *Preoperative 9 gaze photographs showing marked limitation of abduction of RE with widening of the PF on abduction with mild limitation in dextro-elevation and dextro-depression.*

conjunctival limbal approach and standard technique. The patient was followed up onday 1, 7, 14 and 60 postoperatively. Postoperative examinationsrevealed orthophoria with elimination of head posture. Ocular movementsshowed improvement in abduction with reduction in abnormality in PF size in different gazes [Figures 2].A high resolution T1-weighted MRI (including fast imaging enhancing state acquisition [FIESTA]) at the level of brain stem showed intact abducens nerve.

Discussion

A congenital sixth nerve palsy is very rare and may be related to birth trauma. The deficit often is transient, and usually resolves in the first month of life. VIth nerve palsy occurs more commonly in children than in adults. The leading causes are neoplasms (range: 27–39%) such as meningioma, acoustic neurinoma, nasopharyngeal carcinoma and head injuries (range: 34–42%). , , , Less common causes are idiopathic, congenital, hydrocephalus, infections (herpes virus family, leptospirosis), otitis media, and others.7,8,

An esotropic DRS is more common than a congenital sixth nerve palsy. The clinical findings of head posture, ET in

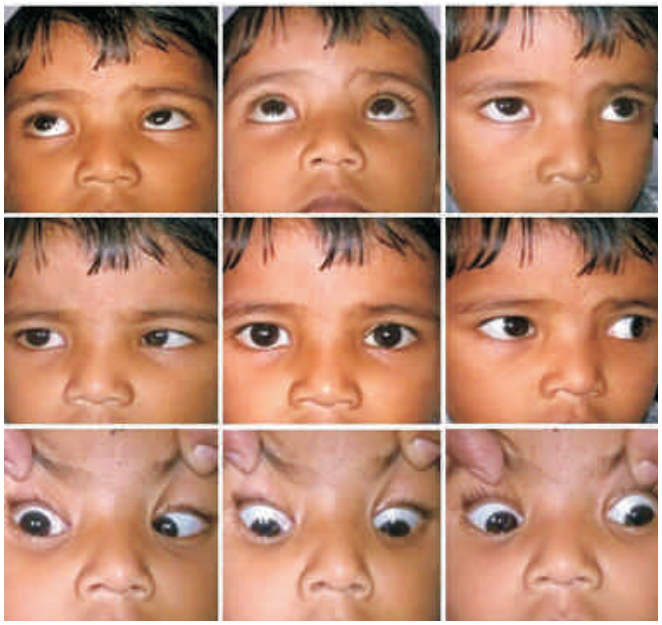

Figure 2: *Postoperative 9 gaze photographs showing orthophoria with elimination of head posture. Ocular movements showed improvement in abduction of RE with reduction in abnormality in PF size in different gazes*

primary position on elimination of head posture with restrictedabduction and associated narrowing of PF in adduction in our patient are consistent with Type I DRS. The primary position ET was relatively small as in DRS (less than 30 PD) compared to LR palsy or paresis. Although widening of PF in abduction is typical of DRS, PF narrowing is not a very dependable diagnostic sign of mild to moderate DRSas narrowing of the PF on adduction is usually interpreted as a passive adjustment of the lids to retracting globe. Our patient was diagnosed as a case of congenital sixth nerve palsy, with forced duction test for MR and absence of 'X' pattern seen due to anomalous LR innervations in upgaze and downgaze in DRS, pointing towards VIth nerve palsy.Besides,presence of abducens nerve innervationtothe affected LR on the MRI also rules out the pathogenesis of type I DRS as studies have shown that abducens nerve is frequently absent in type I DRS. , MR in children with DRS does not exhibit excessive stiffness or contracture in the primary zone, that is, it is normal. The alteration in PF size can be explained by a secondary contracture of right MR muscle due to a longstanding LR palsy, causing retraction of the globe on adduction with consequent narrowing of PF.4

On comparing the clinical features and postoperative results with a similar case reported in the literature by Agrawal et al.4, we found that our case presented with compensatory head posture to right with small ETof 15 PD in contrast to the previously reported case with 70 PD RET with no CHP anddemonstrated no binocular functions on Worth Four Dot and Randot Stereo acuity Tests. Though we could not elicit the binocular function in our case, the CHP proves that the patient had BSV. 5 mm of conventional recession gave satisfactory results in our case with orthotropia in primary position and elimination of CHP. Also there was no limitation of adduction postoperatively in contrast to the previous reported case who developed mild adduction deficit after the surgery. The probable differences in the clinical finding and postoperative results might be because of the difference in age of presentation in both the cases which was 3 years in our's compared to 14 years of previously reported. The MR fibrosis becomes denser with time without any intervention, leading to retraction of the globe on adduction with consequent narrowing of PF and upshoot. Besides, it also decreases the predictability of the results achieved by conventional MR recession.

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