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A Rare Case of Bilateral Duane's Retraction Syndrome with Crocodile Tears, Hearing Loss and Klippel Feil Syndrome (Wildervanck Syndrome)

Case Report

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Abstract

The Duane syndrome is a strabismus syndrome which is characterized by congenital non-progressive horizontal ophthalmoplegia which primarily affects the abducens nerve. Approximately 70% of the individuals with the Duane syndrome have an isolated disease and 30% of cases are associated with other congenital anomalies. A triad of Klippel–Feil anomaly Duane retraction syndrome, and hearing deficits is Wildervanck syndrome the estimated the prevalence is less than 1/1,000,000. We are presenting here, 26 year old male with a very rare case of bilateral Duane syndrome with hearing loss and klippel feil anomaly possible Wildervanck syndrome along with addition feature of crocodile tears.

Keywords: Case report; Duane's retraction syndrome; Klippelfeil syndrome; Wildervanck syndrome

Introduction

Duane Retraction Syndrome, is a congenital and non-progressive strabismus syndrome characterized by complete or less often partial absence of abduction, retraction of globe on adduction and narrowing of palpebral fissure during adduction (induced ptosis) [1]. Advanced neuroimaging, muscle electrophysiology and genetic analysis gave better understanding of this form of strabismus, now considered a congenital cranial dysinnervation disorder (CCDD), giving better insights into the management of this challenging syndrome. This may be primary due to absence of normal innervation or secondary following aberrant innervations from other cranial nerves. CCDD is a non-progressive entity and may also have associated bony abnormalities [2].

Types of DRS (Hubers) [3]

Type 1 (70%–80%): Marked limitation of abduction with minimally defective or normal adduction, globe retraction and palpebral fissure narrowing in adduction, widening in abduction.

Type 2 (7%): Marked limitation of adduction with primary position exotropia of the affected eye abduction normal or slightly limited with globe retraction and palpebral fissure narrowing in attempted adduction.

Type 3 (15%): Limitation or complete absence of adduction and abduction with globe retraction and palpebral fissure narrowing in attempted adduction.

Case Presentation

A 26-year-old male with history of horizontal eyeball movement limitation of the both eyes since birth, profuse lacrimation of both eyes when eating food since childhood. His birth, developmental and family history were normal. On examination his best corrected visual acuity was found to be 20/20 in both eyes. He had -3 abduction deficit in both eyes and retraction of the globe and fissure-narrowing on adduction in the both eyes. He also had short neck (height neck ratio 14), bilateral high frequency hearing loss without anatomical malformation. Other neurological examinations were normal.MRI brain shows bilateral abducens nerve absent and smaller volume of the bilateral lateral rectus muscles. CT spine shows partial fusion of posterior vertebral body C5,C6 and blocked vertebra between D3,D4 with defect in posterior vertebral body (klippelfeil type 3) (Figure 1 and 2).

Discussion

Myofibers of extraocular muscles (EOM) are developed by a condensation of the mesoderm around the eye. When the embryo is 7 mm long, the EOMs form one mass supplied by the oculomotor nerve. When the embryo is 8–12 mm long, this mass divides into separate muscles. It is at this stage that the fourth and sixth nerves arrive. Due to disturbing influences of unknown origin, the abducens nerve fails to develop, causing branches of the oculomotor nerve to remain in contact with the muscle mass that would later become the lateral rectus [4]. Pfaffenbach et al. showed that sporadic forms of DRS are at 10 to 20 times greater risk of having other congenital malformations divided into mainly four categories: skeletal, auricular, ocular, and neural [5].

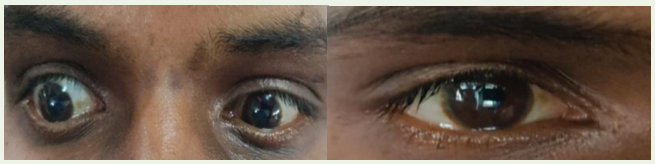

Figure 1: Right eye abduction restriction with left eye retraction and narrow palpebral fissure(left), lacrimation of eyes on chewing food(right).

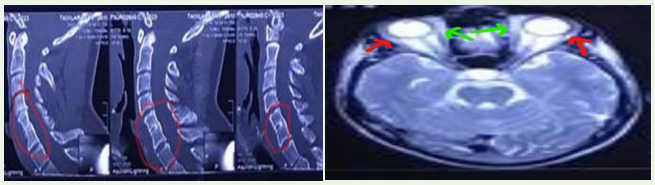

Figure 2: Partial fusion of posterior vertebral body C5,C6 Block vertebra between D3,D4 with defect in posterior vertebral body (klippelfeil type 3) (left), mri brain-bilateral hypoplasia of lateral rectus muscles(right).

Skeletal abnormalities include cleft palate, limb deformities, phocomelia, vertebral anomalies, and spina bifida. Auricular abnormalities include preauricular tags, pinna defects, and sensorineural deafness. Neural defects involve the third, fourth, and sixth cranial nerves.

Wildervanck syndrome, also known as cervico-oculo-acoustic syndrome, constitutes a triad of Klippel–Feil anomaly (fusion of >1 cervical vertebra), Duane retraction syndrome, and hearing deficits with a ten-fold female to male preponderance [6]. The Online Mendelian Inheritance in Man (OMIM) database has estimated the prevalence to be <1/1,000. A Medline search revealed that till date, only 45 cases of Wildervanck syndrome with a complete triad have been described .The interesting fact is that our patient presented with additional clinical signs apart from the complete triad described by Wildervanck.

Conclusion

Duane retraction syndrome may associated systemic defects like Wildervanck syndrome. So it becomes imperative to perform a thorough systemic examination to rule out other associated congenital anomalies. our case is a probable Wildervanck syndrome with crocodile tears which was very rarely reported in medical literature.

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