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Stilling Duane Syndrome: A Case Report

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Abstract | **Case Report**

Stilling-Duane syndrome is a rare congenital neuromuscular disorder. It constitutes a congenital pathology of oculomotor function. The Huber classification remains the most commonly accepted. In terms of treatment, no specific medical management is indicated in the absence of amblyopia. The importance of early detection of congenital oculomotor disorders is emphasized to avoid functional complications. Appropriate management and careful monitoring can preserve good visual function and ensure a satisfactory quality of life. We report the case of a child with Stilling-Duane syndrome.

Keywords: Stilling-duane syndrome, rare, congenital pathology, Huber's classification, early detection.

INTRODUCTION

STILLING DUANE SYNDROME is a rare congenital neuromuscular disorder, more common in females and often discovered in adulthood [1,2]. It results from partial or complete agenesis of the abducens nerve (VI) nucleus, compensated by aberrant innervation of the lateral rectus muscle by branches of the oculomotor nerve (III) [3,4]. The condition is characterized by a limitation of abduction in the affected eye, which may result in its misdiagnosis as an acquired sixth nerve palsy. No specific medical treatment is required, as patients generally compensate for the deficit with a strabismus or mild torticollis, allowing for normal binocular vision in most cases [5].

CASE PESENTATION

A 5-year-old child, born from a non-consanguineous marriage, with no significant past medical history, was referred for a routine pre-school ophthalmologic examination. He presented with an uncorrected visual acuity of 10/10 in both eyes.

Noncycloplegic refraction:

- Right eye : +0.25 (-0.50 X 170°)
- Left eye : +0.50 (-0.50 X 165°)

Cycloplegic refraction:

- Right eye: +1.25 S
- Left eye: + 1.00 S
- Pupillary light reflexes were normal and preserved.

Orthoptic evaluation: Fig: (a)(b)(c)(d)(e)

- Intermittent divergent strabismus accompanied by a vertical component
- Binocular fusion was present on the red glass test at near vision
- Stereoscopic vision: positive Lang Stereotest I and II
- A 45D exotropia with a 10D left hypertropia at near without correction.
- A 50D exophoria with a 10D left hypertropia at distance without correction.
- A 40D exotropia with a 12D left hypertropia at near with correction.
- A 45D exophoria with a 12D left hypertropia at distance with correction.
- Torticollis with head tilt to the left
- Marked limitation of right lateral rectus and right inferior oblique muscles
- Mild limitation of right medial rectus
- Significant overaction of the left lateral rectus
- Moderate overaction of the left inferior oblique and left medial rectus
- Impaired conjugate eye movement.

Examination of the anterior ocular segment revealed no abnormalities.

Funduscopic examination was unremarkable, revealing a normal well-defined and orange optic disc, with a physiological cup. The macula appeared normal with a preserved foveal reflex. Retinal vessels were of normal caliber, and no peripheral degenerative lesions were observed.

Diagnosis: type III Stilling-Duane syndrome.

Management: no additional paraclinical investigations were deemed necessary. Optical correction was prescribed and a regular follow-up was planned to monitor torticollis and prevent the development of amblyopia.

Fig: (a)(b)(c)(d)(e)

DISCUSSION

Stilling–Türk–Duane syndrome is a congenital ocular motility disorder classified within the spectrum of Congenital Cranial Dysinnervation Disorders (CCDDs). It accounts for approximately 1–5% of all pediatric strabismus cases [6].

This condition is classically attributed to partial or complete agenesis of the abducens (VI) nerve nucleus, associated with aberrant innervation of the lateral rectus muscle by branches of the oculomotor (III) nerve, resulting in abnormal innervation of the lateral rectus muscle [3].

Huber's classification remains the most widely accepted system for categorizing Stilling-Duane syndrome and distinguishes three types [7]:

- Type I: limitation of abduction, with normal or minimally defective adduction
- Type II: limitation of adduction, with normal abduction

- Type III: limitation of both abduction and adduction, as observed in the present case.

Type III, the rarest form, presents with a marked disturbance of horizontal ocular motility. This is characterized by the concurrent limitation of both abduction and adduction and is frequently associated with significant strabismus and paradoxical co-contraction of antagonist muscles, reflecting defective and disorganized innervation. The pronounced vertical deviation observed in our patient (constant left hypertropia), together with overaction of contralateral oblique and rectus muscles, suggests the presence of secondary contracture phenomena and compensatory muscular reorganization [8].

Congenital torticollis, represents an adaptive mechanism frequently observed in severe forms of STDS. It helps maintain functional binocular vision. While the patient's torticollis is currently mild at this stage, it requires close follow-up due to the potential for long-term musculoskeletal compensations and cervical deformities [2].

In terms of treatment, no specific medical intervention is indicated in the absence of amblyopia, as in this instance. Preserved visual acuity (10/10 in both eyes) is a favorable prognostic indicator supporting a conservative management strategy. This involves ensuring optimal optical correction and maintaining regular functional follow-up examinations. Surgical intervention is typically reserved for decompensated or poorly compensated cases [9].

Surgical procedures are highly individualized and frequently require a bilateral approach. These interventions may incorporate transpositional procedures, particularly in clinical presentations characterized by significant co-contraction phenomena [10].

CONCLUSION

Stilling-Duane-Turk syndrome is a congenital ocular motility disorder characterized by limitation of horizontal eye movement associated to globe retraction upon adduction. It results from partial or complete agenesis of the abducens nerve (VI) nucleus leading to aberrant innervation of the lateral rectus muscle by branches of the oculomotor nerve (III).

In this patient, the preservation of normal visual acuity and binocular vision justifies a conservative management strategy which involves an optimal optical correction and a regular follow-up.

The presented case emphasizes the clinical value of early detection of congenital ocular motility disorders to prevent long-term functional complications, such as amblyopia and secondary postural imbalances.

Surgical intervention is reserved for instances of significant motor decompensation or pronounced aesthetic concerns.

Ultimately, an individualized management strategy and meticulous follow-up are essential for preserving visual function and ensuring a satisfactory long-term quality of life.

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