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A Rare Case of Duane Retraction Syndrome in an Adolescent Female

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Abstract

Introduction: Duane Retraction Syndrome (DRS), also known as Stilling-Turk-Duane syndrome, is a rare congenital ocular motility disorder that results from anomalous innervation of the lateral rectus muscle due to hypoplasia or absence of the sixth cranial nerve. It is classified under the broader group of Congenital Cranial Dysinnervation Disorders (CCDDs).

Case Report: We report a rare case of Duane Retraction Syndrome type 2 in an adolescent female who presented with systemic symptoms unrelated to her ocular findings. A clinical squint examination led to the incidental diagnosis of DRS during her evaluation for dengue-induced macrocytic anemia.

Conclusion: This case underscores the importance of a multidisciplinary approach in evaluating systemic and ocular findings. Although DRS is typically diagnosed in early childhood, incidental diagnosis in adolescence should prompt thorough neurological and genetic evaluation due to its association with syndromic features.

Keywords: Adolescent, Congenital, Duane's Syndrome, Strabismus, Exotropia

Introduction

Duane Retraction Syndrome (DRS) is a congenital ocular motility disorder caused by misinnervation of the lateral rectus muscle by the oculomotor nerve due to absence or hypoplasia of the abducens nerve. First described by Alexander Duane in 1905, this condition belongs to the group of congenital cranial dysinnervation disorders (CCDDs), which include other conditions like Möbius syndrome and congenital fibrosis of extraocular muscles (CFEOM) [1,2].

Huber's classification remains the most widely accepted clinical scheme:

- **Type 1**: Limited abduction with normal or mildly limited adduction
- **Type 2**: Limited adduction with normal or slightly limited abduction (least common)
- **Type 3**: Limitation of both adduction and abduction

The hallmark features of DRS include globe retraction and narrowing of the palpebral fissure during adduction, with or without vertical overshoots. Type 2 DRS is particularly rare and typically presents with exotropia in primary gaze [3,4].

Case Report

A 13-year-old female was admitted with a 1-week history of headache, generalized weakness, and 2 days of high-grade fever. She presented with breathlessness (NYHA grade 3) and lethargy. Initial evaluations revealed severe anemia and thrombocytopenia. She tested positive for Dengue NS1 antigen, and peripheral smear showed macrocytic anemia.

Oxygen therapy was initiated along with IV fluids, blood transfusion, and antibiotics. During examination, a noticeable lateral deviation of the right eye was observed, prompting an ophthalmological referral. On evaluation, she was diagnosed with

Duane Retraction Syndrome Type 2, characterized by limited adduction in the right eye with globe retraction and narrowing of the palpebral fissure on attempted adduction. No vertical overshoots or downshoots were noted.

Brain imaging with CT showed subtle cytotoxic lesions in the splenium of the corpus callosum, likely dengue-associated encephalopathy. She responded well to supportive care and was discharged with ophthalmology and neurology follow-up. Genetic counselling and audiometry were advised.

Figure 1

Figure 2

Figure 3

Figure 1 Normal eye movement towards right

Figure 2 Lateral upshoot in right eye during abduction of right eye

Figure 3 No upshoot or downshoot

Feature	**Typical DRS Case**	**Present Case**
Age at Diagnosis	Early childhood (0–6 years)	13 years
Type	Type 1 (common), Type 2 (rare)	Type 2 (confirmed on exam)
Laterality	Left eye commonly involved	Right eye involvement
Vertical Overshoot	Frequently present in upshoots	Absent
Genetic/Syndromic Associations	Often syndromic (SALL4, CHN1)	No syndromic features identified
Imaging Findings	Cranial nerve hypoplasia/normal	Cytotoxic lesion in splenium (dengue-related)

Discussion

Duane Retraction Syndrome is often diagnosed in early childhood, with most patients presenting before the age of 10 years. Our case is notable for its **late diagnosis in adolescence** during a hospitalization for **dengue fever**—a contrast to most published literature where DRS is typically an early-life finding [5,6].

In DRS type 2, as in our patient, **adduction is primarily restricted** with relatively preserved abduction, distinguishing it from type 1 (limited abduction) and type 3 (both directions limited) [7]. Furthermore, our patient demonstrated a right-sided involvement with no vertical overshoot—a rare presentation, as vertical misalignments are reported in 25–30% of DRS cases [8].

The presence of **exotropia in primary gaze**, observed in our case, aligns with the typical presentation of DRS type 2. However, the lack of globe retraction on abduction and vertical movement anomalies makes this case even more unusual [9].

Neuroimaging findings of cytotoxic lesions in the splenium of corpus callosum are uncommon but reported in cases of dengue encephalopathy. While not directly related to DRS, this highlights the importance of **systemic evaluation** and **multidisciplinary**

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involvement in children presenting with overlapping neurological and ocular symptoms [10].

Unlike many reported DRS cases with **genetic associations**, such as mutations in **CHN1, SALL1, SALL4**, and **HOXA1**, our patient had **no syndromic features** (e.g., limb malformations, renal anomalies, or hearing loss). However, due to the late discovery and possibility of an underlying genetic etiology, further genetic testing and audiologic assessment were advised [11,12].

Our case diverges from those reported in the literature in several ways:

A recent study by Demer et al. using high-resolution MRI has shown the **absence of the abducens nerve** in type 1 and type 3 DRS, while in type 2, anomalous innervation of the medial rectus may occur [13]. Such detailed imaging was not performed in our case, though it would be recommended for a complete evaluation.

Management in DRS is individualized and often conservative if the deviation is not cosmetically significant or associated with diplopia. Surgical intervention is considered in cases of significant strabismus or head turn. Our patient did not report diplopia or abnormal head posture, so no surgical intervention was pursued [14].

Conclusion

Duane Retraction Syndrome, particularly type 2, remains a rare and often underdiagnosed condition, especially when asymptomatic or overshadowed by systemic illness. This case exemplifies the need for a **holistic and multidisciplinary evaluation** in pediatric patients presenting with systemic illnesses and incidental ocular signs. Early recognition and genetic counseling are crucial, especially considering the potential syndromic and hereditary implications.

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