Hidden in plain sight: Duane retraction syndrome diagnosed in an adult

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A 65-year-old female with diabetes mellitus of 3 years duration presented to the ophthalmology outpatient department with complaints of itching, watering, and foreign body sensation in both eyes for 1 week. On examination, she had features of allergic conjunctivitis in both eyes. She was found to have left lateral rectus palsy and was referred to the neurology department. There was no history of diplopia or headache. There was no history of any obvious squint. The general examination was normal. Her blood pressure was 130/80 mmHg, and pulse was 68/min, regular. Neurological examination showed normal visual acuity. There was no papilledema. Extraocular movement examination showed left lateral palsy (Fig. 1). On attempted right gaze, the left palpebral fissure became narrow, and there was deviation of the left eye downward (Leash phenomenon) (Fig. 2). A diagnosis of left Duane retraction syndrome (DRS) (Type 1) was made. A screening magnetic resonance imaging (MRI) of the brain did not show any obvious abnormalities. As she was asymptomatic, no treatment was offered.

DRS is a congenital ocular motility disorder characterized by limitation of horizontal eye movements associated with globe retraction and narrowing of the palpebral fissure during adduction. It belongs to a group of disorders known as congenital cranial dysinnervation disorders, in which abnormal development of cranial nerves results in aberrant innervation of the extraocular muscles. DRS accounts for approximately 1–4% of all cases of strabismus. It is usually present from birth, although it may not be recognized until later in childhood or adulthood if symptoms are mild. The condition shows a female predominance and most commonly affects the left eye, though bilateral involvement occurs in about 10–20% of cases. In many individuals, the disorder occurs sporadically, but familial cases have also been described. Depending on eye position in primary gaze, DRS can be Eso-DRS, Exo-DRS, or Ortho-DRS [1].

The fundamental abnormality in DRS is the absence or hypoplasia of the abducens nerve, which normally innervates the lateral rectus muscle responsible for eye abduction. In the absence of normal innervation, the lateral rectus muscle often receives aberrant innervation from the oculomotor nerve. Because of this abnormal wiring, both the medial rectus and lateral rectus muscles contract simultaneously during attempted adduction. This co-contraction pulls the globe posteriorly into the orbit, resulting in the characteristic globe retraction

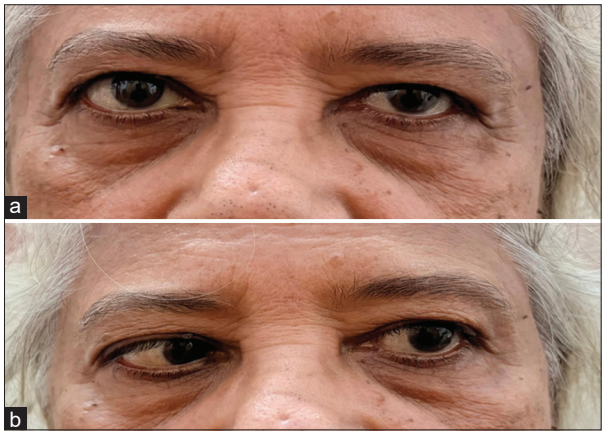

Figure 1: (a) Eyes in primary position; (b) Left lateral rectus palsy on left gaze

Figure 2: (a) Left palpebral narrowing on right gaze; (b) Downward deviation of the left eye on attempted adduction (Leash phenomenon)

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and narrowing of the eyelid fissure. During attempted abduction, the affected eye shows restricted outward movement due to the absence of normal lateral rectus function. Modern imaging studies, such as high-resolution MRI, have demonstrated the absence of the abducens nerve and nucleus in many patients, supporting the concept that DRS is primarily a developmental innervation disorder rather than a primary muscle problem.

DRS is traditionally classified into three clinical types, based on the pattern of ocular motility limitation (Heubner classification) [2]. Type I (70–80%): This is the most common form. There is marked limitation of abduction with relatively preserved adduction. Globe retraction occurs during adduction; Type II (7%): This form shows limitation of adduction, while abduction is relatively normal or only mildly affected; Type III (10–15%): Both abduction and adduction are limited. Globe retraction and narrowing of the palpebral fissure occur during attempted adduction.

The clinical presentation of DRS varies depending on the severity of the motility restriction and associated ocular deviations. Common features include limitation of horizontal eye movements, globe retraction during adduction, narrowing of the palpebral fissure on adduction, upshoot or downshoot of the globe during adduction (leash phenomenon) due to mechanical slippage of the tight lateral rectus muscle, and abnormal head posture (AHP) adopted to maintain binocular vision. In a series of 125 patients, head turn, primary position eye deviation (without head turn), and orthophoria were observed in 71.2%, 13.6%, and 15.2%, respectively [3]. About two-thirds of DRS patients had AHP, one-third had overshoots, and one-sixth had amblyopia [4]. Many patients maintain good visual acuity and binocular vision by compensating with a head turn toward the affected side. However, if significant strabismus is present in primary gaze, patients may experience diplopia or cosmetic concerns. Although many cases are isolated, DRS may occur in association with systemic or congenital anomalies. These include skeletal abnormalities, ear malformations and hearing loss, cervical spine anomalies, and renal abnormalities. DRS can also be part of certain syndromes, such as Okihiro syndrome (Duane–radial ray syndrome) and Wildervanck syndrome, which includes Duane syndrome, cervical vertebral anomalies, and hearing loss.

Diagnosis is primarily clinical, based on characteristic ocular motility findings and eyelid changes. Important diagnostic features include limited abduction or adduction, globe retraction on adduction, narrowing of the palpebral fissure during adduction, and vertical upshoots or downshoots. Neuroimaging, particularly MRI, may be used in selected cases to demonstrate the absence of the abducens nerve or to evaluate atypical presentations. Conditions that may mimic DRS include congenital sixth nerve palsy, congenital fibrosis of the extraocular muscles, Moebius syndrome, and mechanical restriction due to orbital abnormalities. Unlike acquired sixth nerve palsy, Duane syndrome typically shows globe retraction and palpebral fissure narrowing.

Management depends on the severity of symptoms and functional impairment. Many patients with mild disease and good binocular vision require no treatment. Treatment options include observation, optical correction of refractive errors and amblyopia (particularly in children), and surgical management. Surgery may be indicated in patients with significant strabismus in primary gaze, marked AHP, severe upshoots or downshoots, or cosmetic concerns. Procedures typically involve recession of the medial rectus or lateral rectus muscles to reduce co-contraction and improve alignment. However, surgery does not restore normal innervation and mainly aims to improve eye position and head posture.

The overall prognosis of DRS is generally good. Most individuals maintain stable ocular alignment and good vision throughout life, especially when the condition is mild. Early recognition is important in children to prevent amblyopia and to address AHP. Even though DRS is congenital, mild cases may be missed in childhood and can be diagnosed as late as the 60s, in this case.

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