Duane’s Syndrome: A Case Report

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ABSTRACT

The Stilling-Duane syndrome is a congenital disease responsible for restrictions of horizontal ocular movements due to total or partial agenesis of the abducens nerve or nucleus and an abnormal innervation of the lateral rectus by oculomotor nerve fibers. In its most common clinical form, type 1, the eye’s abduction movement is impacted, and it is often diagnosed as an acquired paralysis of the abducens nerve. Through this case, we want to show that it is a common error and that it is crucial to think about this syndrome before any acquired paralysis of the abducens suspicion.

Keywords: Abducens, agenesis, congenital, nerve.

1. INTRODUCTION

Stillling–Turk–Duane syndrome, or Duane retraction syndrome (DRS), is a congenital ocular motility disorder characterized by limitation of horizontal eye movements, globe retraction, and vertical movements in adduction [1]–[3]. It is divided into three types. In its most common form, the abduction limitation of the affected eye can wrongly lead to an acquired paralysis of the sixth cranial pair. Patients with DRS may have strabismus or torticollis and are often diagnosed in childhood [6].

2. CASE REPORT

We present the case of a 13-year-old girl who presented to our ophthalmology department for a follow-up of an abducens paralysis of the left eye diagnosed at the age of 21 months. She had no other past medical history, and her parents had a third-degree consanguinity. A brain and orbital MRI was performed when the diagnosis was made, and it was normal. On ophthalmological examination, her visual acuity was 20/20 in both eyes, and there was a little left-head turn. Motility examination was normal on the right eye. At the same time, it revealed a limitation of abduction of the left eye (Fig. 1) with a great retraction of the eyeball in adduction (Fig. 2). The rest of the motility exam was normal (Fig. 3), and she was orthotropic in primary position.

The slit-lamp examination and the fundus were normal in both eyes. The diagnosis of Type 1 Duane syndrome in the left eye was retained. Considering the good vision and the tolerance of the head position, we selected to follow, and no surgery was performed.

3. DISCUSSION

Duane retraction syndrome (DRS) is one of the congenital cranial dysinnervation disorders, with a prevalence of 0.01–0.05% in the general population; its incidence is 1–4% in strabismus patients [4]. The DRS has a female preponderance of 58% [5]. It is usually unilateral with left eye involvement (59%) [5]. Most DRS cases are sporadic, with only 10% of patients showing a familial pattern. They are often bilateral, and dominant and recessive forms have been documented. In dominant ones, they have shown variable penetrance and expressivity [6], [7]. Duane syndrome is often an isolated finding but may be associated with other malformations such as radial dysplasia, hearing loss, dysmorphic facies, and renal vertebral and cardiac anomalies. DRS can also be associated with other well-defined syndromes: Goldenhar, Okihiro’s, Holt-Oram, Wildervanck, and Möbius syndromes [8]. The DRS is a clinical diagnosis, often made in childhood, but sometimes it can be missed because of the normal binocular vision due to strabismus and torticollis. DS is a congenital cranial dysinnervation disorder (CCDD).

Environmental and genetic factors are known to play a role; this probably occurs between the fourth and the eighth week of pregnancy and is due to poor development of tiny parts of the brainstem that control the eye muscles. In Duane syndrome, the sixth cranial nerve that controls the lateral rectus muscle does not develop properly. In place of the abducens nerve is a branch from the oculomotor
nerve (cranial nerve III), which normally supplies other ocular muscles, including the medial rectus. When patients with DRS attempt to move their eyes inward, the medial and lateral rectus contract simultaneously, resulting in the eyeball retracting inward. Moreover, the eye opening is narrowing. Besides, muscle modifications of the medial and the lateral rectus were noticed (fibrosis and hypertrophy) [7]. Unlike abducens paralysis, RDS has no muscular atrophy due to the III’s innervation of the lateral rectus muscle. However, we can find other abnormalities, which consist of hypertrophy (33%), bumpy muscles (15%), and both hypertrophic and bumpy muscles (11%). We also find an aspect of splitting the lateral rectus; the other muscles affected are the medial rectus (hypertrophy in 56% of cases) and rarely the superior oblique (hypertrophy in 7% of cases) [9]. Huber’s well-known classification divided DRS into three types [10]. IRM also contributes to a better definition of the clinical forms by analyzing the anomalies of the VI nerve [11]. Type 1, the most frequent (78%) [7], is likely to be confused with VI nerve paralysis. It is a marked limitation of abduction explicable by an absence or hypoplasia of the sixth nerve and a maximum innervation reaching the lateral rectus muscle only when the affected eye is adducted. Type 2 (7%) [7] is a limitation of adduction; in this form, there is a partial agenesis of the VI nerve, which remains functional. Thus, the lateral rectus is innervated by twice the sixth nerve and some fiber from the third nerve, which send the same branches to the lateral and the medial rectus; during the adduction, the medial and lateral rectus contract with the same forces, which prevent movement. Type 3 (15%) [7] is a limitation of both adduction and abduction, which can be explained by a co-contraction, accompanied by a loss of innervation to the lateral rectus on attempted abduction. These typical types of DRS are characterized by common features, including globe retraction on adduction and a narrowing of the palpebral fissure during adduction. Management may involve observation, which concerns most patients, who compensate for their trouble with a little head tilt or strabismus. However, sometimes, in some bothersome strabismus or tilt head, specific management can be required, such as open tical correction, amblyopia reduction, surgical methods, such as horizontal muscle recession, vertical muscle transposition, or a combination of these.

4. Conclusion

The Stilling-Duane syndrome is a rare diagnosis, well known by pediatricians and ophthalmologists because it is most frequently observed during childhood. However, it can sometimes go undiagnosed and be discovered only in adults. Therefore, neurologists should be conscious of this differential diagnosis of acquired paralysis of the abducens nerve.

Conflict of Interest

The authors declare that they do not have any conflict of interest.

References
