CASE REPORT

Duane retraction syndrome: MRI features in two cases

Syndrome de rétraction : à propos de deux cas de Stilling-Duane en IRM

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Abstract
Purpose. — Neuroimaging findings in Duane’s retraction syndrome (DRS), through magnetic resonance imaging (MRI), suggest that aplasia of the abducens nerve (VI) can be responsible for several forms of DRS.

Methods. — Brain MRI was performed in two children of 2.5 and 7 years of age with left sided DRS type II and right sided DRS type I, respectively. 3D T2 weighted images through the brain-stem were acquired in order to visualize the cranial nerves especially the abducens (VI) and oculomotor (III) nerves.

Results. — The abducens nerve on the affected side could not be observed in these two different types of DRS with normal morphology of the third nerves.

Conclusion. — Absence of the VI nerve has been described recently in DRS types I and III only, while DRS type II is usually associated with normal VI nerve on MRI. However our results show that aplasia of the VI nerve can also be seen in DRS type II resulting in new insight of the pathogenesis of this clinical entity.
Introduction

Duane retraction syndrome (DRS) is a congenital horizontal eye movement disorder characterized by a marked limitation or absence of abduction, variable limitation of adduction, palpebral fissure narrowing and globe retraction on attempted adduction [2]. Among the four types of DRS described, types I and III are the most frequent while types II and IV are extremely rare [12,8]. We report two cases of DRS types I and II, respectively, with absence of the VI nerve on magnetic resonance imaging (MRI). This MR picture is concordant with the recent literature in DRS type I while it is a totally unusual and possibly paradoxical finding in DRS type II [5].

Case report 1

A 7-year-old girl was referred because of increased headaches since 1 year due to abnormal head posture in relation to DRS. There was no family history of strabismus or congenital anomaly.

Visual acuity was 18/20 in the right eye and 20/20 in the left eye. Refractive error measured under cycloplegia showed hypermetropia of +0.50 in both eyes.

A right head turn was noted with a typical right sided DRS type I: the horizontal duction in the right eye gaze showed unilateral limitation of abduction with enlarged palpebral fissure (Fig. 1a) and palpebral fissure narrowing and globe retraction on attempted adduction (Fig. 1c).

There was no vertical deviation of the right eye in adduction (Fig. 1c). In forced primary position, a convergent strabismus of the right eye was noted as an esotropia of 10 diopters (D) at distance and near vision. When the right eye was fixing, a convergent strabismus of the left eye was also noted as an 18 D esodeviation (Fig. 1b). This contralateral deviation is commonly thought to be related to muscular fibrosis of horizontal ocular muscles.

Slit-lamp and fundus examinations of both eyes were unremarkable. Ear-nose-throat (ENT) and general examinations were normal without associated developmental anomalies such as congenital neurological deficit, crocodile tears, sensorineural hearing loss, structural defects involving skeletal and neural structures.

MRI was ordered for this isolated DRS type I: the abducens nerve on the affected right side could not be detected.

Surgery was performed on both medial and lateral rectus muscles of the right eye. After recession of the two horizontal recti (9 mm for the medial rectus and 5 mm for the lateral rectus), the abnormal head posture disappeared with slight improvement of abduction and headaches resolved. The child is still on follow up.

Case report 2

The patient was a 2.5-year-old boy who was referred because a head tilt was noted since birth. Family history was unremarkable. Pregnancy and delivery were reported normal with no neonatal injury.

Visual acuity was 20/20 for both eyes. Refractive error measured under cycloplegia showed bilateral hypermetropia (+1.25 in the right eye, +1.75 in the left eye) and bilateral astigmatism (+0.75 at 70° in the right eye, +0.25 at 90° in the left eye).

Ophthalmologic assessment noted a right head turn and a typical left sided DRS type II: horizontal ductions showed full abduction of the left eye gaze showed unilateral limitation of abduction with enlarged palpebral fissure (Fig. 2a) and palpebral fissure narrowing and globe retraction on attempted adduction (Fig. 2a).

In forced primary position, a divergent strabismus of the affected eye was noted as an exotropia of 4 D at distance and near vision (not shown).

Slit-lamp and fundus examinations of both eyes were unremarkable. ENT and general examinations were without developmental anomaly. A full optical correction was prescribed.

MRI under anesthesia was ordered for this isolated DRS type II: the abducens nerve on the affected side could not be detected (Fig. 3a, b).

Surgery was not indicated because of the mild abnormal head posture and the young age (binocular vision being not consolidated at that age) and the child is still on follow up.
Brain and orbit MRI were performed on a 1.5 T magnet (Symphony TIM, Siemens, Erlangen). Thin 3D T2 weighted images were obtained with a multichannel head coil by the CISS sequence to visualize cisternal segments of cranial nerves especially the VI and III. This sequence allows section thickness of 0.5 mm and reconstructions with isotropic voxel size of 0.5 × 0.5 × 0.5 mm. The parameters of this sequence are as follow: repetition time 11.2, echo time 5.6, matrix 268 × 384, field of view 250, 64 sections. The brain stem was analyzed from cerebral peduncles to medulla with multiple oblique reconstructions to look at entry/exit zones and cisternal courses of cranial nerves. This brainstem analysis is similar to all MRI performed in cases with oculomotor paralysis.

For both cases the VI nerve of the affected sides could not be detected with no abnormal exit zone (Fig. 3a, b). The third nerves were unremarkable (Fig. 3c). Cerebral parenchyma and oculomotor muscles were normal (Fig. 3d).

**Discussion**

Classification of the DRS is currently based on clinical examination and four types are described with a constant sign as narrowing of palpebral fissure on adduction secondary to the retraction of the eye. DRS I presents limited or absent abduction; DRS II shows limited or absent adduction with relatively normal abduction; DRS III displays limitation of both adduction and abduction; DRS IV consists of synergistic divergence, paradoxal abduction on attempted adduction and limited abduction. Vertical ocular movements are often noted on adduction, most frequently in an upward direction [8,11]. Patients with DRS usually have binocular single vision with good stereopsis in the field of preserved motility.

Approximately 75% of DRS are of type I while type III accounts for most of the rest and type II and synergistic divergence are extremely rare. DRS is usually unilateral and bilateral cases do not exceed 15-20% of cases. Most cases are sporadic but familial cases have been described whether uni- or bilateral [2,6,7,12]. We present here a
common case of DRS type I and an extremely rare DRS type II.

DRS are the consequence of congenital anomaly of cranial nerve nuclei with aberrant innervations suggested by electromyography in the 1970s [4] and confirmed by histology in the 1980s [3,9]. The four forms of DRS are classified with regard to the different distribution of inappropriate neural input from the oculomotor and abducens nerves to lateral and medial recti. Electromyography studies [4] revealed aberrant innervations from the abducens nerve (III) that was thought to be the result of congenitally deficient nucleus of the VI nerve [6], leading to fibrosis of lateral rectus muscle. Therefore branches of abducens nerve innervate the lateral muscle with consequent retraction of the globe due to the co-contraction of medial and lateral rectus muscles [1,4]. In DRS type I characterized clinically by poor abduction and good adduction, the medial rectus muscle receives most of the innervations from the III nerve while minimal innervations are redirected to the lateral rectus muscle. As a consequence Duane’s affected eye is usually fixed in adduction with a convergent strabismus in primary position with a compensatory head tilt toward the affected eye (Fig. 1). In DRS type II characterized clinically by poor adduction and good abduction, EMG recordings showed that the lateral rectus was contracted appropriate on abduction with paradoxical contraction on adduction [4]. Those findings therefore suggested probable partial innervations of the lateral muscle by the sixth nerve (because of the full abduction), plus splitting of the III nerve branches innervating the medial rectus muscle toward the lateral rectus muscle [6]. The lateral rectus muscle receives most of the innervations from the III nerve (as opposed to DRS type I). Consequently the affected eye is abducted with divergent strabismus in primary position with the face turn toward the opposite side.

Histological studies exist only for DRS types I and III [3,9, 10] and show aplasia or hypoplasia of the abducens nucleus and nerve with partial innervations of the lateral rectus muscle by the inferior division of the oculomotor nerve (III) with muscular fibrosis in non-innervated areas.

Absence of the abducens nerve on the affected side has been described in a DRS type I in 1998 [11] by MRI. A recent study from 2005 [5] reported 23 patients with DRS: in 16 patients with type I the abducens nerve could not be observed in all cases whereas the nerve was always present in the two patients with DRS type II; in DRS type III the abducens nerve was visualized in two of five cases.

Our report is suggesting that DRS type II can also be associated with aplasia/hypoplasia of the VI nerve as described in DRS type I. This MR picture is joining and highlighting electromyography reports of aberrant innervations from the III nerve redirected toward the lateral rectus muscle. Therefore our case of DRS type II is displaying a new insight in the understanding of the pathogenesis of this clinical entity: aplasia of the VI nerve is responsible, as in DRS types I and III, for the aberrant innervations from aberrant nerve pathway of the oculomotor nerve explaining the respected abduction of the affected eye in DRS type II. However this clinical entity is extremely rare and aberrant innervations of the III nerve cannot be assessed by MRI even with isotropic voxel size of 0.5 mm. Emerging 3 T magnet will probably help detecting abnormal nuclei within the brainstem and small branches of oculomotor nerve. However, understanding of the pathogenesis of DRS type II requires more cases than already reported.

References