

Case Report

Congenital Multiple Ocular Motor Nerve Palsy Complicated by Splitting of the Lateral Rectus Muscle

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We report a case of congenital multiple ocular motor nerve palsy combined with splitting of the lateral rectus muscle (LR). A 59-year-old Japanese female was investigated for worsening esotropia after corrective surgery. She presented with left hypertropia (35Δ) and esotropia (45-50Δ). Orbital magnetic resonance imaging (MRI) showed reduced belly sizes in the superior rectus, inferior rectus, and superior oblique muscles and splitting of the LR, extending from the origin to the belly, in the left eye. Splitting of the LR belly was detected on MRI in a case of congenital multiple ocular motor nerve palsy.

Key words: multiple ocular motor nerve palsy, congenital cranial dysinnervation disorder, lateral rectus muscle splitting, orbital connective tissue, magnetic resonance imaging

In embryology, the development of the horizontal rectus muscle is completed when the superior mesenchymal complex and inferior mesenchymal complex fuse into one entity [1]. An incomplete fusion of the extraocular muscles is observed in some cases of congenital ocular motor disorders, *e.g.*, congenital cranial dysinnervation disorders (CCDDs) including Duane syndrome, congenital oculomotor nerve palsy, and congenital trochlear nerve palsy [2-4].

Compartmentalized innervation of the primate lateral rectus muscle was reported in a histological study [5]. That study's authors concluded that the consistent intramuscular segregation of the abducens nerve arborization in humans and monkeys suggests that the lateral rectus muscle (LR) has functionally distinct superior and inferior zones. Compartmentalization of the

medial rectus muscle was also reported [6]. The splitting of the horizontal rectus muscles seen in CCDD [2-4] and lateral rectus superior compartment palsy [7] were cited as clinical evidence in support of the compartmentalization of the horizontal rectus muscles. Splitting of the horizontal rectus muscles has been reported in cases of single congenital ocular motor nerve palsy and Duane syndrome, which are CCDD [2-4].

We report a case of congenital multiple ocular motor nerve palsy (a CCDD manifesting as oculomotor nerve palsy and abducens nerve palsy) complicated with splitting of the LR belly into superior and anterior portions. This case was accompanied by degeneration of the orbital connective tissues, presumably due to aging.

Case Report

A 59-year-old Japanese female who had suffered from oculomotor nerve palsy and abducens nerve palsy in her left eye since birth presented at Okayama University Hospital because of worsening ocular misalignment.

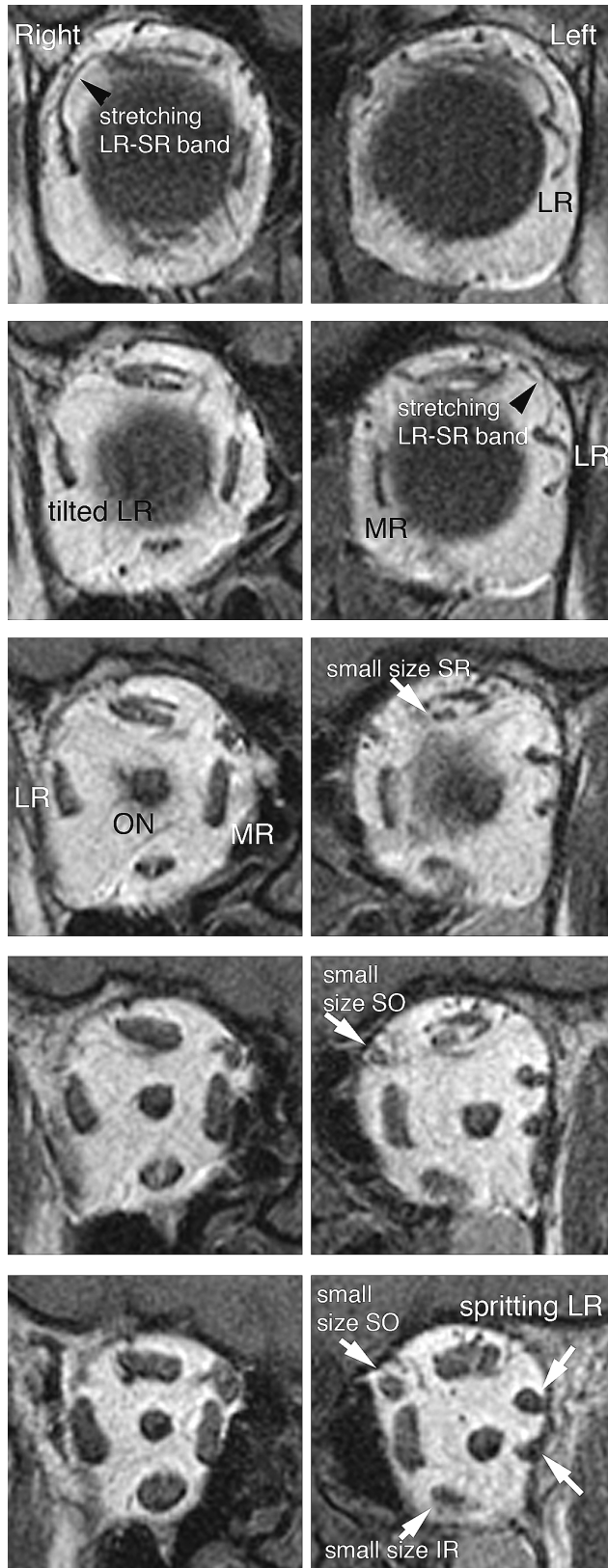
At 36 years of age, the patient presented at our department for the first time. An examination of the left eye revealed abduction and infraduction limitations accompanied by hyperesotropia (esotropia: 4Δ ; hypertropia: $30\text{--}35\Delta$). She was diagnosed with oculomotor nerve palsy and abducens nerve palsy in the left eye. Superior rectus muscle (SR) recession (4 mm) and inferior rectus muscle (IR) resection (5 mm) were conducted in the left eye. At the age of 42 years, she underwent additional IR recession (4 mm) in the right eye as a treatment for residual esotropia of 14Δ and left hypertropia of 25Δ .

At the most recent consultation, her corrected visual acuity was $1.0 \times \text{cyl-}0.5\text{D Ax.}120^\circ$ in the right eye and 0.4 (n.c.) in the left eye. An examination of the left eye showed abduction and infraduction limitations together with hyperesotropia (esotropia: $45\text{--}50\Delta$; hypertropia: 35Δ) (Fig. 1). A marked limitation of infraduction

in abduction and a slight limitation of infraduction in adduction also were observed in the left eye. Magnetic resonance imaging (MRI) did not reveal any marked abnormalities in the cavernous sinus segments; the vicinity of the cisternal segments of the oculomotor or abducens nerves; or the region surrounding the oculomotor nucleus, trochlear nucleus, and abducens nucleus. However, hypoplasia of the SR and IR, superior oblique muscle (SO), and LR, as well as splitting of the LR from its origin to the middle of its belly were observed in the left eye (Figs. 2, 3). Temporal stretching and displacement of the connective tissue band connecting the LR and SR (the LR-SR band) as well as superior displacement of the LR were observed bilaterally. Temporal tilting of the upper part of the LR was noted in the right eye. Based on these findings, we diagnosed congenital multiple ocular motor nerve palsy complicated by morphological abnormalities of the extraocular muscles and orbital connective tissue degeneration. Additional medial rectus muscle (MR) recession (6 mm) and LR resection (8 mm) had been performed in the left eye when the patient was 36 years old. However, the results of this surgical correction were not as good as expected, with residual esotropia of 18Δ and left hypertropia of 45Δ detected.



Fig. 1 Photographs of the patient in 9 gaze positions at 59 years of age. In the primary position, esotropia and left-sided hypertropia were observed. In the left eye, a limitation of abduction (movement beyond the midline), a limitation of infraduction (movement only to the primary position), and a limitation of supraduction (movement beyond the midline) were observed. A marked limitation of infraduction in abduction and a slight limitation of infraduction in adduction were also observed in the left eye.



Discussion

According to MRI studies, morphological abnormalities are detected at relatively high frequencies in CCDDs. Demer *et al.* [2] detected hypoplasia of the LR in the affected eye or both eyes in 5 of 7 patients (71%) with Duane syndrome who exhibited splitting of the LR into superior and inferior portions. In a series of 35 cases of CCDD studied by Okanobu *et al.* [4], muscle belly splitting was observed in the MR in one of four cases (25%) of oculomotor palsy, and in the LR in one of 26 cases (4%) of congenital superior oblique palsy, and in two of five cases (40%) of Duane syndrome. In a study by Okanobu *et al.*, splitting of the horizontal rectus muscles was detected in CCDD, but not in acquired ocular motor palsy or normal subjects [4]. The splitting of the horizontal rectus muscles seen in CCDDs was not only cited as clinical evidence in support of the compartmentalization of the horizontal rectus muscles; it was suggested that it might also be related to the etiology of CCDDs.

In the present case, the cross-sectional areas of the bellies of the SR, IR, and SO were smaller in the affected eye than in the contralateral eye. In addition, the LR was split from its origin to its belly into superior and inferior portions, and the belly of the LR had a markedly reduced cross-sectional area from the origin to the scleral insertion. These MRI findings led to a diagnosis of multiple ocular motor nerve palsy, comprising oculomotor nerve palsy, trochlear nerve palsy, and abduction nerve palsy in the left eye. To the best of

Fig. 2 Quasi-coronal T1-weighted MR images (thickness: 3 mm) of the left and right eyes in the central gaze position. *Upper row:* Anterior side. *Lower row:* posterior side. *Middle slice:* Cross-section around the globe-optic nerve junction. In the left eye, the posterior portion of the LR was completely split in two, whereas the anterior portion remained in one piece. Compared with that seen in the right eye, the LR in the left eye was smaller, and the size difference was particularly marked in the posterior portion. The IR, SR, and SO in the left eye were also smaller than those in the right eye. Bilateral temporal stretching and displacement of the connective tissue band connecting the LR and SR (the LR-SR band) were observed (*black arrowheads*), whereas temporal tilting of the upper portion of the LR was seen in the right eye (*uppermost row and second row from the top*). In both eyes, the LR was displaced superiorly compared with the MR. IR, inferior rectus muscle; LR, lateral rectus muscle; MR, medial rectus muscle; ON, optic nerve; SO, superior oblique muscle; SR, superior rectus muscle.

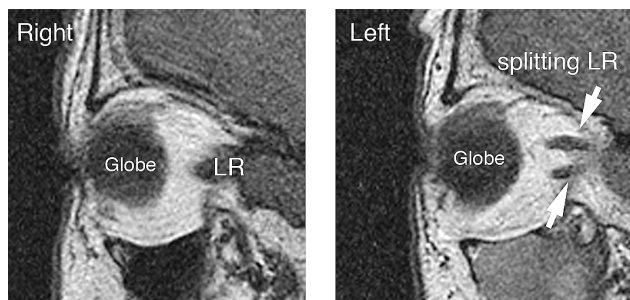


Fig. 3 Quasi-sagittal T1-weighted MR images of the right and left eyes obtained at 4-10 mm temporal to the globe-optic nerve junction in the central gaze position. In the left eye, the belly of the posterior lateral rectus muscle (LR) was split into superior and inferior portions.

our knowledge, this is the first report of splitting of the LR into superior and inferior portions being detected on MRI in a case of congenital multiple ocular motor nerve palsy.

Our patient's case also involved bilateral temporal stretching and displacement of the LR-SR band and temporal tilting of the upper LR in the right eye, which are known to be due to age-related degeneration of the orbital connective tissue [8,9]. Abnormal LR-SR band findings are detected in approx. 30% of healthy individuals aged ≥ 50 [10]. On the other hand, our patient's case did not involve inferior displacement of the LR, which is known to be an age-related change [8,9], but it did display bilateral superior displacement of the LR, which is a characteristic of A-pattern strabismus in congenital pulley heterotopy [11].

In the present case, left vertical rectus muscle surgery was performed during the first operation, which had been carried out 24 years ago. In the third operation, left horizontal rectus muscle surgery was conducted. Quadruple rectus muscle surgery might increase the risk of anterior segment ischemia. However, we judged that the risk of complications was low because the anterior segment would have healed over the 24-year period between the operations. There were no findings that were indicative of anterior segment ischemia or ocular phthisis at one postoperative year. The patient subsequently decided to change hospitals, but careful ophthalmological follow-up will be necessary in the future.

Conclusion

We report that splitting of the LR belly was detected on MRI in a case of congenital multiple ocular motor nerve palsy. The splitting of the horizontal rectus muscles seen in CCDDs was cited as clinical evidence in support of the compartmentalization of the horizontal rectus muscles, and this splitting might be one of the characteristics of CCDDs.

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