Neurological Sciences (2019) 40:2645–2647
https://doi.org/10.1007/s10072-019-03961-w

LETTER TO THE EDITOR

Combined superior rectus hypoplasia and superior oblique palsy without a trochlear nerve

Hee Kyung Yang [1] · Jae Hyoung Kim [2] · Jeong-Min Hwang [1]

Received: 3 April 2019 / Accepted: 31 May 2019 / Published online: 18 June 2019

Dear Editor,

Congenital cranial dysinnervation disorders (CCDDs) represent a group of neurodevelopmental diseases of the brainstem and cranial nerves [1]. Congenital superior oblique palsy (SOP) is one of the most representative CCDD because 73% of patients show an absent trochlear nerve and superior oblique hypoplasia [2]. CCDD is not limited to a single nerve and sometimes affects two or more nerves [3–7]. Coexistence of SOP with other CCDDs such as Duane's retraction syndrome has been reported [2]. However, combined superior rectus hypoplasia and superior oblique palsy without a trochlear nerve has never been reported.

A 35-year-old man presented with persistent diplopia for the past 5 years and an abnormal head posture consisting of a face turn to the right, head tilt to the left, and chin up position. He had noticeable asymmetry of the face. He had previously undergone strabismus surgery twice of a right superior rectus resection followed by left superior rectus recession 1 year apart in another university hospital. Diplopia had improved immediately after surgery, but recurred a few weeks later. His past medical history was unremarkable. He denied any previous event of head trauma.

On examination, his best-corrected visual acuities were 20/20 OU. He showed right hypotropia (RHoT) of 18 prism diopters (Δ) at distance and RHoT 12Δ at near on alternate prism and cover test in the primary position. He showed RHoT 25Δ in the right gaze, RHoT 18Δ in the left gaze, RHoT 6Δ on right head tilt, and RHoT 16Δ on left head tilt. The right eye showed limited elevation (−2) and depression on adduction (−1) (Fig. 1). He showed pseudoptosis in the right eye. The Lancaster red-green test revealed RHoT and extorsion (Fig. 2). Ocular versions and the Lancaster red-green test results before the first surgery in an outside university hospital were similar. The thyroid function tests, anti-acetylcholine receptor-antibody test, and repetitive nerve stimulation test were all negative.

T2-weighted coronal imaging was obtained with 1-mm slice thickness for the orbit to evaluate extraocular muscles.

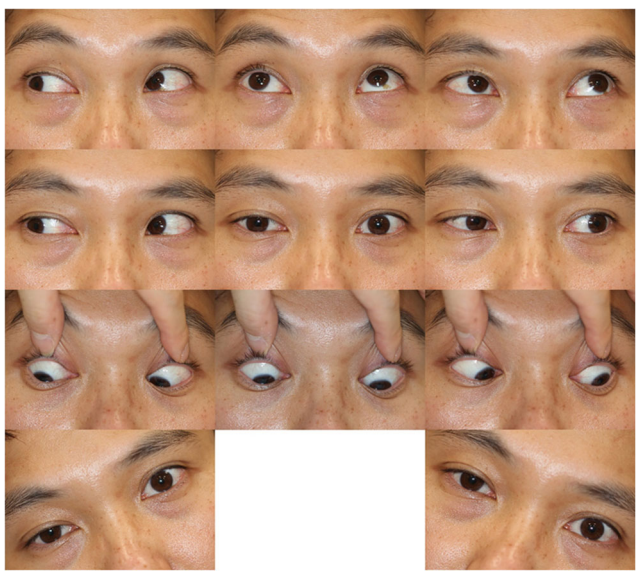

Fig. 1 Ocular versions demonstrating limited elevation (−2) and depression on adduction (−1) of the right eye

Hee Kyung Yang and Jae Hyoung Kim contributed equally to the work; therefore, they should be regarded as equivalent authors.

✉ Jeong-Min Hwang
hjm@snubh.org

Hee Kyung Yang
eye@snubh.org

Jae Hyoung Kim
jaehkim@snubh.org

[1] Department of Ophthalmology, Seoul National University College of Medicine, Seoul National University Bundang Hospital, 166, Gumiro, Bundang-gu, Seongnam, Gyeonggi-do 463-707, South Korea

[2] Department of Radiology, Seoul National University College of Medicine, Seoul National University Bundang Hospital, Seongnam, South Korea

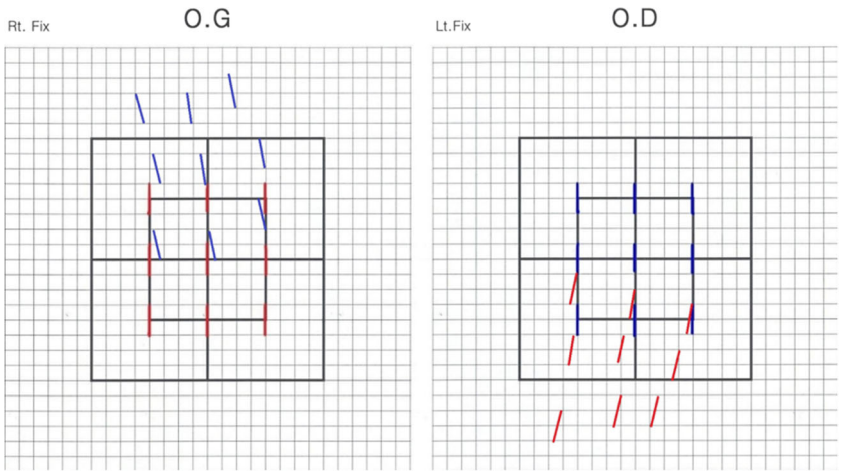

Fig. 2 The Lancaster red-green test revealed right hypotropia and extorsion

T2-weighted axial imaging was conducted with 0.25-mm thickness for the trochlear nerve and 1.4-mm thickness for the oculomotor nerve and abducens nerve in the basal cistern using a 3-Tesla MRI system (Intera Achieva; Philips Healthcare, Best, the Netherlands). The right superior oblique was hypoplastic (Fig. 3a), and the right trochlear nerve was absent (Fig. 3b). The right superior rectus was hypoplastic (Fig. 3a), and the right oculomotor nerve was normal in size (Fig. 3c). All the other extraocular muscles and the abducens nerves were normal in size.

In this report, we showed a patient with combined superior rectus hypoplasia and superior oblique palsy without a trochlear nerve. Elevation deficiency can be caused by many conditions such as superior rectus palsy, inferior oblique palsy, monocular elevation deficiency, Brown's syndrome, superior oblique overaction, blowout fracture, adherence syndrome, and congenital ocular fibrosis syndrome. Among them, superior rectus atrophy may be found in superior rectus palsy. There is a chance that he may have acquired superior rectus palsy. However, he did not report any history of head trauma

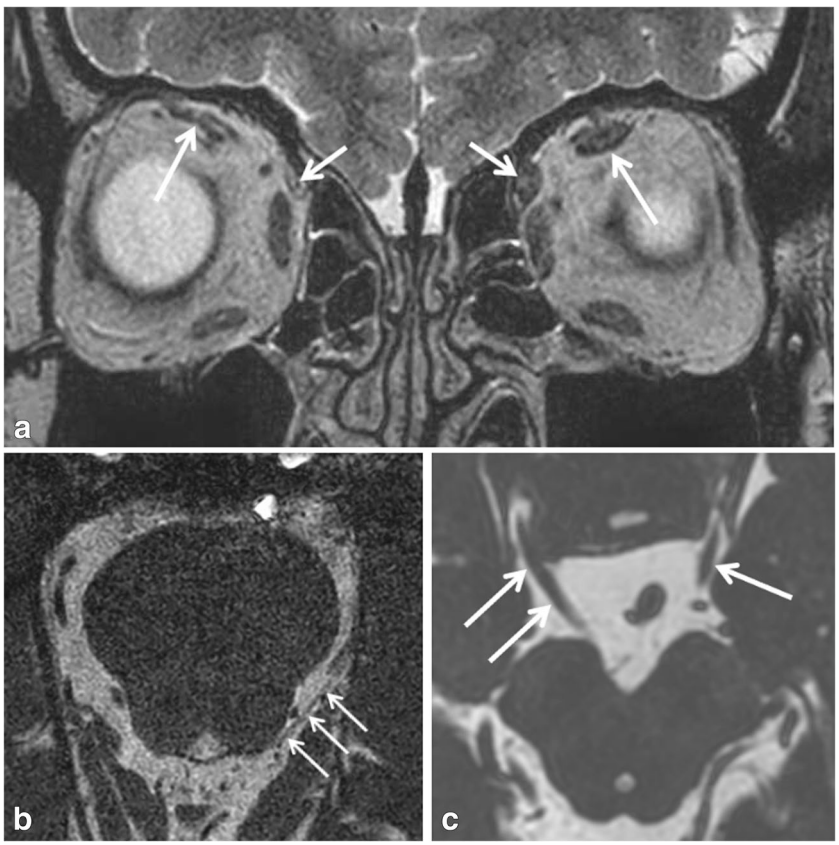

Fig. 3 **a** On coronal MR imaging of the orbit, superior oblique (short arrows) and superior rectus muscles (long arrows) are hypoplastic on the right side. **b** On axial MR imaging at the upper pons, the left trochlear nerve is well identified (arrows), but the right is not observed. **c** On axial MR imaging at the lower midbrain, right and left oculomotor nerves are symmetrically normal in size (arrows)

and absence of the right trochlear nerve mostly likely indicates a congeintal etiology; therefore, the chance of CCDD including part of the oculomotor nerve accompanied by trochlear nerve hypoplasia could not be denied. In conclusion, CCDD can affect more than one nerve with a combination of the trochlear nerve and part of the oculomotor nerve as in this case.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

References

1. Gutowski NJ, Bosley TM, Engle EC (2003) 110th ENMC International Workshop: the congenital cranial dysinnervation disorders (CCDDs). Naarden, The Netherlands, 25-27 October, 2002. Neuromuscul Disord 13:573–578
2. Yang HK, Kim JH, Hwang JM (2012) Congenital superior oblique palsy and trochlear nerve absence: a clinical and radiological study. Ophthalmology 119:170–178
3. Kim JH, Kim SK, Hwang JM (2014) Combined bilateral absence of trochlear and abducens nerve. Neurol Sci 35:1457–1459
4. Kim JH, Hwang JM (2005) Hypoplastic oculomotor nerve and absent abducens nerve in congenital fibrosis syndrome and synergistic divergence with magnetic resonance imaging. Ophthalmology 112: 728–732
5. Park C, Kim JH, Hwang JM (2012) Coexistence of Mobius syndrome and Duane's retraction syndrome. Graefes Arch Clin Exp Ophthalmol 250:1707–1709
6. Kim JH, Hwang JM (2017) Imaging of cranial nerves III, IV, VI in congenital cranial dysinnervation disorders. Korean J Ophthalmol 31:183–193
7. Hwang JM, Park SH (2001) A case of Marcus Gunn jaw winking and pseudo inferior oblique overaction. Am J Ophthalmol 131:148–150