

Development of acquired non-accommodative esotropia in a patient without a trochlear nerve

Hee Kyung Yang¹, Jae Hyoung Kim², Jeong-Min Hwang¹

¹Department of Ophthalmology, Seoul National University College of Medicine, Seoul National University Bundang Hospital, Seongnam 13620, Republic of Korea

²Department of Radiology, Seoul National University College of Medicine, Seoul National University Bundang Hospital, Seongnam 13620, Republic of Korea

Co-first authors: Hee Kyung Yang and Jae Hyoung Kim

Correspondence to: Jeong-Min Hwang. Department of Ophthalmology, Seoul National University Bundang Hospital, 82, Gumi-ro, 173-beon-gil, Bundang-gu, Seongnam 13620, Republic of Korea. hjm@snuh.org

Received: 2021-03-02 Accepted: 2021-05-07

DOI: [10.18240/ijo.2022.06.26](https://doi.org/10.18240/ijo.2022.06.26)

Citation: Yang HK, Kim JH, Hwang JM. Development of acquired non-accommodative esotropia in a patient without a trochlear nerve.

Int J Ophthalmol 2022;15(6):1030-1031

Dear Editor,

Congenital cranial dysinnervation disorders (CCDD) are neurodevelopmental diseases of the brainstem and cranial nerves^[1]. Congenital superior oblique palsy (SOP) is one of the most representative CCDDs because approximately three quarters of patients show trochlear nerve agenesis and superior oblique (SO) hypoplasia^[2]. Vertical diplopia associated with congenital SOP could emerge during the later life due to acute decompensation of vertical fusion^[3-4]. Meanwhile, acquired non-accommodative esotropia (ANAET) is one of the common causes of horizontal diplopia^[5]. Neuroradiological investigation has revealed Chiari malformation or intracranial masses in patients with ANAET^[5]. However, the association of trochlear nerve aplasia and ANAET has never been reported. Herein, we found a patient with ANAET in whom trochlear nerve aplasia was confirmed with high-resolution thin-section magnetic resonance imaging (MRI).

A 10-year-old boy presented with intermittent esotropia which developed one month prior. He was born full term with no significant perinatal history. He denied any history of previous head trauma or family history of strabismus. His parents denied any history of longstanding or intermittent head tilt

which was also not found in old photographs.

On ophthalmologic examination, he showed no head tilt or facial asymmetry. His visual acuities were 20/25 OU. Prism and alternate cover test revealed constant esotropia of 45 prism diopters (PD) at distance and at near in the primary position. He showed additional right hypertropia of 4 PD only with the head tilted to the right. Ductions and versions showed a mild limitation of depression on adduction OD (Figure 1A). The pupils were round and isocoric in both eyes. Dilated fundus examination was unremarkable in both eyes. Cycloplegic refraction showed +1.25 D OD and +1.50 D OS.

Using a 3-tesla MRI system (Ingenia CX, Philips, Best, the Netherlands), T2-weighted coronal imaging of the orbit and high-resolution transverse imaging of the cranial nerves were performed. Mild hypoplasia of the right SO muscle was observed (Figure 1B), and the right trochlear nerve was absent (Figure 1C-1E).

In this report, a patient with ANAET was found to have trochlear nerve aplasia which was unexpected because he showed no vertical diplopia and vertical deviation in the primary position, nor any head tilt and facial asymmetry. The only evidence of trochlear nerve aplasia was a right hypertropia of 4 PD only with the head tilted to the right and a mild limitation of depression on adduction. Therefore, it is hard to believe that ANAET developed associated with decompensated congenital SOP.

The typical characteristics of congenital SOP patients with trochlear nerve aplasia include an early onset of severe head tilt before 1 year of age, and apparent facial asymmetry^[2]. However, our patient did not show any of the characteristics of trochlear nerve aplasia, thus congenital SOP could not be expected. Dosunmu *et al*^[6] found that manifestation of symptoms in presumed congenital SOP patients peaked in the fourth decade, therefore there is a chance that our patient may develop vertical diplopia later in his life.

Regarding the horizontal deviation associated with SOP, horizontal strabismus was present in 42% and exotropia was eight times more prevalent than esotropia in Asian patients without the trochlear nerve^[2]. Helveston *et al*^[7] also reported that 75% were exotropes in unilateral SOP patients who received surgical treatment. In contrast, Hata *et al*^[8] reported

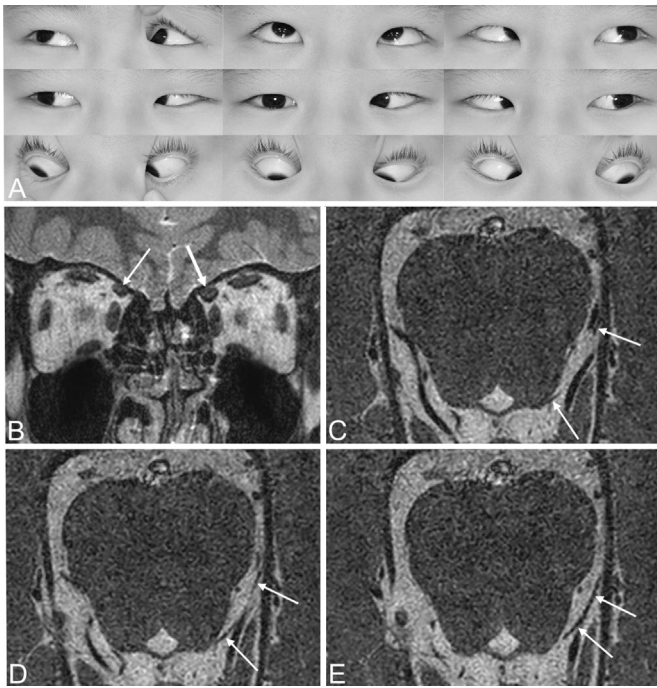


Figure 1 Acquired non-accommodative esotropia accompanied with right trochlear nerve aplasia A: Ocular versions show esotropia OS and a mild limited depression on adduction OD; B: On T2-weighted orbital image, the right superior oblique muscle (thin arrow) is slightly hypoplastic when compared with the left (thick arrow); C-E: On three consecutive high-resolution images obtained at the pons-midbrain junction level, the left trochlear nerve (arrows) is clearly identified as a curvilinear non-branching structure emerging from the inferior margin of the inferior colliculus and coursing in the anterolateral direction toward the ipsilateral tentorium. The right trochlear nerve is not identified.

that esotropia was more common than exotropia in isolated decompensated SOP which was contrary to the previous reports. Therefore, further studies would be necessary to

elucidate the characteristics of horizontal deviation associated with congenital SOP.

In conclusion, trochlear nerve aplasia could be found in a patient with acquired, comitant non-accommodative esotropia.

ACKNOWLEDGEMENTS

Conflicts of Interest: Yang HK, None; Kim JH, None; Hwang JM, None.

REFERENCES

- 1 Kim JH, Hwang JM. Imaging of cranial nerves III, IV, VI in congenital cranial dysinnervation disorders. *Korean J Ophthalmol* 2017;31(3): 183-193.
- 2 Yang HK, Kim JH, Hwang JM. Congenital superior oblique palsy and trochlear nerve absence: a clinical and radiological study. *Ophthalmology* 2012;119(1):170-177.
- 3 Kim JH, Hwang JM. Usefulness of magnetic resonance imaging in a patient with diplopia after cataract surgery. *Graefes Arch Clin Exp Ophthalmol* 2012;250(1):151-153.
- 4 Lee S, Kim SH, Yang HK, Hwang JM, Kim JH, Kim JS, Park SH. Imaging demonstration of trochlear nerve agenesis in superior oblique palsy emerging during the later life. *Clin Neurol Neurosurg* 2015;139:269-271.
- 5 Weeks CL, Hamed LM. Treatment of acute comitant esotropia in chiari I malformation. *Ophthalmology* 1999;106(12):2368-2371.
- 6 Dosunmu EO, Hatt SR, Leske DA, Hodge DO, Holmes JM. Incidence and etiology of presumed fourth cranial nerve palsy: a population-based study. *Am J Ophthalmol* 2018;185:110-114.
- 7 Helveston EM, Mora JS, Lipsky SN, Plager DA, Ellis FD, Sprunger DT, Sondhi N. Surgical treatment of superior oblique palsy. *Trans Am Ophthalmol Soc* 1996;94:315-328; discussion 328-334.
- 8 Hata M, Miyamoto K, Nakagawa S, Horii T, Yoshimura N. Horizontal deviation as diagnostic and prognostic values in isolated fourth nerve palsy. *Br J Ophthalmol* 2013;97(2):180-183.