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Case Report

Two Pediatric Patients with Acute Acquired Comitant Esotropia as the First Symptom of Brainstem Tumor: A Case Report

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Keywords
Acute acquired comitant esotropia · Hess red-green test · Magnetic resonance imaging · Brainstem tumor

Abstract
Introduction: Acute acquired comitant esotropia (AACE) is an acquired strabismus with uncrossed sudden-onset diplopia due to esodeviation, comitant esotropia without accommodation factor, or paretic eye movement. The diagnosis of AACE entails differentiation from incomitant esotropia caused by abnormalities in the central nervous system. We present 2 pediatric patients with AACE as the first symptom of brainstem tumor. ***Case Presentation:*** The 2 patients were aware of their diplopia and had no other neurological abnormalities. There were no special findings in the anterior segment, ocular media, or fundus. Esotropia with a difference of no more than 10Δ between distant and near fixations was observed. Eye movements were normal, and Hess red-green test under prism neutralization did not reveal abduction restriction. The presumed cause of AACE in both patients was excessive use of digital displays, and brain magnetic resonance imaging (MRI) was performed to confirm the absence of neurological abnormality. Using MRI, a definitive diagnosis of AACE was made based on comitant esotropia associated with diffuse median glioma and medullary pilocytic astrocytoma without abducens nerve palsy. ***Conclusion:*** Although the incidence of AACE

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caused by brainstem tumors may be low, it is necessary to perform head imaging to confirm etiology. Furthermore, Hess red-green test under prism neutralization is considered important for the differentiation of abducens nerve palsy.

Introduction

Acute acquired comitant esotropia (AACE) is an acquired strabismus with uncrossed sudden-onset diplopia due to esodeviation, comitant esotropia without accommodation factor, or paretic eye movement [1–4]. It is a rare disease that occurs in about 0.3% of children with strabismus and is classically classified based on clinical characteristics and origin into the following three types: Swan type, Burian-Franceschetti type, and Bielschowsky type [1–4]. However, in recent years, cases of adolescents and young adults who develop AACE due to long-term excessive use of digital devices have been reported and a new classification of AACE was proposed [5]. The diagnosis of AACE entails differentiation from incomitant esotropia caused by abnormalities in the central nervous system. Thus, it is important to perform diagnostic neurological imaging tests such as magnetic resonance imaging (MRI) and computed tomography. Nevertheless, cases of esotropia with comitant eye movement, including acute esotropia associated with intracranial lesions such as brain tumors, have been reported as AACE [6–8]. To establish a diagnosis of comitant or incomitant esotropia, examinations such as Hess red-green test are essential to differentiate AACE from paretic esotropia [9]. When performing Hess red-green test for patients with large-angle strabismus, it is objectively difficult to diagnose comitant or incomitant strabismus because the indicator falls out of bounds or the inner square measurements exceed the outer fields. In such cases, Hess red-green test under prism neutralization is reported to be useful [9, 10]. In this series, we present the cases of 2 pediatric patients with AACE diagnosed as brainstem tumor following neurological examination for acute-onset diplopia, and the diagnosis was confirmed using Hess red-green test under prism neutralization. The CARE Checklist has been completed by the authors for this case report, attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000534709).

Case Report

Case 1

An 11-year-old girl noticed horizontal diplopia when gazing at distant objects, and the diplopia did not improve after 2 weeks. On a typical day, her time of near-work activity was about 3 h. She was otherwise in good health with no history of head trauma, systemic illness, or psychogenic stress.

Her uncorrected visual acuity was better than 20/20 in both eyes, and her exact refraction was as follows: R (S −0.50D C + 0.50D A 170°) L (S + 0.00D C + 0.25D A 175°). Intraocular pressure was 19 mm Hg in her right eye and 17 mm Hg in her left eye. Her pupils were round and equal, light reflex was complete in both eyes, and nystagmus was not observed. Examinations of the anterior segment, ocular media, and fundus of each eye yielded normal findings. The single prism cover test revealed basic esotropia of 30 prism diopters (PD) at near fixation and 35 PD at distant fixation. The ocular rotations were full without nystagmus. Although Hess red-green test revealed inward displacement of both eyes (Fig. 1a),

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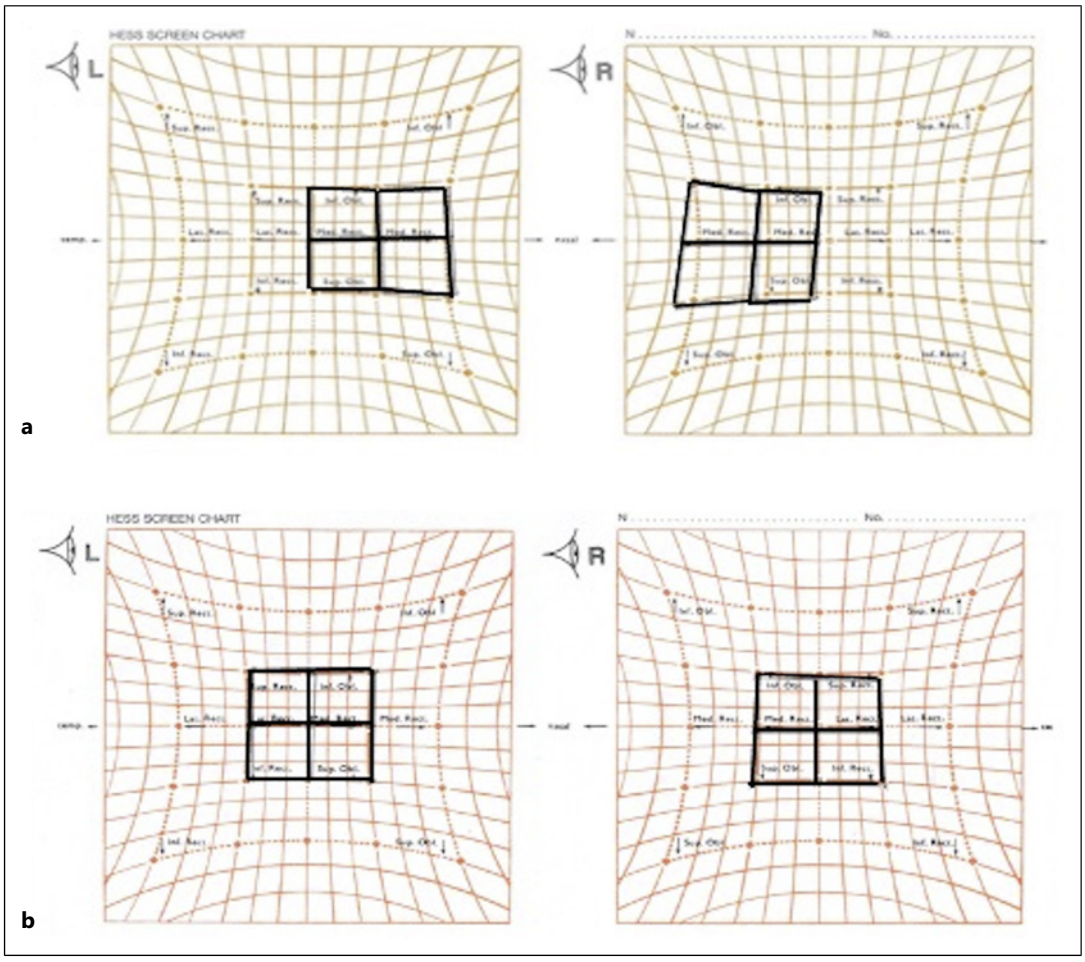

Fig. 1. Hess red-green test record in case 1. **a** Inward displacement of the dots. **b** Visual fields of equal size with the record conducted under 45Δ base-out prism neutralization.

restriction of eye movement was not observed with Hess red-green test under prism neutralization (Fig. 1b), and comitant esotropia was confirmed. As AACE was suspected, brain MRI was performed to detect intracranial disease. Brain MRI revealed a swollen internal nonuniform high-signal area on T2-weighted image (Fig. 2a). On gadolinium contrast-enhanced brain MRI, the T1-weighted image had poor contrast (Fig. 2b). Additional head computed tomography showed a lesion with unclear borders and poor contrast enhancement at the same site. There were no signs of meningism or increased intracranial pressure. These results were considered suggestive of diffuse midline glioma. She was immediately referred to neurosurgery and diagnosed with diffuse intrinsic pontine glioma. Local radiotherapy (1.8 Gy administered 30 times) was performed, and brain MRI showed tumor shrinkage after radiotherapy (Fig. 2c).

Case 2

A 4-year-old boy presented with esotropia, which was noticed by his parent and did not improve after 2 months. He was in good health with no history of head trauma or systemic illness. His uncorrected visual acuity was better than 20/20 in both eyes, and his refraction was as follows: R (S + 0.00D C + 2.00D A 180°) L (S + 0.00D C + 0.75D A 180°). Intraocular pressure was 14 mm Hg in his right eye and 15 mm Hg in his left eye. His pupils were round

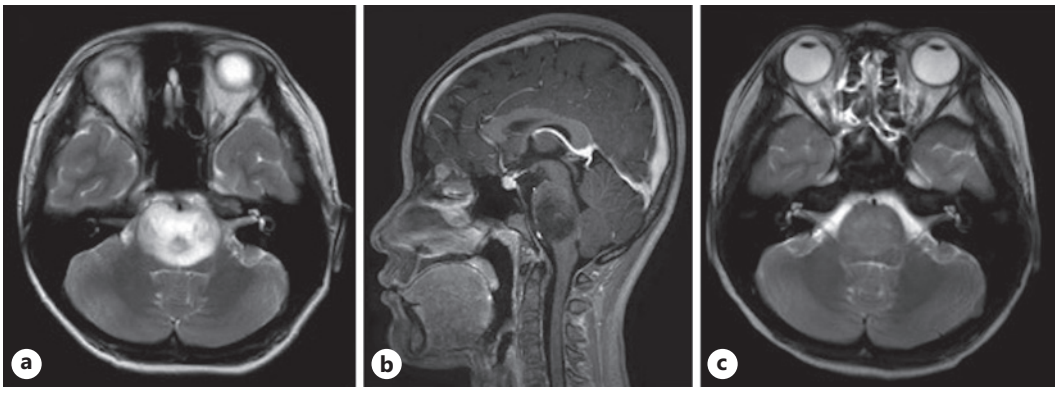

Fig. 2. Magnetic resonance image (MRI) of the brain in case 1. **a** Swollen internal nonuniform high-signal area on T2-weighted image. **b** Gadolinium (Gd) contrast-enhanced brain MRI model showing poor contrast on T1-weighted image. **c** Posttreatment MRI showing clear shrinkage of tumor.

and equal, light reflex was complete in both eyes, and nystagmus was not observed. Examinations of the anterior segment, ocular media, and fundus of each eye yielded normal results. The single prism cover test revealed basic esotropia of 40 PD at near fixation and at distant fixation. The ocular rotations were full without nystagmus. The simple Hess red-green test was not performed; however, restriction of eye movement was not observed with the Hess red-green test under prism neutralization (Fig. 3a), and comitant esotropia was confirmed.

To differentiate between AACE and other types of esotropia associated with intracranial disease, MRI was performed, and the MRI revealed a medullary tumor. Then he was diagnosed with pilocytic astrocytoma (Fig. 3b, c). Since the tumor was low-grade, he was followed up without treatment, but use of prism glasses for diplopia did not result in any changes in the esotropia. Therefore, he underwent bilateral medial rectus recession surgery approximately 1 year after his initial visit.

Discussion

We encountered 2 patients with brainstem tumors diagnosed using cranial imaging after the onset of AACE. Other neurological findings were essentially normal, and there were no abnormal findings in the anterior segment, intermediate periphery, or fundus. In both patients, esotropia was of the basic type, and eye movements were normal with no abduction restriction. Hess red-green test under prism neutralization was significantly useful in establishing a diagnosis of comitant esotropia, particularly in case 2. After diagnosing AACE, we assumed that excessive use of DDs was the main cause of AACE in the patients, and cranial imaging was performed for differential diagnosis. Unexpectedly, the imaging confirmed comitant esotropia due to brainstem tumor without abducens nerve palsy.

The characteristics of the AACE are as follows: acute or subacute onset, diplopia or hemianopsia, comitant esotropia, no abduction restriction, no change in strabismus angle, and no central nervous system disease [1–5]. Involvement of accommodative factors or paralytic esotropia is another feature considered in the differential diagnosis. Classically, AACE is classified into the following three types according to clinical features and etiology: Swan type which occurs due to uniocular occlusion or visual loss, Burian–Franceschetti type which triggered by stress, and Bielschowsky type which could be induced by low-corrected myopia [1–5]. However, in recent studies, a new classification that considers excessive use of DDs as a

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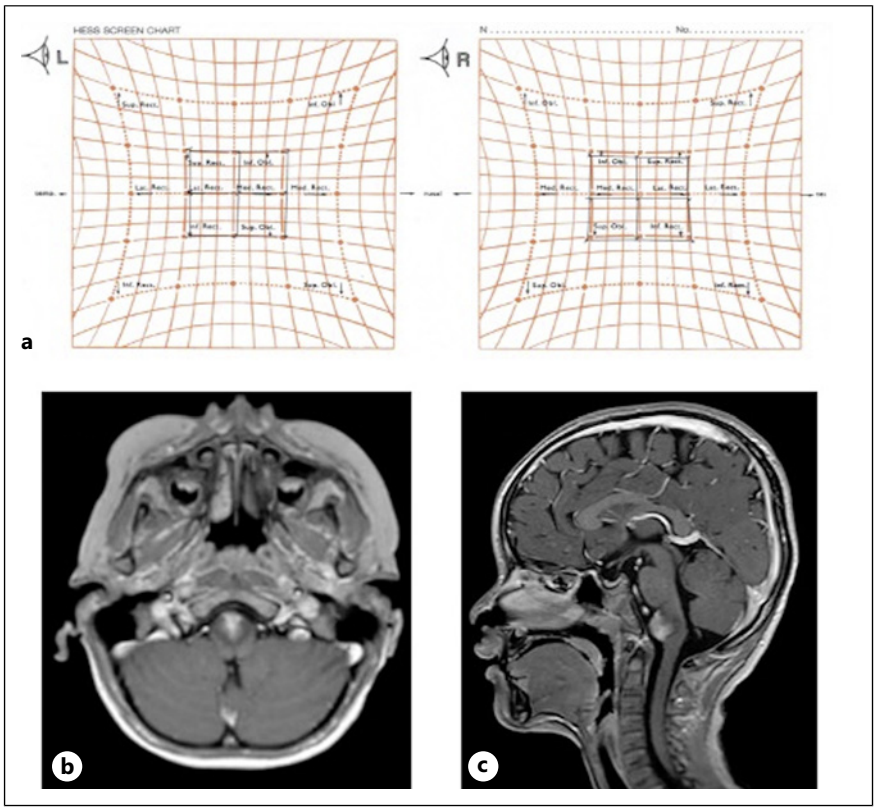

Fig. 3. Hess red-green test record and magnetic resonance image (MRI) of the brain in case 2. **a** Hess red-green test record. Showing visual fields of equal size with the record conducted under 40Δ base-out prism neutralization. Axial plane (**b**) and sagittal plane (**c**) Gd contrast-enhanced MRI of the brain. Showing a 16 mm × 18 mm mass protruding ventrally from the medulla oblongata on T1-weighted image. The border is indistinct, and the interior is heterogeneous.

cause of AACE was proposed [5, 11, 12]. Cranial imaging is recommended for the diagnosis of all four types of AACE. However, the morbidity of AACE with intracranial disease diagnosed using cranial imaging was reported to be 0–10% [7, 8, 13–15]. Buch et al. [7] reviewed 48 patients with AACE and found intracranial lesions in 3 patients (6%). The intracranial lesions found were one of hydrocephalus, diffuse intrinsic pontine glioma, or thalamic glioma in each of the 3 patients. The clinical features included hyperopia of less than +3D, diplopia at onset, and few signs of intracranial disease (in 1 of the three patients). Further, the following risk factors were reported: increased strabismus angle in distant vision, recurrence, congestive papillae, and late onset at 6 years of age or older [7]. In our series, the patients presented at the clinic with diplopia (but without congestive papillae or obvious central nervous system symptoms), refractive values less than +3D in both eyes, and were aged 11 years with late onset at 6 years of age or older. These features are similar to those reported by Buch et al. [7]. Therefore, we highly recommend that the presence of intracranial disease should always be ruled out when evaluating patients with AACE who have the risk factors reported by Buch et al. [7].

It may be difficult to differentiate between AACE and abducens nerve palsy. The most important point in the diagnosis of AACE is to establish comitancy of eye movement. Hess red-green test is recommended as one of the convenient tools for assessing ocular misalignment in patients with paralytic or restrictive strabismus [9]. However, for some patients with large-

angle strabismus, it is difficult to differentiate between normal eye movements and small restrictive eye movements based on the results of Hess red-green test because the indicator falls out of bounds or on the outer field. Koh et al. [10] reported that Hess screen test can be performed for patients with paralytic strabismus of greater than 15 PD using a Fresnel prism. We also assessed comitancy of eye movement in our 2 patients using Hess red-green test under prism neutralization.

In Hess red-green test under prism neutralization, the amount of prism is divided equally between the two eyes, and the difference in the size of the Hess chart is governed by Hering's law of equal innervation. The results of Hess red-green test under prism neutralization, with the smaller chart indicating the eye with the underacting muscle (i.e., primary deviation) and the larger chart indicating the eye with the overacting muscle (i.e., secondary deviation), establish incomitancy of eye movement. On the other hand, in cases where the test reveals that both fields are of equal size, ocular deviations are regarded as comitant strabismus [9]. If undiagnosed esotropia shows large deviations, Hess red-green test under prism neutralization can be used to differentiate between AACE and other types of paralytic esotropia. In conclusion, we examined 2 rare cases of AACE associated with brainstem tumors. To establish a diagnosis of AACE, it is necessary to carefully consider ophthalmological and neurological symptoms (e.g., pupillary reaction, eye position, and eye movement) and systemic findings, and to actively search for head images even if there is no obvious abducens nerve palsy or finding of congested papillae on fundus examination. More importantly, comitancy of eye movement should be determined, and Hess red-green test under prism neutralization may be a powerful tool for the diagnosis of AACE.

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Statement of Ethics

The Ethics Committee of Gifu University waived the need for approval of this study that involved a retrospective review of medical records. This report adhered to the tenets of the Declaration of Helsinki 1964. Verbal consent from each patient and written informed consent from their guardians were obtained for the publication of this case report and any accompanying images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Ayaka Yagasaki, Taishi Miyase, Shota Sakai, Naoyuki Ohe, Shiho Yasue, Saori Endo, and Michio Ozeki collected the clinical data. Kiyofumi Mochizuki and Teiji Yagasaki analyzed the findings and provided critical suggestions. Ayaka Yagasaki contributed to the original draft preparation. Kiyofumi Mochizuki, Hirokazu Sakaguchi, and Teiji Yagasaki reviewed and edited the manuscript. Ayaka Yagasaki, Kiyofumi Mochizuki, Hirokazu Sakaguchi, Teiji Yagasaki, Naoyuki Ohe, Shiho Yasue, Saori Endo, and Michio Ozeki agree to be accountable for all aspects of work. Ayaka Yagasaki, Kiyofumi Mochizuki, Hirokazu Sakaguchi, and Teiji Yagasaki approve the final version of the manuscript for publication.

Data Availability Statement

All data analyzed in this study are included in this article and its online supplementary material files. Further inquiries can be directed to the corresponding author.

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