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Acute acquired comitant esotropia associated with Lhermitte–Duclos disease: a case report

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Abstract

Background Acute acquired comitant esotropia caused by prolonged near work, such as the use of digital devices, has been frequently reported in recent years. However, intracranial examination is necessary even for patients with nonparalytic comitant esotropia. Lhermitte–Duclos disease is a rare tumor that grows in layers in the cerebellum. Among those with this disease, cases of esotropia have been reported due to abduction limitation of the eye, but there have been no reports of comitant esotropia. Here, we report the case of a young woman with acute acquired comitant esotropia who was found to have Lhermitte–Duclos disease.

Case presentation A 16-year-old Japanese female patient, whose ethnicity was Asian, was referred to our hospital for acute acquired comitant esotropia. Fundus examination revealed papilledema in both eyes, and magnetic resonance imaging of the head revealed a cerebellar tumor in the right cerebellum with obstructive hydrocephalus. She underwent partial tumor resection, and a histopathological diagnosis of Lhermitte–Duclos disease was obtained. However, comitant esotropia status remained unchanged, and she underwent strabismus surgery. Finally, diplopia disappeared completely.

Conclusion Neurological and intracranial imaging examinations are essential when acute acquired comitant esotropia is observed. Acute acquired comitant esotropia by Lhermitte–Duclos disease did not improve with partial tumor resection and required strabismus surgery, but good surgical results were obtained.

Keywords Acute acquired comitant esotropia, Lhermitte–Duclos disease, Papilledema, Hydrocephalus, A case report

Background

A comitant esotropia is one in which the magnitude of the esotropia is the same in all fields of gaze. Acute acquired comitant esotropia (AACE) is a nonparalytic, nonlimiting, comitant esotropia with sudden onset after the age of 6 months. Many patients experience difficulties in their daily lives due to diplopia. Although the etiology and mechanism of AACE are still unknown, Burian *et al.* classified the causes of AACE into three categories in 1958: an interruption of fusion, physical and psychological stress, and myopia [1]. Later, with the availability of modern imaging technology, Buch *et al.* classified the causes of AACE into seven categories in 2015: monocular occlusion, idiopathic, acute accommodative,

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decompensated monofixation, neurological, cyclic, and secondary [2]. Furthermore, cases of AACE due to viewing three-dimensional images and using digital devices such as smartphones have been frequently reported in recent years [3, 4]. During the coronavirus disease 2019 (COVID-19) pandemic starting in 2020, the risk of AACE was an issue due to the significant increase in device use time by children [5].

Lhermitte–Duclos disease, first described by Lhermitte and Duclos in 1920, is a localized lesion of large, abnormal neuronal cells that grows in layers in a portion of the cerebellum, and it is not known whether the lesion is neoplastic or malignant. The exact incidence of this very rare tumor is unknown, but more than 300 cases have been reported worldwide [6]. It is also known as a brain lesion associated with autosomal dominant Cowden's disease [7]. It presents with cerebellar ataxia symptoms, headache due to increased intracranial pressure, vomiting, visual disturbance, and diplopia due to focal neurological symptoms [6, 8].

Here, we report the case of a young woman with AACE who was found to have Lhermitte–Duclos disease. As far as we have searched, no association between AACE and this brain tumor has been reported. Thus, we report this case here as a new complication of Lhermitte–Duclos disease.

Case presentation

A 16-year-old Japanese female patient, whose ethnicity was Asian, was referred to our hospital for acute esotropia 3 weeks after a sudden onset of diplopia. She had been followed up for hydrocephalus since she was 10 months old, but the follow-up was stopped before she started elementary school. Since no medical records from that time were available, the details are unknown, as is her relationship with Lhermitte–Duclos disease, which would later be identified. There was no other medical history or family history of note. At the initial examination, the best-corrected visual acuity was 20/33 in right eye (OD) and 20/20 in left eye (OS), and the equivalent spherical power was −0.25 D OD and −0.75 D OS. Slit-lamp microscopy revealed clear corneas, anterior chambers, lenses, and vitreous cavities in both eyes. Fundus examination revealed no abnormalities in the retina, including the macula, but papilledema was observed in both eyes. Both eyes had prompt light reflexes. Mild upward gaze palsy and horizontal nystagmus during bilateral viewing were observed, but horizontal eye movements were not limited. In the alternate prism cover test with refractive correction, the patient had 35–40 prism diopter (PD) comitant esotropia both near and distance, with alternating fixation, and was aware of uncrossed diplopia. No abnormal head position was observed. Antiacetylcholine receptor antibody, antithyroid stimulating hormone receptor antibody, antithyroid peroxidase antibody, and antithyroglobulin antibody were all negative. Magnetic resonance imaging (MRI) of the head revealed a cerebellar tumor in the right cerebellum that compressed the brainstem from behind (Fig. 1a, b). Stenosis of the midbrain aqueduct and enlargement of the cerebral ventricles were observed, and Lhermitte–Duclos disease, complicated with obstructive hydrocephalus, was subsequently suspected. Based on these findings, a diagnosis of AACE caused by intracranial disease was made. The neurological examination revealed mild ataxia of the right upper extremity, but no other apparent cranial nerve abnormalities or motor paralysis of the extremities. Partial tumor resection was performed to treat hydrocephalus

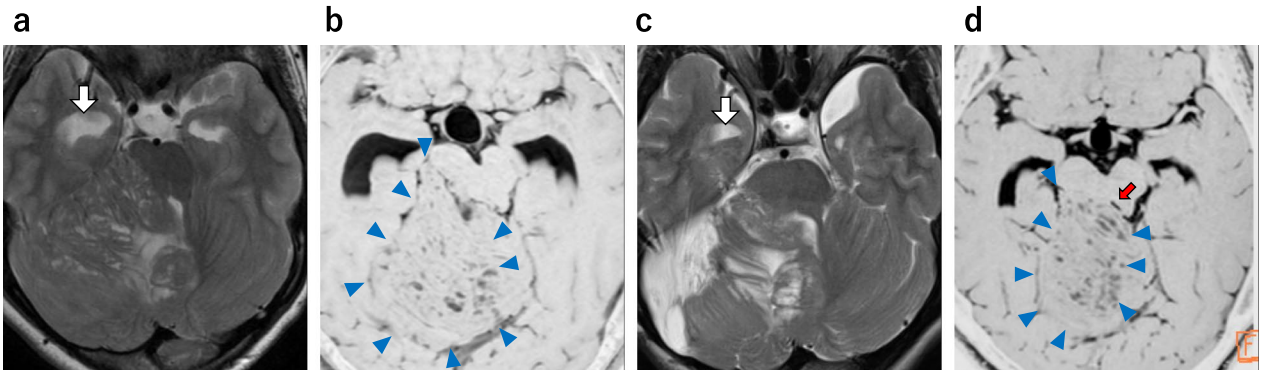

Fig. 1 Magnetic resonance images before craniotomy (**a, b**) and 11 months after surgery (**c, d**). **a** T2-weighted image. A cerebellar tumor was observed centered in the right cerebellum, compressing the brainstem posteriorly. The inferior horn of the lateral ventricle is enlarged (white arrow). **b** Constructive interference in the steady state image. The location of the tumor was clearly visible (blue arrowheads). **c** The tumor was partially removed, and compression to the brainstem improved. The inferior horn of the lateral ventricle also shrank (white arrow), and the hydrocephalus improved. **d** Constructive interference in the steady state image. Compression of the brainstem by tumor (blue arrowheads) decreased, and stenosis of the midbrain aqueduct improved (red arrow)

and decompress the brainstem. Pathological examination revealed a marked decrease in the granular cell layer and thickening of the molecular layer (Fig. 2), and the diagnosis of Lhermitte–Duclos disease was confirmed. MRI showed decreased brainstem compression and shrinkage of the ventricles (Fig. 1c, d).

At 11 months postsurgery, there was no vision loss or improvement in the papilledema in either eye. The alternate prism cover test showed residual esotropia, with 40Δ at near and 35Δ at distance. The only ocular motor limitation was upward gaze palsy similar to the preoperative level, with no horizontal motor limitation (Fig. 3a, b). Because the strabismus angle was stable, strabismus surgery was performed 14 months after the head surgery. Target correction was determined by the prism adaptation test, and lateral rectus resection and medial rectus recession were performed on the right eye. Six months after strabismus surgery, ocular alignment was 8 PD and 6 PD of esophoria at near and distance, respectively, and diplopia disappeared completely. The upward gaze palsy was unchanged before and after strabismus surgery (Fig. 3c). The stereoacuity was 200 s of arc in the Titmus Stereo Test. Neurologically, mild ataxia of the right upper extremity did not improve. MRI showed no change in the residual tumor, and the cerebral ventricles tended to shrink.

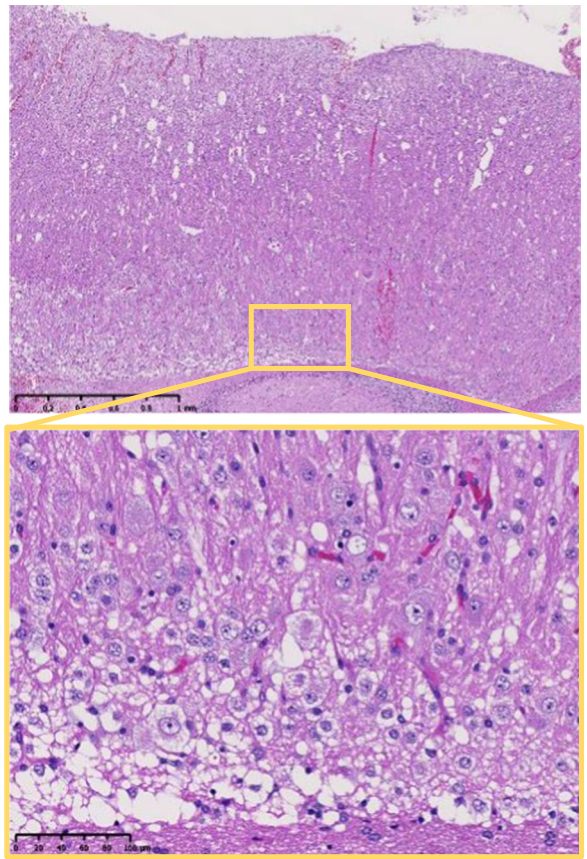

Fig. 2 Histopathological image of the excised cerebellar tumor. Histopathological analysis of the cerebellar tumor revealed a marked decrease in the granular cell layer and a thickening of the molecular layer. A pathological diagnosis of Lhermitte–Duclos disease was made

Discussion

In Lhermitte–Duclos disease, MRI T2-weighted images show high-signal and characteristic tiger-striping structure [7, 9], making it possible to suspect this disease even before surgery. Histopathologically, tumors are characterized by a marked decrease in the granular cell layer, thickening of the molecular layer, and increased neuronal cell proliferation [7, 10]. Treatment generally involves craniotomy with partial tumor resection and does not require total resection [7]. A systematic review reported the incidence of diplopia and strabismus in Lhermitte–Duclos disease to be 9.6%, usually involving eye abduction secondary to abducens cranial nerve palsy and elevated intracranial pressure [6]. However, there has been no detailed description of strabismus to date, and this case is the first report of a nonparalytic AACE associated with this rare tumor.

The current case falls under Buch's classification of neurological AACE arising from intracranial disease [2]. AACE associated with the central nervous system due to intracranial disease has been reported for a long time. Its differential diagnosis is essential because it can be clinically fatal. Causative diseases include Chiari malformation type 1 [11], cerebellar medulloblastoma [12], cerebellar astrocytoma [13–15], diffuse intrinsic pontine glioma [16], and herpes encephalitis [2]. Liu *et al.* studied 30 cases of acquired esodeviation caused by brain tumors, meningitis, or pseudotumor cerebri [17]. They reported that 40% were comitant and 60% were incomitant, with the comitant type being common in children. In recent years, the incidence of AACE caused by prolonged near work has been increasing [18]; therefore, AACE is often easily judged to be caused by near work. However, it is necessary to confirm the presence or absence of neurological abnormalities via interviews and general examinations and to perform fundus examinations to detect papilledema. It is controversial whether intracranial imaging should be performed when those tests are not abnormal. Buch *et al.* investigated risk factors for intracranial disease in patients with AACE . The authors found that risk factors included greater esodeviation at distance than near, the presence of neurological findings and symptoms, recurrent AACE, and onset at age 7 or older [2]. On the other hand, in a report investigating 20 children presenting with AACE and otherwise normal

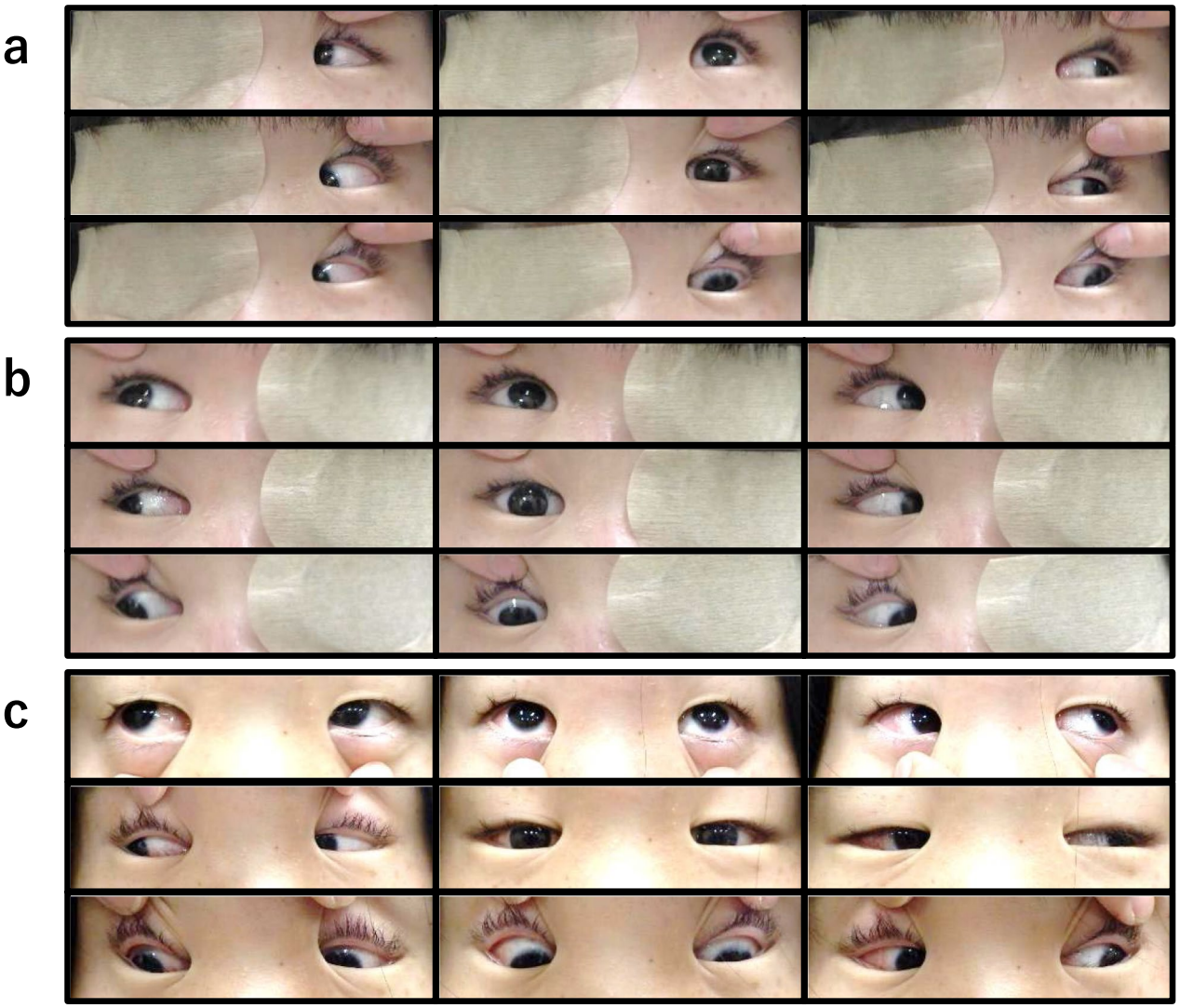

Fig. 3 Eye movements in nine directions. **a, b** The ductions in all directions of gaze after removal of the cerebellar tumor. The patient had upward gaze palsy with no other limitations. There were no changes compared with those in the preoperative period. **c** The versions in all directions of gaze after strabismus surgery. Upward gaze palsy was still present, and no other limitations were observed

neurological and ophthalmological evaluations, no children had intracranial disease as a cause. Nevertheless, the authors noted that they try to perform intracranial imaging studies [19]. This is because intracranial disease may be found, although it is rare, and neurological symptoms may appear late. Zweifach reported the case of a 10-year-old boy who developed sudden esotropia, had normal neurological and neuroradiologic evaluations, underwent strabismus surgery, and 18 months later developed clinical signs that led to a diagnosis of cerebellar medulloblastoma [12]. In addition, some of these diseases are serious and can be fatal [16]. Therefore, we also believe that computed tomography or MRI is essential in patients with AACE.

Table 1 compares the present case with previously reported cases of AACE caused by cerebellar tumors [12–15]. Papilledema was present in four of the six patients but not in two, suggesting hydrocephalus was not involved in developing AACE. Since all the tumor sites rested on the cerebellar vermis, compression, and/or damage to the cerebellar vermis may be responsible for the development of AACE. However, the lack of remission of strabismus after tumor treatment suggested that the neurons had already undergone irreversible changes.

In the present case, mild upward gaze palsy was also observed. Upper gaze disorders have been reported to occur with bilateral rostral interstitial nucleus of the medial longitudinal fasciculus (riMLF) disorders,

Table 1 Comparison of this case with previously reported acquired comitant esotropia caused by a cerebellar tumor

	Age (years), sex	Papilledema [yes (y)/no (n)]	Cerebellar tumor	Site of origin	Strabismus after tumor treatment
Zweifach [12]	10, male	y	Medulloblastoma	Cerebellum	N/A
Musazadeh [13]	3, female	n	Pilocyte astrocytoma	Upper dorsal vermis	No changes
Lee [14]	3, male	y	Pilocyte astrocytoma	The midline of the cerebellum	No changes
Dikici [15]	5, female	y	Astrocytoma	Cerebellar midline	No changes
Present case	16, female	y	Lhermitte–Duclos disease	Right cerebellar hemisphere	No changes

posterior commissure (PC) disorders, unilateral riMLF disorders, and disorders after the connecting fibers cross the contralateral side at the PC [20]. In the present case, the tumor compressed the brainstem from the right dorsal side at the level of the midbrain superior colliculus, suggesting that her upper gaze disorder was caused by unilateral riMLF disorder or disorder after the crossing of the connecting fibers at the PC.

Conclusion

The patient with Lhermitte–Duclos disease was found to not only have papilledema but also had other neurological abnormalities, such as upward gaze palsy and ataxia of the upper extremities. When AACE is observed, neurological and intracranial imaging examinations are essential. AACE by Lhermitte–Duclos disease did not improve with partial tumor resection and required strabismus surgery, but good surgical results were obtained.

Abbreviations

AACE Acute acquired comitant esotropia
MRI Magnetic resonance imaging
PC Posterior commissure
PD Prism diopter
riMLF Rostral interstitial nucleus of medial longitudinal fasciculus

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Not applicable.

Author contributions

JO and RA substantially contributed to the report conceptualization, data analysis and interpretation of ophthalmology-related content and completed the draft manuscript. HM contributed to data analysis and interpretation of neurosurgery-related content. HS and TM contributed to data analysis and interpretation of pathology-related content. All authors besides JO and RA critically reviewed and revised the manuscript draft and approved the final version for submission.

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Availability of data and materials

The datasets used in this case report are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate

Ethical approval was not required in accordance with local guidelines of the Institutional Review Board at the Hokkaido University Hospital.

Consent for publication

Written informed consent was obtained from the patient's legal guardian for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests

The authors declare that they have no competing interests.

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