Epilepsy & Behavior Reports 26 (2024) 100674

Contents lists available at ScienceDirect

Epilepsy & Behavior Reports

journal homepage: www.elsevier.com/locate/ebcr

MRI detection of mild malformation of cortical development with oligodendroglial hyperplasia (MOGHE) on T1WI-CHESS

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ARTICLE INFO

Keywords:
MOGHE
MRI
CHESS
epilepsy

ABSTRACT

Mild malformation of cortical development with oligodendroglial hyperplasia (MOGHE) is a recently proposed epileptogenic entity that is difficult to detect on MRI. We present a case of MOGHE that was successfully detected on T1WI-chemical shift-selective saturation (CHESS) MRI. The clinical presentation, MRI including T1WI-CHESS, functional images, and pathology findings of a 14-year-old Japanese girl diagnosed with MOGHE are described. T1WI-CHESS revealed an abnormal high signal along the affected lesion, whereas the findings shown by the other MR sequences were less obvious; interictal fluorodeoxyglucose-positron emission tomography indicated slightly decreased accumulation in the lesion, and subtraction ictal single photon emission computed tomography co-registered to MRI showed an increased blood flow. Together these observations suggest that T1WI-CHESS may be a useful MR sequence for detecting the lesions in patients with MOGHE.

Introduction

Mild malformation of cortical development with oligodendroglial hyperplasia (MOGHE) is a histopathologic entity characterized by blurred gray/white matter boundaries due to an increased density of oligodendroglia in subcortical white matter [1]. MOGHE was proposed as a new category in the International League Against Epilepsy (ILAE) consensus classification of focal cortical dysplasia (FCD) in 2022 [1,2]. MOGHE lesions are mainly in the frontal lobes, and *SLC35A2* mutation has been identified [2,3]. Chemical shift selective (CHESS) is the fat-suppression pulse technique that is the most frequently used clinically. T1WI-CHESS examinations of the brain have been reported to detect lesions as abnormally high signals such as bottom of sulcus and transmantle signs in FCD type IIb, and the cortical tubers and radial migration lines in the tuberous sclerosis complex. We report the case details of a patient with MOGHE that was also well detected by T1WI-CHESS on MRI.

Case report

A 14-year-old Japanese girl who had developed epileptic spasms at the age of 2 months was diagnosed with West syndrome at 5 months and was controlled with anti-seizure medications. At age 3, she developed startle seizures in which she raised both upper limbs in response to unexpected sounds, and at age 6, she had daily seizures in which she fell over at the sound, resulting in an intractable course. After the age of 10 years, atonic, behavior arrest, and asymmetrical tonic seizures, occurred daily. Her intelligence quotient (IQ) was 88 on the Wechsler Intelligence Scale for Children −IV test, which was within the normal range at age 7, but her IQ had declined to 70 at age 10 and to 53 at age 13. At age 11, her electroencephalogram showed a series of spike-and-wave discharges in the right frontal region, which led to the diagnosis of right frontal lobe

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https://doi.org/10.1016/j.ebr.2024.100674
Received 16 February 2024; Received in revised form 7 May 2024; Accepted 7 May 2024
Available online 8 May 2024

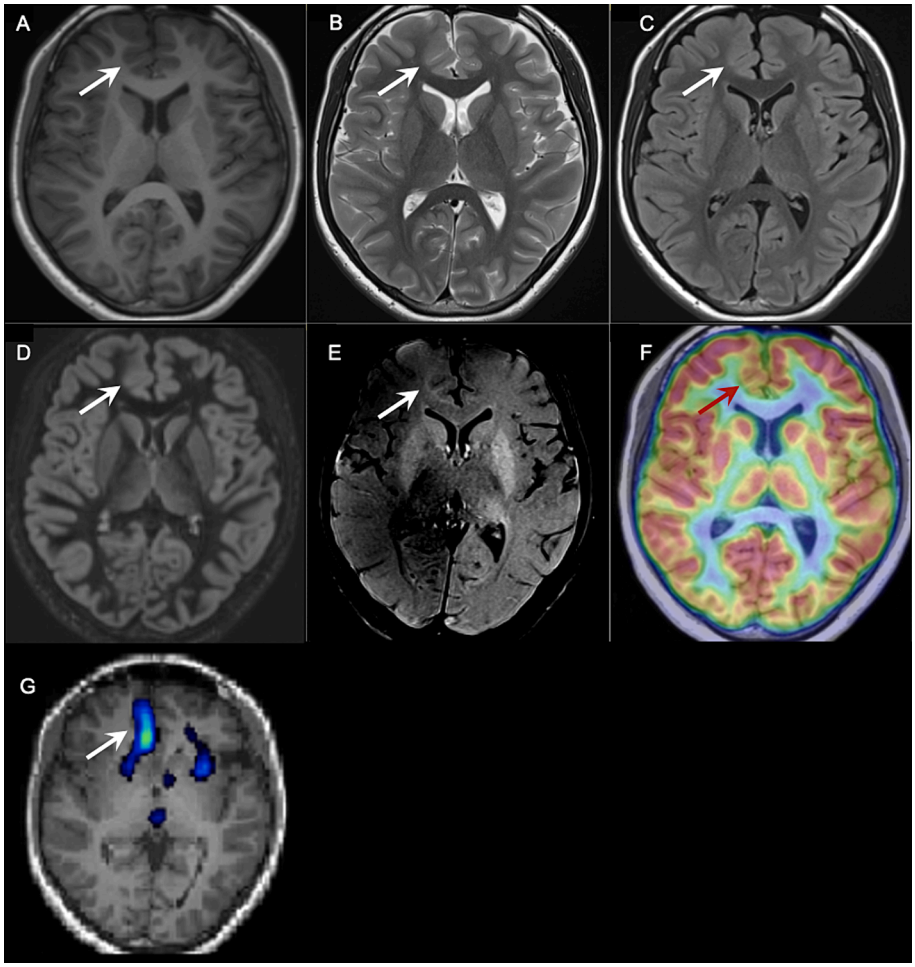

Fig. 1. (A) T1WI, (B) T2WI, (C) FLAIR, (D) DIR, (E) T1WI-CHESS, (F) interictal FDG-PET MRI fusion image, and (G) subtraction ictal SPECT co-registered to MRI. The right medial frontal lobe showed subtle cortical thickening and blurring of gray/white matter boundaries on T1WI /T2WI/FLAIR/DIR, without signal abnormality (*arrows*, *A–D*). However, T1WI-CHESS detected a subcortical linear high signal along the affected gyri (*arrows*, *E*). Interictal FDG-PET showed a slightly decreased accumulation consistent with the lesion, compared to the opposite cortex (*arrow*, *F*). Subtraction ictal SPECT co-registered to MRI indicated increased blood flow in the lesion (*arrows*, *G*). CHESS: chemical shift selective, DIR: double inversion recovery, FDG: fluorodeoxyglucose, FLAIR: fluid-attenuated inversion recovery, PET: positron emission tomography, SPECT: single photon emission computed tomography.

epilepsy. No lesion had been evident on previous MRI, but when the patient was 14 years old, 3 T MRI and functional images were acquired to identify the epileptogenic lesion.

T1WI/T2WI/fluid-attenuated inversion recovery (FLAIR)/double inversion recovery (DIR) imaging demonstrated subtle cortical thickening and blurring of gray/white matter boundaries in the right medial frontal lobe, a weak subcortical high signal in the right superior frontal gyrus, and decreased white matter volume in the right central posterior gyrus (Fig. 1A–D, Fig. 2A–D). However, T1WI-CHESS detected a subcortical linear high signal along the affected medial frontal and postcentral gyri (Fig. 1E, Fig. 2E). In some parts of the affected areas, a slight decrease in accumulation on interictal fluorodeoxyglucose-positron emission tomography (FDG-PET) and increased blood flow on subtraction ictal single photon emission computed tomography (SPECT) co-registered to MRI were observed (Fig. 1F, G). In the ictal-phase SPECT, the radionuclide was injected 10 s after the tonic seizure.

The patient's right medial frontal lobe, including the right superior frontal gyrus and anterior cingulate gyrus, was extensively resected. Pathologically, the gray/white matter boundaries were blurred with increased oligodendroglia and heterotopic neurons subjacent to the white matter. These areas revealed increased densities of subcortical oligodendroglia with oligodendrocyte transcription factor 2 (Olig2) staining, patchy loss of myelin basic protein (MBP) staining, and increased subcortical neurons with neuronal nuclear protein (NeuN) staining. The MIB-1 positivity rate was low (Fig. 3). The diagnosis was MOGHE. *SLC35A2* mutation, c.359-360del TC was confirmed by a genetic analysis of a surgical specimen from the patient. The patient had no seizures after surgery.

Discussion

We presented a case of MOGHE showing high signal along the lesion on T1WI-CHESS, whereas the signal shown by the other MR sequences was less obvious. This report is the first to demonstrate characteristic findings of MOGHE visualized by T1WI-CHESS.

Regarding MRI findings in MOGHE, cortical thickening and gray/white matter blurring in affected areas corresponding to the histologic pattern of oligodendroglial hyperplasia were first reported in 2009 [4]. After MOGHE was proposed in 2017, two MRI subtypes were reported [1,5]. Subtype I has shown a laminar T2WI/FLAIR high signal at the gray/white matter junction in children < 5 years old, and subtype II has shown gray/white matter blurring due to a reduced subcortical T2WI/FLAIR high signal in older children and young adults. These MRI findings seem to shift from subtype I to subtype II with age, possibly related to myelination [5]. The MRI in our patient's case was suggestive of subtype II based on the weak T2WI/FLAIR high signal. Even with subtype II, T1WI-CHESS would likely show a clearer high signal than T2WI/FLAIR.

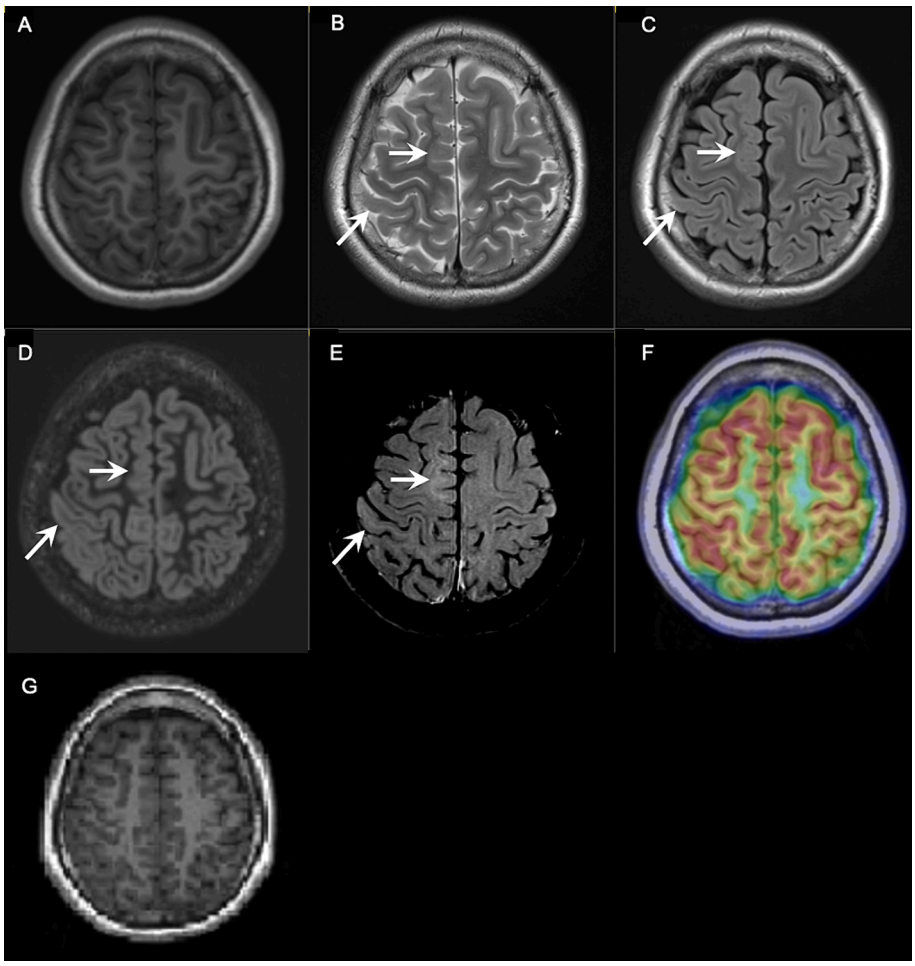

Fig. 2. (A) T1WI, (B) T2WI, (C) FLAIR, (D) DIR, (E) T1WI-CHESS, (F) interictal FDG-PET MRI fusion image, and (G) subtraction ictal SPECT co-registered to MRI. T2WI/FLAIR/DIR showed minimally blurred gray/white matter boundaries with a weak subcortical high signal in the right superior frontal gyrus and decreased white matter volume in the right central posterior gyrus *(arrows, B–D)*. T1WI-CHESS detected a high signal in the right superior frontal gyrus to the anterior cingulate gyrus and in the right central posterior gyrus (*arrows, E*). In these lesions, no abnormalities were detected on interictal FDG-PET and subtraction ictal SPECT co-registered to MRI were detected (F, G).

T1WI-CHESS clarifies abnormal high-signal areas in lesions based on signal suppression by the magnetization transfer effect and a fat suppression of myelin fat-rich normal white matter. The usefulness of high signals on T1WI-CHESS in brain lesions has been reported in two papers from our institution; the Kusama et al. report concerned the bottom of sulcus and transmantle signs in FCD type IIb [6], and the Fujii et al. paper described a high signal in the cortical tubers and radial migration lines in the tuberous sclerosis complex [7]. Both groups observed a very strong high signal on T1WI-CHESS compared to the present case and described numerous balloon cells in the pathology findings. In our present patient's case, the T1WI-CHESS signal was relatively weak and no balloon cells were detected pathologically, but oligodendroglial proliferation was characteristic. Thus, the very strong high signal on T1WI-CHESS is likely attributable to the massive balloon cells, whereas, the high signal was weak and linearly extended along the subcortical areas, which is probably associated with the oligodendroglial proliferation. These MRI findings may be useful for the differential diagnosis. An additional advantage is that T1WI-CHESS can point out lesions of MOGHE that are difficult to by conventional imaging modalities such as T2WI, T1WI, FLAIR, and DIR.

MOGHE is diagnosed pathologically, but is considered when MRI shows extensive lesions, mainly in the frontal lobe with blurred gray and white matter boundaries. Even if the T2WI/FLAIR high signal is weak, a high signal on T1WI-CHESS is characteristic of MOGHE and may be useful in investigations of MOGHE.

Funding

The study was supported in part by a grant from the Japan Agency for Medical Research and Development (AMED), no. JP23wm0425005.

Ethical statement

The authors declare that this report was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

CRediT authorship contribution statement

Elly Arizono: Writing – original draft. **Zen-ichi Tanei:** Writing – review & editing. **Keiya Iijima:** Writing – review & editing. **Yukio Kimura:** Writing – review & editing. **Yoko Shigemoto:** Writing – review & editing. **Hiroyuki Maki:** Writing – review & editing. **Midori Kusama:** Writing – review & editing. **Kumiko Murayama:** Writing – review & editing. **Masaki Iwasaki:** Writing – review & editing. **Takashi Saito:** Writing – review & editing. **Yuko Saito:** Writing – review & editing. **Kazuhiro Saito:** Supervision. **Noriko Sato:** Writing – review & editing, Supervision.

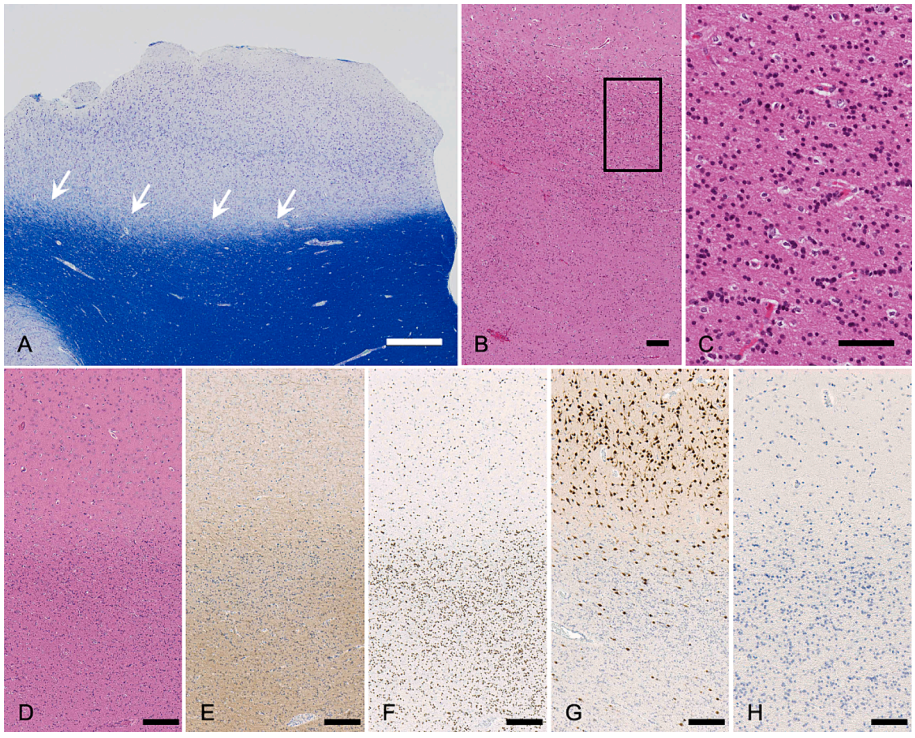

Fig. 3. The histopathological specimens shown here are the parts of the lesion indicated in Fig. 2. (A) Klüver-Barrera (KB), (B) hematoxylin and eosin-stained (H&E), (C) enlarged image of the marked area in panel B, (D) H&E, (E) myelin basic protein (MBP), (F) oligodendrocyte transcription factor 2 (Olig2), (G) neuronal nuclear protein (NeuN), and (H) MIB-1 in the anterior of right superior frontal gyrus. KB staining showed partial blurring of gray/white matter boundaries (*arrows*, *A*). B: H&E staining showed a focal increase in cell density at the gray/white matter boundaries. C: High-power magnification of the H&E image showed increased oligodendroglia and neurons. E–G: Immunohistochemical staining of the same area as the gray/white matter boundaries (D); MBP staining showed patchy loss (E), and Olig2- and NeuN-positive cells showed a heterogeneous increase (F, G). The MIB-1 positivity rate was low. Scale bar = 500 μm (A), 100 μm (B, H), 50 μm (C), and 200 μm (D–G).

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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