

Chiari Type 1 Malformation-induced Intracranial Hypertension with Diffuse Brain Edema Treated with Foramen Magnum Decompression: A Case Report

Toshiki Fukuoka,¹ Yusuke Nishimura,¹ Masahito Hara,² Shoichi Haimoto,¹ Kaoru Eguchi,¹ Satoshi Yoshikawa,¹ Toshihiko Wakabayashi,¹ and Howard J. Ginsberg³

Chiari type 1 malformation (CM1) rarely causes papilloedema, which is indicative of high intracranial pressure with or without ventricular dilatation. Furthermore, concomitant brain parenchymal abnormalities have not been reported to date. In this paper, the authors report on a young woman of CM1-induced intracranial hypertension (ICH) with diffuse brain edema with a focus on venous sinus assessment, and discuss the surgical strategy. A 24-year-old woman presented to Nagoya University Hospital complaining of 4-year history of severe occipital headache and blurry vision with slowly progressive worsening. Head and whole spine MRI showed a CM1 with diffuse white matter hyperintensities (WMH) on T2-weighted imaging and narrowed brain sulci without hydrocephalus. Lumbar puncture revealed extremely high opening pressure. Detailed blood examination and other radiographical imaging studies denied the presence of tumor, collagen disease, encephalitis and other entities. Head magnetic resonance venography and angiography demonstrated severe transverse sinus stenosis on both sides. Foramen magnum decompression was performed to alleviate the ICH by restoration of cerebrospinal fluid (CSF) stagnation at the foramen magnum with successful outcome. The patient completely recovered from preoperative symptoms immediately after surgery. The diffuse WMH and narrowing brain sulci have been resolving. The most feasible explanation for this complicated pathophysiology was ICH induced by CM1 led to transverse sinus collapse, resulting in diffuse WMH as a result of venous hypertension. This case report is the first illustration of successful surgical treatment of CM1 with diffuse brain edema with a focus on venous sinus assessment.

Keywords: Chiari type 1 malformation, intracranial hypertension, diffuse brain edema, foramen magnum decompression, venous sinus stenosis

Introduction

Chiari malformation type 1 (CM1) is first described by Hans Chiari in 1891¹⁾ and defined as the downward displacement of the cerebellar tonsils and inferior cerebellar lobules through the foramen magnum into the upper cervical spinal canal. CM1 could cause headache, visual disturbance, hydrocephalus, syringomyelia, lower cranial nerve palsies²⁾ and rarely papilloedema,³⁻⁵⁾ which is indicative of high intracranial pressure. There are some reports to describe the relationship between CM1 and intracranial hypertension (ICH).⁶⁻⁹⁾

Diffuse brain edema, described as white matter hyperintensities (WMH) and narrowing of brain sulci on T2-weighted magnetic resonance imaging (T2WI-MRI), is correlated with venous hypertension, which could lead to ICH. This case report describes an extremely rare case of CM1 with diffuse brain edema provoked by combination of CM1-induced ICH and venous sinus stenosis (VSSs), which was successfully treated with foramen magnum decompression (FMD). There are no previous reports referring to the association between diffuse brain edema and CM1. The pathophysiology is discussed with a focus on the assessment of venous outflow.

Case Report

A 24-year-old female without any past medical history had suffered from slowly-progressive blurring vision and severe occipital headache over a four-year period. She presented to an ophthalmologist, where the ophthalmic assessment revealed bilateral papilloedema, elevated intraocular pressure and inferior nasal quadrantanopia. Visual acuity was 1.0 on both sides and non-contact tonometry revealed 23.3 mmHg on the right side and 23.0 mmHg on the left side. She was referred to our neurosurgical clinic with presumptive diagnosis of ICH. She was otherwise neurologically intact. Head T2WI-MRI showed CM1 and narrowed brain sulci as well as high intensity areas of bilateral cerebral white matter, basal ganglia and thalamus without ventricular dilatation (Figs. 1a–1d). Diffusion weighted MRI (DWI) showed no abnormal findings and apparent diffusion coefficient (ADC) map demonstrated high intensity area corresponding with high intensity area on T2WI-MRI (Figs. 1e–1f). At first, the differential diagnosis was considered including brain tumors, vascular diseases, inflammatory diseases, demyelinating

¹Department of Neurosurgery, Nagoya University, Nagoya, Japan

²Department of Neurosurgery, Inazawa Municipal Hospital, Inazawa Japan

³Division of Neurosurgery, St. Michael's Hospital, University of Toronto, Toronto, Canada

Received: November 16, 2016; Accepted: March 28, 2017
Online September 12, 2017

Copyright© 2017 by The Japan Neurosurgical Society
This work is licensed under a Creative Commons Attribution-NonCommercial-NoDerivatives International License.

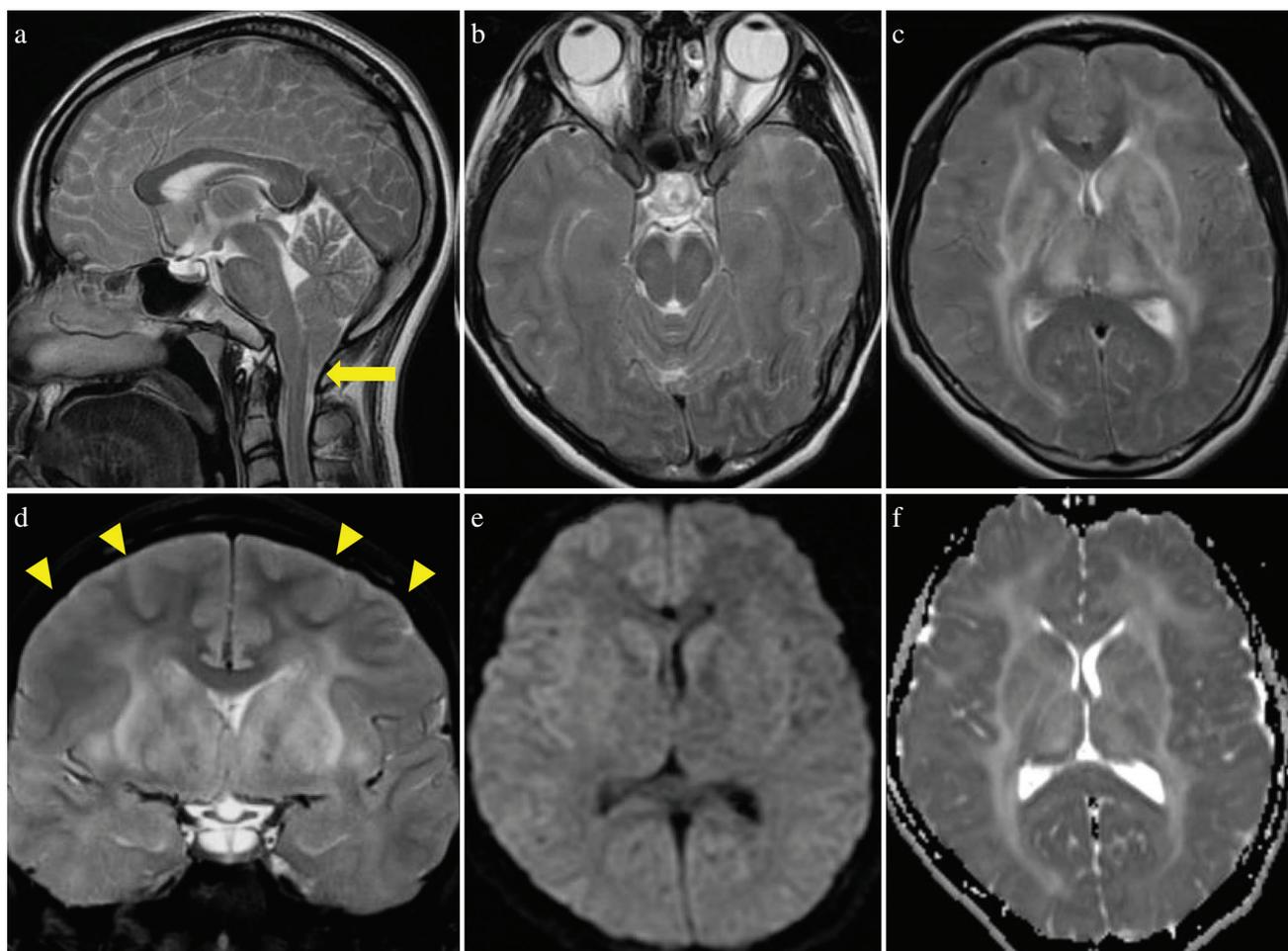


Fig. 1 Preoperative cervical spine sagittal image of T2WI-MRI (a) showed downward displacement of cerebellar tonsils by 13 mm below the base of the foramen magnum without evidence of syringomyelia. Preoperative head T2WI-MRI (b and c; axial image, d; coronal image) showed brain edema, narrowed brain sulci (d; arrowheads) and diffuse white matter hypertensities affecting bilateral cerebral white matter, basal ganglia and thalamus without ventricular dilatation. Supratentorial brain herniation was not observed (b and d). Diffusion-weighted image (DWI) demonstrated no abnormal findings (e) and apparent diffusion coefficient (ADC) map (f) showed high intensity area corresponding with T2WI.

disease and collagen disease, which could show diffuse WMH. Head MRI with gadolinium enhancement, positron emission tomography (PET) did not show any tumor presence. Magnetic resonance spectroscopy (MRS) indicated no abnormalities (Fig. 2a). The presence of a tumor was ruled out and we referred this patient to neurologists to investigate other possible pathologies. They performed detailed blood exams and cerebrospinal fluid (CSF) study, which were all within normal range except for markedly elevated initial CSF pressure (32 cm H₂O) during lumbar puncture. Magnetic resonance venography (MRV) and angiography disclosed significant bilateral transverse-sigmoid sinus junction stenosis (Figs. 2b and 2c). Cine flow MRI showed severely stagnant CSF flow around foramen magnum. There were not any physical and radiological indications of idiopathic intracranial hypertension (IIH) except for tonsillar herniation and VSs. At this stage, we thought ICH was attributable to CM1 and resulted in secondary VSs. We formulated a hypothesis that each element could become a contributing factor to one another, which eventually ended up with diffuse brain edema

(Fig. 3). As collateral development of other venous sinuses at foramen magnum which could potentially interfere with duraplasty was absent, we decided to proceed to duraplasty following occipital craniotomy and C1 laminectomy. She successfully underwent FMD combined with C1 laminectomy and duraplasty. Adequate 180-degree decompression of the foramen magnum was achieved (Fig. 4a) with arachnoid membrane left intact to prevent postoperative arachnoid adhesion and CSF leak. The patient made an excellent recovery without any detectable subjective neurological complications. Her headache and blurring vision rapidly and dramatically improved immediately after surgery. On postoperative head MRI, foramen magnum was successfully decompressed (Fig. 4b). Postoperative MRV and cine flow MRI showed a marked improvement of venous sinus flow and CSF circulation around the foramen magnum (Figs. 4c and 4d). Diffuse brain edema and narrowed brain sulci were significantly reduced with time on T2WI-MRI and ADC map (Figs. 5a–5i), while there was still a certain degree of residual white matter change. At one-year postoperative

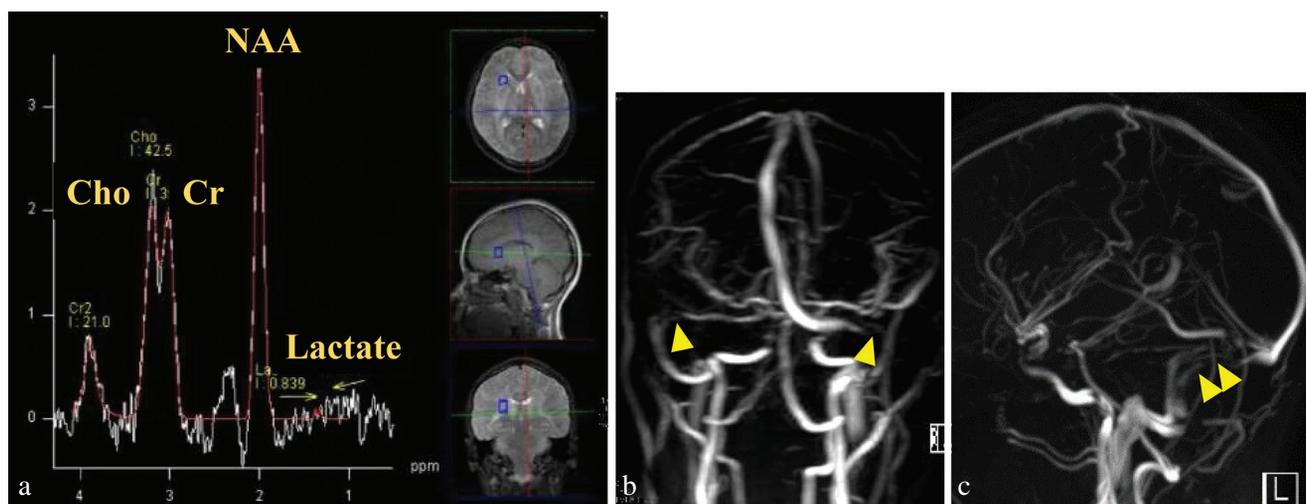


Fig. 2 Preoperative magnetic resonance spectroscopy (MRS) indicated no abnormalities (a). MRV (magnetic resonance venography) (b; AP view and c; lateral view) disclosed significant bilateral transverse-sigmoid sinus junction stenosis (arrow heads).

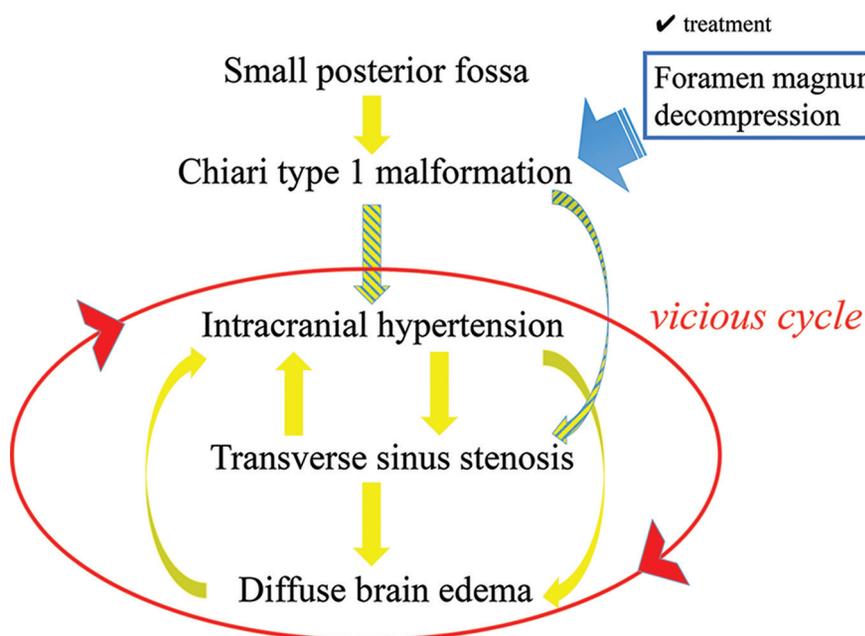


Fig. 3 The hypothesized pathophysiology in the present case. We formulated a hypothesis that each element could become a contributing factor to one another, which eventually ended up with diffuse brain edema.

follow-up, her subjective symptoms completely resolved, and bilateral papilledema, elevated intraocular pressure and inferior nasal quadrantanopia are steadily improving in accordance with ongoing recovery of residual white matter change.

Discussion

The unique radiological findings in the present CM1 case was diffuse brain edema. This diffuse brain edema proved to be vascular edema characterized by no abnormality on DWI and high intensity areas on ADC map. Differential diagnosis has to include various kinds of entities

such as: drug-associated progressive multifocal leukoencephalopathy,¹⁰ vasculitis,¹¹ acute disseminated encephalomyelitis, CADASIL^{12,13}/CARASIL,¹⁴ malignant tumors like gliomatosis cerebri, demyelinating disease like multiple sclerosis, collagen disease such as systemic lupus erythematosus (SLE)¹⁵⁻¹⁷ and Sjogren's disease,¹⁸ metabolic abnormality like adrenoleukodystrophy,¹⁹ Alexander's disease²⁰ and VSs.^{15,21} Some types of autoimmune or virus-induced encephalitis could demonstrate spontaneous resolution without specific treatment. However, contrary to that kind of encephalitis cases, the patient had diffuse and bilateral brain edema without focal neurological symptoms,

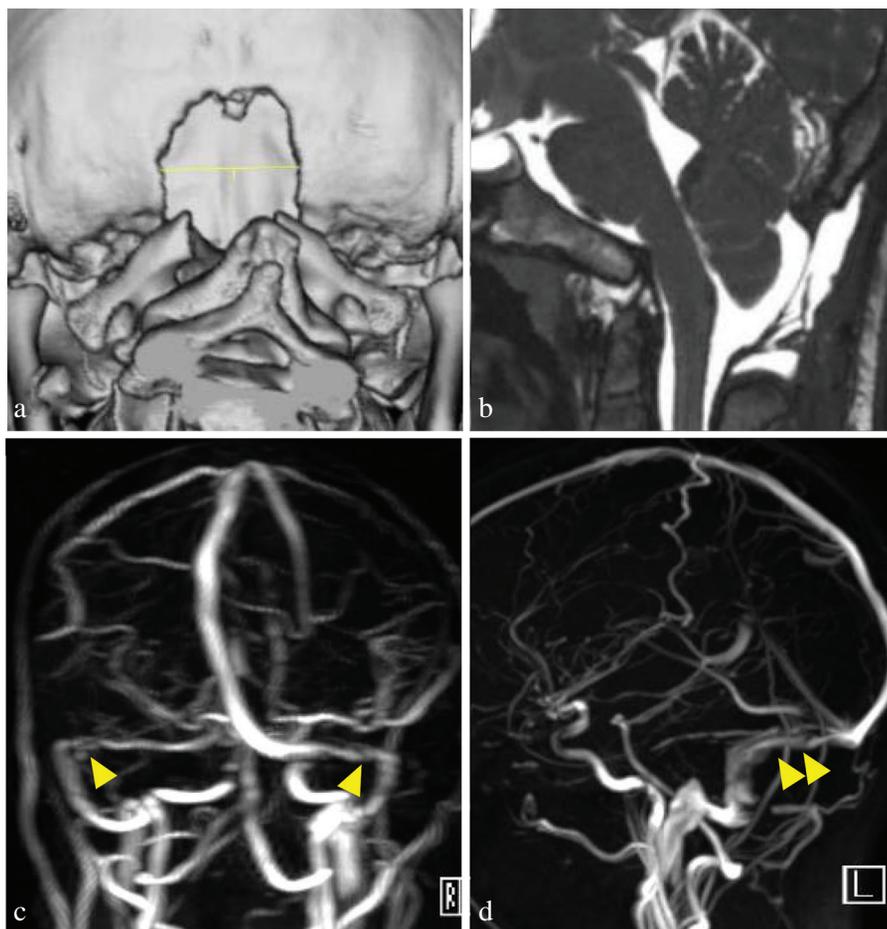


Fig. 4 Postoperative cervical spine 3D-CT image (a) and cervical spine T2WI-MRI (b) obtained seven days after surgery showed successful decompression of the foramen magnum. Postoperative MRV (c and d) obtained at one-year postoperative follow-up showed an improved flow of transverse sigmoid sinus junction (arrow heads).

seizures and fever. Furthermore, hormonal blood exams, CSF studies and possible autoimmune antibodies were completely negative. Diffuse vascular edema could be attributed to VSs, which is well described as a particularly treatable cause among them and is also the key factor which connects CM1 with diffuse brain edema. Some surgically-treated cases of VSs demonstrated gradual radiological recovery like the present case.²²⁾

There are two kinds of CM1, congenital or acquired.^{6,23)} Acquired CM1 is reportedly led by IIH.^{6,23,24)} IIH has close relation to obesity²⁵⁾ and specific neuroimaging findings suggestive of long-standing intracranial hypertension, including empty sella, flattening of the posterior globes, optic nerve head protrusion, distention of the optic nerve sheaths, tortuosity of the optic nerve, cerebellar tonsillar herniation, meningoceles, CSF leaks, and VSs.²⁶⁾ However, there was not any physical and radiological indications of IIH other than tonsillar herniation and VSs in the present case. In congenital CM1, tonsillar herniation caused by overcrowding of small posterior fossa is the primary change. Given the narrowness of posterior cranial fossa and lack of evidence of supratentorial brain herniation as well as retrospective assessment of dramatic effectiveness of FMD in the present case, it is natural to judge the tonsillar herniation was likely to be caused by overcrowding of congenital small

posterior fossa. A subset of patients with congenital CM1 progresses to secondary ICH^{7-9,27,28)} without hydrocephalus with an overall incidence of 1.3% to 2.7% based on MRI.^{22,28,29)} The proposed mechanism is intermittent cranio-spinal dissociation of the CSF fluid pressure brought on by primary stagnation, not complete obstruction, of CSF flow at the foramen magnum.³⁻⁵⁾ Another proposed pathophysiology is seen in patients with slit-ventricle syndrome, which may also pertain to CM1 patients. Slit-ventricle syndrome is a term used to denote patients with headaches, small ventricles on cranial neuroimaging, and a slow refill of the ventricular shunt pumping devices. Authors postulated that increased brain turgor occurs in these patients, which may interfere with CSF absorption and increase the ICP.^{4,27,30)} Given several authors advocate that VSs is a significant contributing factor to IIH,^{22,26)} elevated intracranial pressure of the present case, primarily arising from significant CSF stagnation at foramen magnum, might give rise to VSs and be deteriorated by VSs at the same time like IIH.³¹⁾ Our hypothesis is that combination of high CSF pressure created by CSF stagnation at the foramen magnum and secondary high venous pressure caused by VSs are thought to trigger vicious cycle of worsening of high intracranial pressure (Fig. 3). Furthermore, there was not collateral venous sinus flow formation after VSs, which could lead to significant

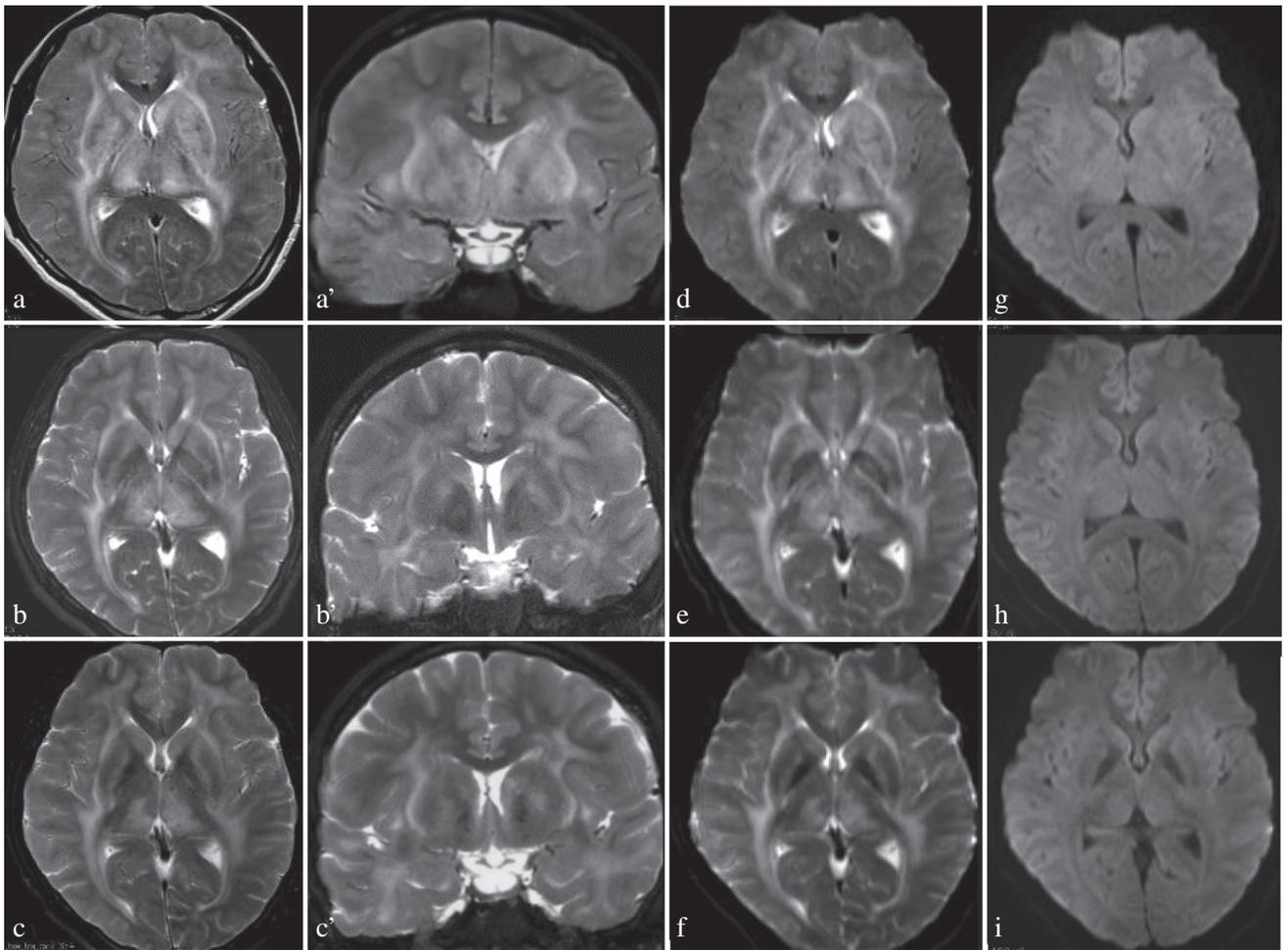


Fig. 5 Head T2WI-MRI (a-c; axial images, a'-c'; coronal images), ADC map (d-f) and DWI (g-i) were obtained preoperatively (a, d, g), at 6-month postoperative follow-up (b, e, h) and 1-year postoperative follow-up (c, f, i). T2WI-MRI and ADC map revealed gradual improvement of diffuse white matter hyperintensities and narrowed brain sulci with time. There were no detectable pathologies on DWI.

venous hypertension. In the end, further progression of VSs as a result of vicious cycle ended up with brain parenchymal edema without ventricular dilatation. The combination of CSF stagnation and VSs is thought to play an important role for creation of extremely high intracranial pressure. CSF flow was not completely blocked at the foramen magnum as confirmed with cine MRI and that is why lumbar puncture demonstrated high pressure, instead of normal pressure in the case of complete block. However, in the presence of an increased intracranial pressure like the present case, lumbar puncture puts patients at increased risk of deteriorating neurologically, and it is not recommended even without complete CSF block.

There is no consensus regarding patient selection, timing for surgical intervention and surgical approach for CM1. However, CM1-induced ICH should be alleviated properly not to leave neurological sequelae on a timely basis. Ramon et al. described that the main objective in the surgical treatment of congenital CM1 should be restoration of normal CSF dynamics at the craniovertebral junction.²³⁾ In the present case, vicious cycle of high intracranial pressure

was thought to be originally initiated by CSF flow stagnation at the foramen magnum. Therefore, FMD has to be first considered to remove the primary cause and break the vicious cycle.

The preoperative assessment of venous outflow is extremely important in the case of CM1 with ICH in consideration of the presence of VSs as well as other collateral venous outflows.³²⁾ If there could exist collateral venous sinuses development, it could interfere with duraplasty and complicate the surgical procedure. To avoid excessive bleeding and other complications, duraplasty should be skipped depending on venous anatomy.³³⁾

The present case is the first illustration of successful surgical treatment of CM1-induced ICH with diffuse brain edema led by VSs. Appropriate surgical planning based on preoperative angiography and MRV is crucial for CM1 with ICH.

Conflicts of Interest Disclosure

There is no conflict of interest to disclose for any of the authors.

References

- 1) [Chiari H: Über Veränderungen des Kleinhirns infolge von Hydrocephalie des Grosshirns]. *Dtsch Med Wochenschr* 17: 1172–1175, 1891 (German)
- 2) Milhorat TH, Chou MW, Trinidad EM, et al.: Chiari I malformation redefined: clinical and radiographic findings for 364 symptomatic patients. *Neurosurgery* 44: 1005–1017, 1999
- 3) Choudhari KA, Cooke C, Tan MH, Gray WJ: Papilloedema as the sole presenting feature of Chiari I malformation. *Br J Neurosurg* 16: 398–400, 2002
- 4) Vaphiades MS, Eggenberger ER, Miller NR, et al.: Resolution of papilloedema after neurosurgical decompression for primary Chiari I malformation. *Am J Ophthalmol* 133: 673–678, 2002
- 5) Zhang JC, Bakir B, Lee A, Yalamanchili SS: Papilloedema due to Chiari I malformation. *BMJ Case Rep* 2011 Oct 16; 2011 doi: 10.1136/bcr.2011.4721
- 6) Istek S: Chiari type I malformation in a pseudotumour cerebri patient: is it an acquired or congenital Chiari malformation? *BMJ Case Rep* 2014 Jun 4; 2014 doi: 10.1136/bcr-2013-201845
- 7) Banik R, Lin D, Miller NR: Prevalence of Chiari I malformation and cerebellar ectopia in patients with pseudotumour cerebri. *J Neurol Sci* 247: 71–75, 2006
- 8) Bejjani GK: Association of the adult Chiari malformation and idiopathic intracranial hypertension: more than a coincidence. *Med Hypotheses* 60: 859–863, 2003
- 9) Lancione RR, Kosmorsky GS: When does low mean high? Isolated cerebral ventricular increased intracranial pressure in a patient with a Chiari I malformation. *J Neuroophthalmol* 21: 118–120, 2001
- 10) Maas RP, Muller-Hansma AH, Esselink RA, et al.: Drug-associated progressive multifocal leukoencephalopathy: a clinical, radiological, and cerebrospinal fluid analysis of 326 cases. *J Neurol* 263: 2004–2021, 2016
- 11) Marra AM, Barilaro G, Vilella V, Granata M: Eosinophilic granulomatosis with polyangiitis (EGPA) and PRES: a case-based review of literature in ANCA-associated vasculitides. *Rheumatol Int* 35: 1591–1595, 2015
- 12) Zhu S, Nahas SJ: CADASIL: imaging characteristics and clinical correlation. *Curr Pain Headache Rep* 20: 57, 2016
- 13) Chabriat H, Vahedi K, Iba-Zizen MT, et al.: Clinical spectrum of CADASIL: a study of 7 families. Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy. *Lancet* 346: 934–939, 1995
- 14) Nozaki H, Sekine Y, Fukutake T, et al.: Characteristic features and progression of abnormalities on MRI for CARASIL. *Neurology* 85: 459–463, 2015
- 15) Yasude T, Sekijima Y, Mitsuhashi S, Gono T, Matsuda M, Ikeda S: Benign intracranial hypertension and leukoencephalopathy due to venous sinus stenosis in an SLE patient. *Lupus* 16: 839–840, 2007
- 16) Ogawa M, Ishimaru K, Shirotu T, Baba M, Matsunaga M: [A case of benign intracranial hypertension associated with systemic lupus erythematosus (SLE) showing diffuse white matter lesions on MRI]. *Rinsho Shinkeigaku* 34: 577–581, 1994 (Japanese)
- 17) Chaves-Carballo E, Dabbagh O, Bahabri S: Pseudotumor cerebri and leukoencephalopathy in childhood lupus. *Lupus* 8: 81–84, 1999
- 18) Jeong HN, Suh BC, Kim YB, Chung PW, Moon HS, Yoon WT: Posterior reversible encephalopathy syndrome as an initial neurological manifestation of primary Sjögren's syndrome. *Clin Auton Res* 25: 259–262, 2015
- 19) Chaves-Carballo E, Frank LM, Chrenka BA: Increased intracranial pressure in adrenoleukodystrophy. *Arch Neurol* 41: 339–340, 1984
- 20) Garcia L, Gascon G, Ozand P, Yaish H: Increased intracranial pressure in Alexander disease: a rare presentation of white-matter disease. *J Child Neurol* 7: 168–171, 1992
- 21) Waragai M, Takeuchi H, Fukushima T, Haisa T, Yonemitsu T: MRI and SPECT studies of dural arteriovenous fistulas presenting as pure progressive dementia with leukoencephalopathy: a cause of treatable dementia. *Eur J Neurol* 13: 754–759, 2006
- 22) Onder H, Gocmen R, Gursoy-Ozdemir Y: Reversible transverse sinus collapse in a patient with idiopathic intracranial hypertension. *BMJ Case Rep* 2015 May 15; 2015. doi: 10.1136/bcr-2014-011606
- 23) Ramón C, Gonzáles-Mandly A, Pascual J: What differences exist in the appropriate treatment of congenital versus acquired adult Chiari type I malformation? *Curr Pain Headache Rep* 15: 157–163, 2011
- 24) Aiken AH, Hoots JA, Saindane AM, Hudgins PA: Incidence of cerebellar tonsillar ectopia in idiopathic intracranial hypertension: a mimic of the Chiari I malformation. *Am J Neuroradiol* 33: 1901–1906, 2012
- 25) Jensen RH, Radojicic A, Yri H: The diagnosis and management of idiopathic intracranial hypertension and the associated headache. *Ther Adv Neurol Disord* 9: 317–326, 2016
- 26) Bidot S, Saindane AM, Peragallo JH, Bruce BB, Newman NJ, Biousse V: Brain imaging in idiopathic intracranial hypertension. *J Neuroophthalmol* 35: 400–411, 2015
- 27) Heiss JD, Patronas N, DeVroom HL, et al.: Elucidating the pathophysiology of syringomyelia. *J Neurosurg* 91: 553–562, 1999
- 28) Saindane AM, Bruce BB, Desai NK, Roller LA, Newman NJ, Biousse V: Transverse sinus stenosis in adult patients with Chiari malformation type I. *Am J Roentgenol* 203: 890–896, 2014
- 29) Sinclair N, Assaad N, Johnston I: Pseudotumour cerebri occurring in association with the Chiari malformation. *J Clin Neurosci* 9: 99–101, 2002
- 30) Rekatte HL: Classification of slit-ventricle syndromes using intracranial pressure monitoring. *Pediatr Neurosurg* 19: 15–20, 1993
- 31) Owlser BK, Parker G, Halmagyi GM, et al.: Pseudotumor cerebri syndrome: venous sinus obstruction and its treatment with stent placement. *J Neurosurg* 98: 1045–1055, 2003
- 32) Bateman GA: Arterial inflow and venous outflow in idiopathic intracranial hypertension associated with venous outflow stenoses. *J Clin Neurosci* 15: 402–408, 2008
- 33) Gürbüz MS, Berkman MZ, Ünal E, et al.: Foramen magnum decompression and duraplasty is superior to only foramen magnum decompression in Chiari I malformation type I associated with syringomyelia in adults. *Asian Spine J* 9: 721–727, 2015

Corresponding author:

Toshiki Fukuoka, MD, Department of Neurosurgery, Nagoya University School of Medicine, 65 Tsurumai-cho, Showa-ku, Nagoya, Aichi 466-8550, Japan.

✉rugby104@gmail.com