Surgical Neurology International

OPEN ACCESS

For entire Editorial Board visit : http://www.surgicalneurologyint.com

Editor-in-Chief: **James I. Ausman**, MD, PhD University of California, Los Angeles, CA, USA

Case Report

Rapidly progressing monoparesis caused by Chiari malformation type I without syringomyelia

Masahiro Oishi, Yasuhiko Hayashi, Daisuke Kita, Issei Fukui, Moeko Shinohara[1], John D. Heiss[2], Jun-ichiro Hamada

Departments of Neurosurgery, and [1]Neurology, Kanazawa University, Kanazawa, Ishikawa, Japan, [2]Surgical Neurology Branch, National Institute of Neurological Disorders and Strokes, National Institutes of Health, Bethesda, Maryland

E-mail: Masahiro Oishi - ma-oishi@med.kanazawa-u.ac.jp; *Yasuhiko Hayashi - yahayashi@med.kanazawa-u.ac.jp; Daisuke Kita - kitad@med.kanazawa-u.ac.jp; Issei Fukui - ifukui@med.kanazawa-u.ac.jp; Moeko Shinohara - m-nohora@med.kanazawa-u.ac.jp; John D. Heiss - HeissJ@ninds,nih,gov; Jun-ichiro Hamada - jhamada@med.kanazawa-u.ac.jp
*Corresponding author

Received: 31 January 13 Accepted: 13 May 13 Published: 12 June 13

This article may be cited as:
Oishi M, Hayashi Y, Kita D, Fukui I, Shinohara M, Heiss JD, *et al.* Rapidly progressing monoparesis caused by Chiari malformation type I without syringomyelia. Surg Neurol Int 2013;4:79.
Available FREE in open access from: http://www.surgicalneurologyint.com/text.asp?2013/4/1/79/113355

Abstract

Background: Patients with Chiari malformation type I (CM-I) can manifest neurological symptoms, such as headache, neck pain, dysesthesia, swallowing disturbance, and paresis, which are usually stable or slowly progressive even if syringomyelia is coexistent. In some instances, however, acute onset of neurological symptoms has been reported but the pathogenetic mechanism and subsequent clinical course have not been explained. In those cases, it was reported that urgent treatment of foramen magnum decompression (FMD) was very effective. This work reports that an 11-year-old girl with CM-I subacutely developed unique symptoms and that urgent treatment of FMD was very effective.

Case Description: We present here an 11-year-old girl with CM-I who subacutely developed dysphagia, left upper extremity monoparesis and sensory dysesthesia, with the limb assuming a peculiar posture at rest, with the wrist in extension and the elbow joint in flexion. Although her symptoms were assumed to be due to previously diagnosed CM-I without syringomyelia, no differences on magnetic resonance imaging (MRI) could be found except for slight change in the shape of tonsils compared with the previous ones. FMD and C1 removal with duraplasty was performed and resulted in an excellent neurological recovery.

Conclusion: This case is a reminder that the presence of a new neurological deficit referable to nuclei within, or tracts that traverse, the cerebromedullary junction is a firm surgical indication for FMD in a patient with CM-I. MRI was nearly identical during the asymptomatic and symptomatic periods in this case, and did not explain the timing of symptom onset.

Key Words: Acute onset, Chiari malformation type I, monoparesis, syringomyelia

Access this article online

Website: www.surgicalneurologyint.com

DOI: 10.4103/2152-7806.113355

Quick Response Code:

INTRODUCTION

Chiari malformation type I (CM-I) is typically defined as cerebellar tonsillar herniation of more than 5 mm from the foramen magnum and may be associated with syringomyelia, hydrocephalus, and/or scoliosis.[1,16,18,19]

Many authors have suggested that tonsillar descent can result from decreased volume of the posterior cranial fossa and skull base dysplasia.[11,19]

Some symptoms of CM-I patients initially appear variable and nonspecific, including headache, neck pain, brainstem or cerebellar dysfunction, cranial nerve signs, and spinal cord dysfunction.[2,6,7,10,11,13,18,19] The majority of patients with CM-I present with stable or slowly progressive symptoms and clinical courses and are diagnosed during late childhood or adulthood.[5,7,11,13,18]

The widespread availability and use of magnetic resonance imaging (MRI) allow earlier and more frequent detection of CM-I during the developmental stage of this disease, when no or minimal neurological deficits have emerged.[7,11,13] The management of asymptomatic patients with CM-I is still controversial because the natural history of such patients is not clear. Foramen magnum decompression (FMD) is usually not recommended in asymptomatic patients without syringomyelia. Some authors have reported a relatively benign natural history for asymptomatic or minimally symptomatic CM-I and the results of these studies could support observation of asymptomatic CM-I patients.[2,11,12] However, in previously asymptomatic patients with CM-I and syringomyelia, acute onset of CM-I is sometimes reported. Acute clinical deterioration in patients with previously asymptomatic CM-I without syringomyelia is rarer and its pathogenetic mechanism remains unknown.[7,18,19]

We report a case of an 11-year-old girl with CM-I diagnosed when she was aged 1 year and remained asymptomatic for 10 years before manifesting acute focal symptoms, including motor and sensory deficits in left upper forearm and dysphagia. Despite her new neurological deficits, MRI could detect only minute change from previous imaging.

CASE REPORT

An 11-year-old girl presented with the gradual onset over one day of sensory dysesthesia of her left forearm followed the day of admission by severe monoparesis of the ipsilateral forearm, increasing dysesthesia, and dysphagia. She recalled that the day the symptoms developed, she swam faster and for a longer duration than usual. She was referred to our hospital one week from the onset because her symptoms had not resolved. Her past history was significant for hydrocephalus from aqueductal stenosis that was diagnosed and treated when she was aged 1 year. Her ventriculomegaly and symptoms at that time resolved completely following endoscopic third ventriculoscopy. CM-I was diagnosed by radiographic criteria. During the subsequent 10 years, she received annual MRI and neurological examinations that documented persistent tonsillar ectopia but no neurological deficits.

On admission, her physical examination revealed spastic paresis of left upper limb, with the limb assuming a peculiar posture at rest, with the wrist in extension and the elbow joint in flexion [Figure 1]. The other elements of neurological examination, mental status, cranial nerves, motor, sensory, cerebellar, and deep tendon reflexes were normal except for decreased gag reflex. Besides, she had neither headache, neck pain nor motor deficits of lower extremities, and denied experiencing any Valsalva-induced symptoms. There was no known history of head trauma.

MRI a week from the onset showed cerebellar tonsillar herniation of 8 mm below McRae's (basion-opisthion) line, which was the same amount as on previous imaging. No recurrence of hydrocephalus was demonstrated [Figure 2a-c]. There was no evidence of syringomyelia or abnormal T2 signal in the spinal cord [Figure 2d and e]. Sagittal fast imaging employing steady state acquisition (FIESTA) MRI clearly showed that compared with the previous in detail, the tonsil changed into beaked shape and was asymmetry [Figure 3]. Clinical signs and symptoms pointed to dysfunction of the cervicomedullary junction despite stable imaging findings. Surgical treatment consisting of FMD, C1 laminectomy and duraplasty was performed without complication.

After the operation, dysesthesia and monoparesis of left forearm and dysphagia improved steadily. She was discharged on the 14th postoperative day with complete neurological recovery. At the follow-up 6 months after FMD, she maintained her recovery. Postoperative MRI demonstrated an ascent of the cerebellar tonsil of 4 mm compared with before FMD [Figure 4].

DISCUSSION

We report on a child presenting with the acute onset of focal neurological findings attributed to CM-I despite having asymptomatic CM-I for the preceding 10 years. There was no apparent difference between MRI before

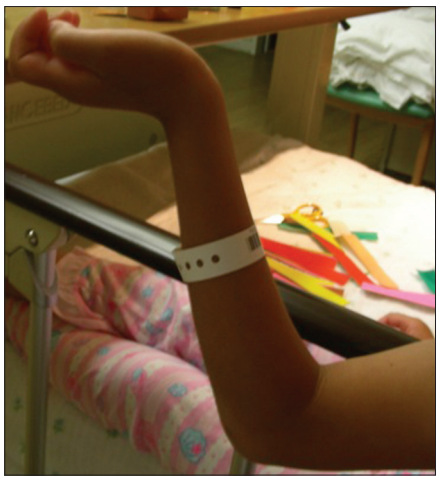

Figure 1: Her left upper limb holding in a peculiar position, with the wrist in extension and the elbow joint in flexion

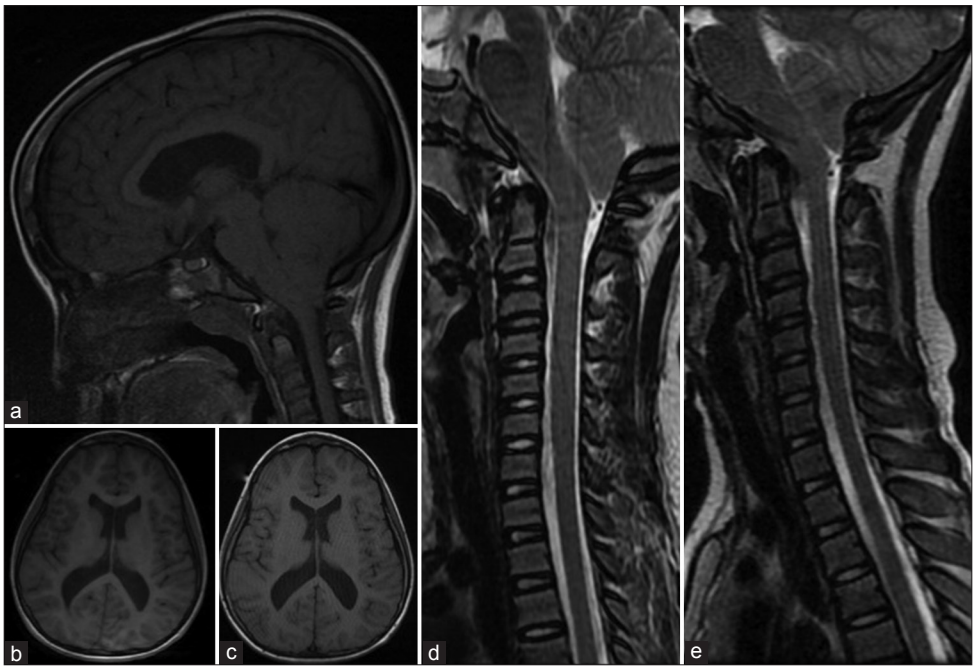

Figure 2: (a) Preoperative sagittal T1-weighted MRI revealing the cerebellar tonsillar herniation of 8 mm. (b) Axial T1-weighted MRI 6 months before the onset and (c) one week after the onset, and (d) sagittal T2-weighted MRI 6 months before the onset and (e) one week after the onset manifesting no progression of cerebellar tonsillar herniation and no syringomyelia

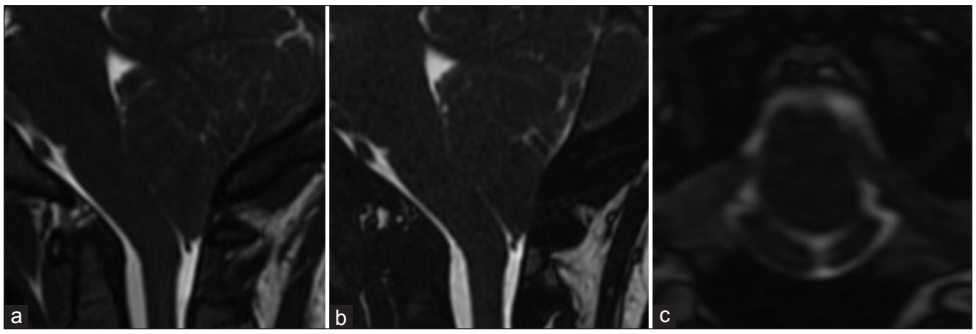

Figure 3: (a) Sagittal fast imaging employing steadystate acquisition (FIESTA) MRI 6 months before the onset and (b) one week after the onset of monoparesis revealed different shapes of tonsils. (c) Axial FIESTA MRI one week after the onset showed the right-sided dominant tonsillar herniation

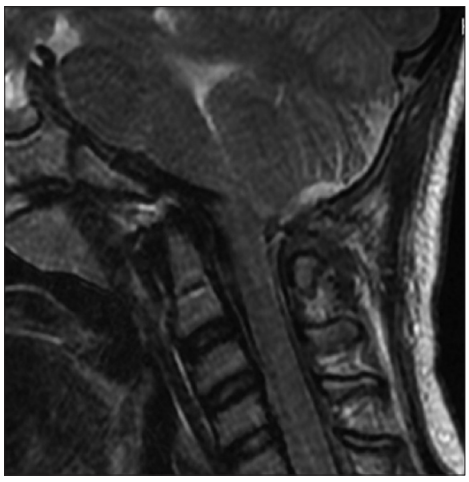

Figure 4: Postoperative sagittal T2-weighted MRI one week after FMD showing the improvement of the cerebellar tonsillar herniation

and after onset of symptoms, except for slight change in the shape of tonsils. Patients with CM-I who have remained asymptomatic for a long period usually would be expected to have relatively benign outcomes.[11,12,14] But, this supposition is brought into question by some authors who have reported patients with CM-I presenting acutely, although it is difficult to discern whether the symptoms resulted from CM-I or associated syringomyelia.[7,18]

The papers about acute onset of CM-I implied that acute onset of symptoms was quite sporadic and infrequent. Some authors proposed a mechanism in which mild head and/or neck injury triggers the abrupt onset of CM-I and neurological deterioration.[17-19] The trauma is purported to cause increased tonsillar ectopia that disrupts the previous equilibrium, resulting in ischemic dysfunction of the tonsils and brainstem or

malcirculation of cerebrospinal fluid (CSF) around the cervicomedullary junction.[7] The blockage and subsequent elevated pressure due to the descendant cerebellar tonsils directs CSF into the central canal at the cervical spinal cord, compresses the brainstem and upper cervical spinal cord, and may trigger acute neurological deterioration.[7]

Most of the subjects in those papers had syringomyelia, and patients with CM-I and syringomyelia may be at higher risk of acute onset and serious neurological deterioration than those without syrinx.[7,19] In some cases with acute onset, sudden cardiopulmonary arrest or death has occurred.[5,7] In contrast, it was reported that the degree of tonsillar herniation at presentation might not be associated with the worsening of neurological symptoms.[3,8] In our case, the degree of tonsillar descendent was mild and was not associated with syringomyelia. The time course of symptoms can suggest an acute condition, such as would occur with compression, edema and ischemia or acute inflammatory lesion at the craniocervical junction. In our case, compression seemed most likely because MRI did not show an enhancing lesion. The patient's immediate response to decompressive surgery also validates the view that it was caused by compression of cervicomedullary junction at the foramen magnum. Judging from little change into the beaked shape of tonsil on detailed MRI, it was suggested that slight increase compression of the craniocervical junction could cause the abrupt onset of symptomatic CM-I. The craniocervical junction could be left vulnerable by a rhythmic, systematic, and prolonged neck turning in the same direction during swimming that would be no consequence in a normal individual. In addition to the repeated microtrauma, the halting of respiration during swimming would be considered to cause the paroxysmal elevation of intracranial pressure. We advocate that the moderate or sporadic increases in intracranial pressure could be enough to develop symptomatic compression of the craniocervical junction of patient with CM-I.

One of the most common acute manifestations of CM-I is the respiratory distress, which is followed by dysphagia and hemiparesis [Table 1].[7,18,19] In our case, she presented with spastic monoparesis and dysesthesia of the left upper extremity and dysphagia. In the syndrome of the decussation of the pyramids, there should be contralateral spastic paresis of the lower and upper extremity. In this case, by the right-sided dominant cerebellar tonsillar

Table 1: Patients presenting with acute neurological symptoms from CM-I

Authors and year	Age (years), sex	Days between onset and operation	Syrinx	Trauma	Neurological deficits	Outcome	Follow-up (month)
Yarbrough *et al*., 2011	12, F	2	Yes	History of falls	Quadriparesis	Complete resolution	26
	13, F	5	Yes	No	Lt UE and LE paresthesias	Complete resolution	12
	3, M	3	Increased T2 signal intensity in spinal cord	No	Vocal cord paresis	Complete resolution	14
	14, M	14	Yes	Blow to head during football game	Rt UE paresthesias	Complete resolution	11
	10, M	14	Yes	Fall from standing	Paraparesis	Complete resolution	24
	13, F	8	Yes	Flip on trampoline, landing on feet	Rt hemianesthesia	Complete resolution	5
Wellons JC III *et al*., 2007	16, M	3	Unknown	No	Dysphagia, Rt hemiparesis, respiratory distress	Mild hemiparesis	Unknown
	7, M	2	Yes	No	Rt UE and LE paresis and paresthesias, left anisocoria	Mild hemiparesis	12
Massami *et al*., 2011	2, M	1	Yes	Head turned forcibly by older boy	Tetraparesis, hypesthesia, dyspnia	Mild hemiparesis	10
	38, M	12	Yes	No	Acute respiratory failure	Complete resolution	36
	1, M	7	No	Knock his head on the foot of table	Lt hemiparesis and palpebral ptosis, Lt myosis, dysphagia	Complete resolution	36
Present case	11, F	14	No	No	Lt UE monoparesis and dysesthesia, dysphagia	Complete resolution	6

Lt: Left side, Rt: Right side, UE: Upper extremity, LE: Lower extremity, CM-I: Chiari malformation type I

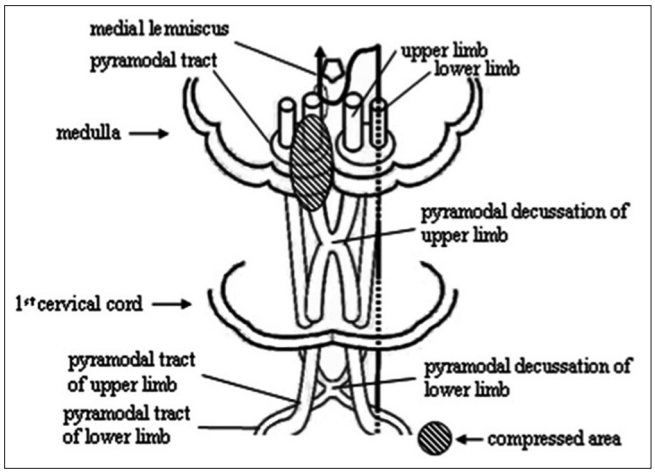

Figure 5: The pyramidal tract to the upper extremity crosses before one to the lower extremity and the medial lemniscus decussates just superior to the pyramidal decussation

herniation the compression of the medulla affected the corticospinal fibers that control the left upper extremity, which cross before the corticospinal tract fibers to the lower extremity, and the medial lemniscus [Figure 5]. Our case was not associated with syringomyelia and the tonsillar ectopia was of a mild degree (8 mm). Besides, MRI demonstrated little differences compared with the previous ones obtained while she was asymptomatic. For these reasons, we believe that a restricted area at the level of the foramen magnum was affected to cause monoparesis and dysesthesia.

Although the surgical treatment of CM-I is well known among neurosurgeons, the decision whether to operate on asymptomatic patients with CM-I is quite difficult and controversial.[14] Some authors have advised performing prophylactic surgery in asymptomatic children to avoid the risks of CM-I progression.[4,9,15] However, it seems to be considerably infrequent for untreated CM-I to cause permanent neurological deficits after an unexpected neurological deterioration, based on the reports of the natural history of CM-I.[2,10,12] Accordingly, several authors currently prefer conservative management for asymptomatic children with CM-I.[7,11,15]

In spite of the initial severity, the outcome of the patients surviving after an abrupt onset of neurological deficit is generally good with early FMD.[18,19] The short time between clinical onset and surgical decompression certainly plays a crucial role in achieving such a good outcome. Neurosurgeons and neurologists should keep in mind that surgical intervention in the form of FMD should be performed as early as possible if acute brainstem or long tract signs develop.

CONCLUSION

It is very important to recognize that acute neurological decline in a previously asymptomatic patient with CM-I is an indication for urgent treatment with FMD, even if MRI is unchanged between the symptomatic and asymptomatic state. The outcome of FMD appears to be excellent even for the patients with acute neurological deterioration.

REFERENCES

1. Aboulezz AO, Sartor K, Geyer CA, Gado MH. Position of cerebellar tonsils in the normal population and in patients with Chiari I malformation: A quantitative approach with MR imaging. J Comput Assist Tomogr 1985;9:1033-6.
2. Benglis D Jr, Covington D, Bhatia R, Bhatia S, Elhammady MS, Ragheb J, *et al*. Outcomes in pediatric patients with Chiari malformation Type I followed up without surgery. Clinical article. J Neurosurg Pediatr 2011;7:375-9.
3. Elster AD, Chen MY. Chiari I malformations: Clinical and radiological reappraisal. Radiology 1992;183:347-53.
4. Genitori L, Peretta P, Nurisso C, Macinante L, Mussa F. Chiari type I anomalies in children and adolescents: Minimally invasive management in a series of 53 cases. Childs Nerv Syst 2000;16:707-18.
5. Gentry JB, Gonzalez JM, Blacklock JB. Respiratory failure caused by Chiari I malformation with associated syringomyelia. Clin Neurol Neurosurg 2001;103:43-5.
6. Laufer I, Engel M, Feldstein N, Souweidane MM. Chiari malformation presenting as a focal motor deficit: Report of 2 cases. J Neurosurg Pediatr 2008;1:392-5.
7. Massami L, Della Peta GM, Tamburrini G, Di Rocco C. Sudden onset of Chiari malformation Type I in previously asymptomatic patients. Report of 3 cases. J Neurosurg Pediatr 2011;8:438-42.
8. Meadows J, Kraut M, Guarnieri M, Haroun RI, Carson BS. Asymptomatic Chiari type I malformations identified on magnetic resonance imaging. J Neurosurg 2000;92:920-6.
9. Navarro R, Olavarria G, Seshadri R, Gonzales-Portillo G, McLone DG, Tomita T. Surgical results of posterior fossa decompression for patients with Chiari I malformation. Childs Nerv Syst 2004;20:349-56.
10. Nishizawa S, Yokoyama T, Yokota N, Tokuyama T, Ohta S. Incidentally identified syringomyelia associated with Chiari I malformations: Is early interventional surgery necessary? Neurosurgery 2001;49:637-41.
11. Novegno F, Caldarelli M, Massa A, Chieffo D, Massami L, Pettorini B, *et al*. The natural history of the Chiari type I anomaly. J Neurosurg Pediatr 2008;2:179-87.
12. Schijman E, Steinbok P. International survey on the management of Chiari I malformation and syringomyelia. Childs Nerv Syst 2004;20:341-8.
13. Steinbok P. Clinical features of Chiari I malformations. Childs Nerv Syst 2004;20:329-31.
14. Strahle J, Muraszko KM, Kapurch J, Bapuraj JR, Garton HJL, Maher CO. Natural history of Chiari malformation Type I following decision for conservative treatment. J Neurosurg Pediatr 2011;8:214-21.
15. Tubbs RS, Lyerly MJ, Loukas M, Shoja MM, Oakes WJ. The pediatric Chiari I malformation: A review. Childs Nerv Syst 2007;23:1239-50.
16. Tubbs RS, McGirt MJ, Oakes WJ. Surgical experience in 130 pediatrics patients with Chiari I malformations. J Neurosurg 2003;99:291-6.
17. Wan MJ, Nomura H, Tator CH. Conversion to symptomatic Chiari I malformation after minor head or neck trauma. Neurosurgery 2007;63:748-53.
18. Wellons JC III, Tubbs RS, Bui CJ, Grabb PA, Oakes WJ. Urgent surgical intervention in pediatric patients with Chiari malformation type I. Report of two cases. J Neurosurg 2007;107 (1 Suppl):S49-52.
19. Yarbrough CK, Powers AK, Park TS, Leonard JR, Limbrick DD, Smyth MD. Patients with Chiari malformation Type I presenting with acute neurological deficits: Case series. J Neurosurg Pediatr 2011;7:244-7.

Disclaimer: The authors of this article have no conflicts of interest to disclose, and have adhered to *SNI's* policies regarding human/animal rights, and informed consent. Advertisers in *SNI* did not ask for, nor did they receive access to this article prior to publication.