

A case of bilateral medial medullary infarction Presenting with “heart appearance” sign

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Bilateral medial medullary infarction (bilateral MMI) is an extremely rare cerebrovascular accident presenting with quadriplegia as the initial symptom and resulting in poor functional prognosis. Diagnosis of bilateral MMI has become possible based on brain MRI findings in recent years, but is still very difficult to diagnose. In the present case, brain MRI was performed 9 hours after the onset, and the infarcted area was detected only by diffusion-weighted MRI. However, changes over time were clearly detected by FLAIR-MRI on days 3, 5 and 7, but it is essential to confirm the disease by DW-MRI in the early stage. The infarct observed on horizontal MRI sections showed the characteristic “heart appearance” sign. For an early diagnosis of bilateral MMI, it is essential to bear in mind that characteristic findings may be obtained by diffusion-weighted MRI.

Key words: bilateral medial medullary infarction, MRI, diffusion-weighted image

INTRODUCTION

Medial medullary infarction (MMI) accounts for only less than 1% of all cases of brain infarctions, and very rarely occurs bilaterally [1]. MMI develops acutely or subacutely, presenting with dysarthria, dysphagia and quadriplegia, sometimes complicated by respiratory disorder, resulting in poor prognosis [2, 3]. Diagnosis of bilateral MMI has become possible based on brain MRI findings in recent years, but is still very difficult to diagnose. We encountered a patient with early stage bilateral MMI in whom the infarcted area with its characteristic morphology was identified by diffusion-weighted MRI of the brain. We report herein this case, along with the sequential changes in MRI findings.

CASE REPORT

On June 24, 2006, a 62-year-old man was referred to us with symptoms of dysarthria and mild hemiparesis and the suspected diagnosis of cerebral infarction or Guillain-Barré syndrome. The patient had been diagnosed hypertension, hyperlipidemia and diabetes mellitus in 1986; had suffered a cerebral infarction in 1992 and myocardial infarction in 1994. This time, he had complained of discomfort in the left upper and lower extremities two days previously and had started to experience weakness of the limbs and gait difficulty around noon on June 24. On admission to our

hospital, his blood pressure was 178/84 mmHg, pulse was 78/min (normal), and body temperature was 36.6 °C; he was lucid but had mild dyslalia. Pupils and eye movement were normal. Facial sense and movement showed no asymmetry, nor was hearing impairment observed. The soft palate did not show any asymmetry but pharyngeal reflex was absent. Tongue protrusion was possible and not deviated. Left upper and lower extremities showed mild hemiparesis and there was no sensation below the neck. The deep tendon reflex was enhanced bilaterally, with both bilateral Babinski's reflex and bilateral Chaddock's reflex being positive. Hematological test showed no abnormality in peripheral blood. Biochemistry showed elevated levels of Glu (157 mg/dL), HbA1c (7.5%), TG (180 mg/dL), Cre (1.6 mg/dL) and UA (8.5 mg/dL). Qualitative urinalysis showed proteinuria (3+).

Brain diffusion-weighted (DW) magnetic resonance imaging (MRI) at 1.5 T revealed heart-shaped hyperintensity areas in the bilateral ventral medulla (Fig. 1). No abnormal signal was observed in the same region by fluid-attenuated inversion recovery (FLAIR). Based on these findings, the patient was diagnosed as having an acute phase bilateral medial medullary infarction, and he was treated with aspirin, argatroban, and edaravone. However, the symptoms gradually progressed to quadriplegia on hospital day 5. Signs of bulbar palsy such as dysarthria and dysphagia led to aspiration pneumonia. Brain MRI performed on hospital

days 3 and 5 showed heart-shaped hyperintensity areas in bilateral ventral medulla both by diffusion-weighted and FLAIR MRI (Fig. 2). On week 14 of hospitalization, no change was observed by DW-MRI, while the infarcts showed edematous change on FLAIR-MRI.

Subsequently, the quadriplegia improved slightly but the patient did not recover motor function to a usable level. The patient was transferred to another hospital on day 120 from the onset of symptoms.

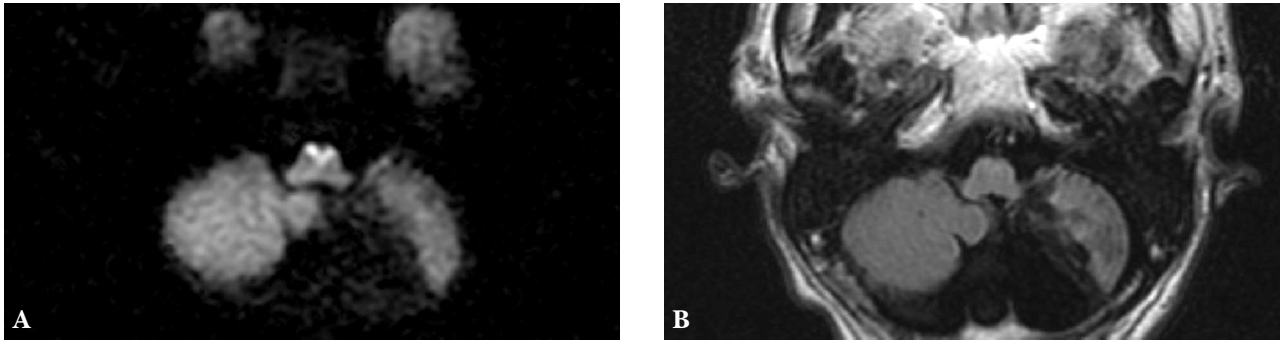


Fig. 1 MRI images (A: DW (SE-EPI; b value=1000), B: FLAIR (TR=8000, TE=130)). Hyperintensity areas are observed in the bilateral ventral medulla by DW-MRI but not by FLAIR-MRI.

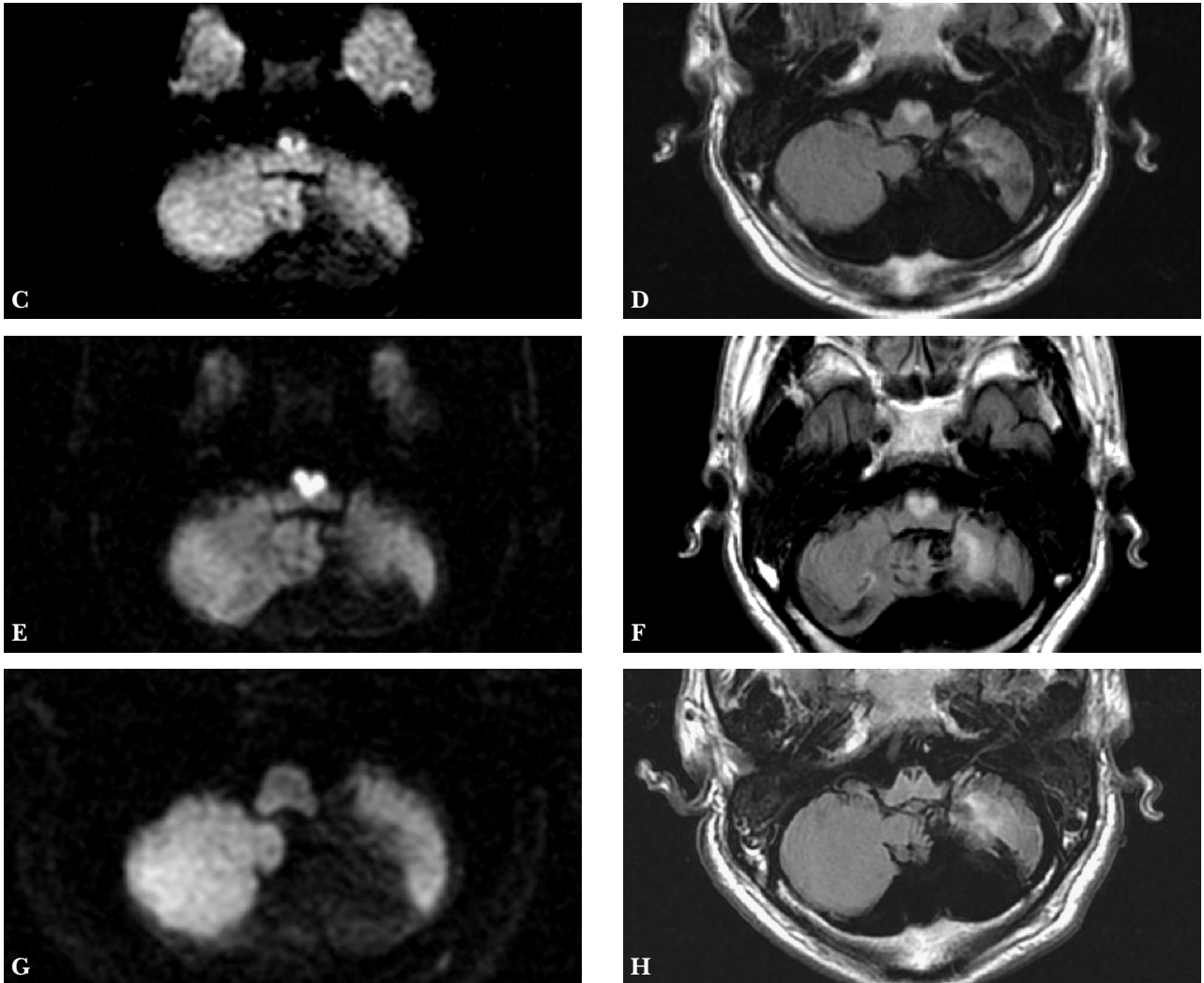
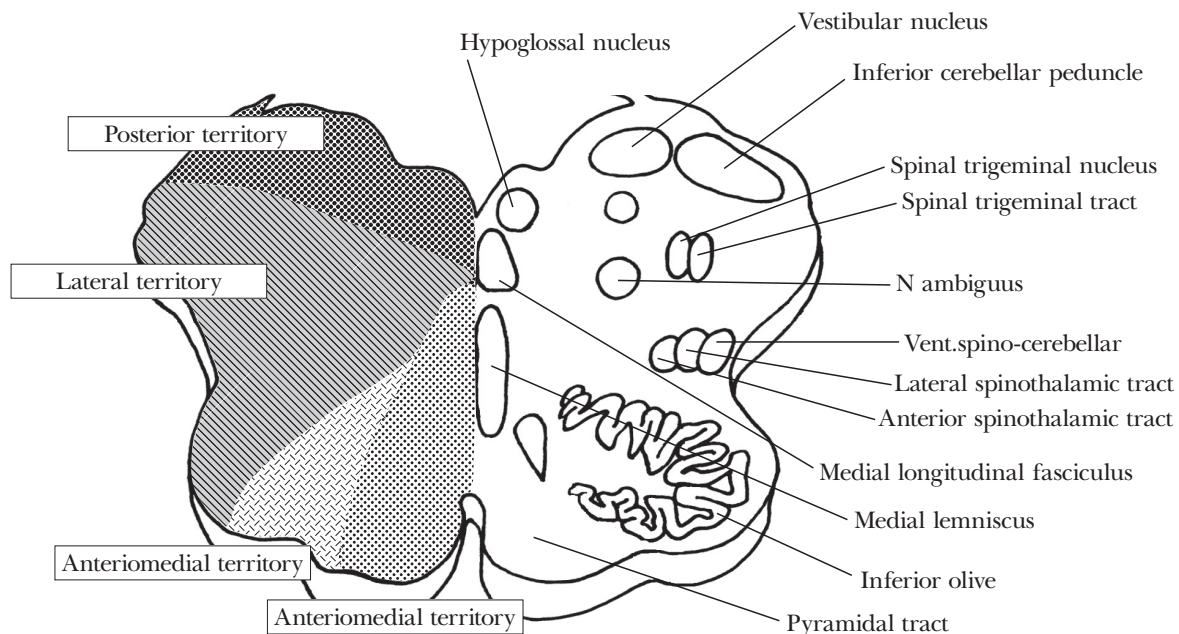


Fig. 2 MRI images obtained on day 3, day 5 and week 14 after the onset of symptoms (C, E, G: DW-MRI (SE-EPI; b value=1000), D, F, H: FLAIR-MRI (TR=8000, TE=130)). Hyperintensity areas are observed in the bilateral ventral medulla both by FLAIR-MRI as well as by DW-MRI on day 3 and day 5. On week 14, no change was observed in DW-MRI signals, while the infarcts showed edematous change.

Table 1 Reports of bilateral medial medullary syndrome confirmed by MRI

Authors	Year	MRI	DWI	Symptoms
Toyoda K et al	1992	Day 10	-	Quadriplegia, ophthalmoplegia
Kleinert G et al	1993	Day 21	-	Quadriplegia, sensory loss, lingual palsy
Ohashi T et al	1995	Day 11	-	Quadriplegia, sensory loss, lingual palsy
Wada K et al	1996	Day 5	-	Hemiparesis, facial palsy, ophthalmoplegia
J.-K Roh et al	1996	Day 5	-	Ophthalmoplegia, sensory loss
Hoshino M et al	1997	Day 4	-	Quadriplegia, ophthalmoplegia, sensory loss, lingual palsy
Del Brutto OH et al	1998	-	-	Quadriplegia, lingual palsy
Baible M et al	2000	-	-	Quadriplegia, sensory loss, lingual palsy
Katoh M et al	2000	Day 3	-	Quadriplegia, ophthalmoplegia
Thijs RD et al	2001	After 10 hr	+	Quadriplegia
Kobayashi Z et al	2003	Day 1, 5	-	Quadriplegia
Takano K et al	2003	Day 5	-	Quadriplegia, sensory loss, lingual palsy
Fukuda M et al	2004	Day 3	-	Hemiparesis
Maeda M et al	2004	After 35 hr	+	Quadriplegia, sensory loss, lingual palsy
Jikumaru M et al	2006	Day 8	+	Quadriplegia, ophthalmoplegia
Present case	2006	After 9 hr	+	Quadriplegia, sensory loss

**Fig. 3** Diagrammatic representation of the MRI findings in medulla oblongata and vascularization

DISCUSSION

Bilateral medial medullary infarction (bilateral MMI) is an extremely rare cerebrovascular accident [1] presenting with quadriplegia as the initial symptom and resulting in poor functional prognosis [2, 3]. In the early stage, bilateral MMI is sometimes misdiagnosed as Guillain-Barré syndrome. There have been reports of approximately 40 cases of bilateral MMI so far, but the diagnosis is often extremely difficult. Among them, there are 15 cases of bilateral MMI that were confirmed by MRI [4-18] (Table 1). In 2 [13, 17] of these cases, brain MRI was performed 10 to 35

hours after the onset of symptoms and the infarction was confirmed by diffusion-weighted imaging, whereas in most other cases brain MRI was performed several days after the onset or hospitalization, with cerebral infarcts confirmed only by T2-weighted imaging. In the present case, brain MRI was performed 9 hours after the onset, and the infarcted area was detected only by diffusion-weighted MRI. However, changes over time were clearly detected by FLAIR-MRI on days 3, 5 and 7, but it is essential to confirm the disease by DW-MRI in the early stage.

The infarct observed on horizontal MRI sections showed the characteristic “heart appearance” sign [13].

The medulla oblongata is divided into anterior-medial territory, anterior-lateral territory, lateral territory, and posterior territory, according to vascular supply [19] (Fig. 3). It is considered that blood is supplied to these areas by the vertebral artery and the anterior spinal artery [6, 20, 21, 22], but it is often difficult to identify the occluded blood vessel because of the vastly complex network formed by these blood vessels. The “heart appearance” sign is considered to appear when the infarct occurs in the former two regions (anterior-medial territory, anterior-lateral territory).

For an early diagnosis of bilateral MMI, it is essential to bear in mind that characteristic findings may be obtained by diffusion-weighted MRI. Early diagnosis and treatment may delay the progression of complications due to the infarction.

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