Bilateral Medial Medullary Infarct with Unique Radiological Presentation

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Abstract

Bilateral medial medullary infarction is a rare subtype of stroke.The typical heart-shaped appearance on magnetic resonance imaging is pathognomonic for bilateral medial medullary syndrome. Vertebrobasilar dolichoectasia is a condition characterized by tortuous dilatation and marked enlargement of the basilar and vertebral arteries, and it may cause posterior circulation infarction.

We present the case of a 55-year-old female patient with complaints of speech disorder, regression in consciousness, and difficulty breathing. Diffusion-weighted imaging examination was normal on arrival. In the cranial imaging after 24 hours, acute infarction was observed in the bilateral medial medullary area. Time-of-flight magnetic resonance angiography revealed vertebrobasilar dolichoectasia.

In this report, a case of bilateral medial medullary infarction with a unique radiological appearance accompanied by vertebrobasilar dolichoectasia, which is rarely reported in the literature, is presented.

Keywords: **Stroke, infarct, bilateral medial medullary infarction, heart appearance**

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INTRODUCTION

Bilateral medial medullary infarction (MMI) is a rare subtype of stroke that usually presents with the sudden onset of tetraparesis, deep sensation loss, hypoglossal paralysis, and bulbar dysfunction. Respiratory dysfunction may also accompany this stroke subtype. Diagnosis may be difficult in the acute phase due to the heterogeneous clinical presentation and the inability to show radiological pathology in the early period. In particular, MMI cases may provide false-negative results on diffusion-weighted imaging (DWI) early in the development of medullary lesions [(1,2)]. Therefore, careful evaluation of neurological findings is important. A typical heart-shaped appearance on magnetic resonance imaging (MRI) is pathognomonic for bilateral medial medullary syndrome [(3)].

Dolichoectasia is defined as dilated, tortuous, and elongated arteries, and it is most commonly found in intracranial vertebral and basilar arteries. The prevalence of intracranial dolichoectasia is in the range of approximately 0.06-5.8% [(4)]. Hypertension, atherosclerosis, and a genetic predisposition are possible etiologies [(5,6)]. Vertebrobasilar dolichoectasia (VBD) can be asymptomatic in many cases. In some cases, VBD can cause brainstem infarctions, hemorrhages, and compression of brainstem structures. Mortality is higher than expected in cases of VBD-induced

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stroke [4]. This report presents a rare submitted case of bilateral MMI with a unique radiological appearance accompanied by VBD.

CASE PRESENTATION

A 55-year-old female patient with hypertension, diabetes mellitus, and coronary artery disease was brought to the emergency department of a local health center by ambulance with complaints of speech disorder, regression in consciousness, and difficulty breathing. At admission, her blood pressure, heart rate, and respiratory rate were 140/100 mm Hg, 88/min, and 35 per minute, respectively. On neurological examination, consciousness was drowsy, and speech was dysarthric. Facial movements were symmetrical and active. Light reflexes and pupils were normal, and eye movements were normal. Gag reflex was absent, and muscle strength was 1/5 tetraparesis. There was also a bilateral extensor response in the plantar reflex. Hypoglossal nerve functions could not be evaluated because they could not be communicated with the patient. Laboratory examinations were normal. Acute stroke was considered the primary diagnosis. No abnormality was observed in cranial computerized tomography (CT) and DWI examinations. Due to the sudden development of respiratory failure during the follow-up, the patient was intubated and transferred by ambulance to our hospital, which has a regional stroke center. The patient was admitted to the neurology intensive care unit immediately after her arrival and was connected to a mechanical ventilator. Intravenous thrombolysis could not be performed as it was outside the treatment window. Low molecular weight heparin and supportive therapy were started.

On the 24th hour FLAIR and DWI images, the hyperintense lesion extended to the dorsal part and included the bilateral medial medulla oblongata (Figure 1). In addition, VBD was detected by three-dimensional time-of-flight magnetic resonance angiography (3D-TOF MRA) (Figure 2). No thrombus source was detected in the cardiological examinations for the etiology of stroke. There was no significant change in the neurological examination in the clinical follow-ups, and respiratory functions did not improve. Tracheotomy was performed in the third week of hospitalization. The patient died at eight weeks due to sepsis.

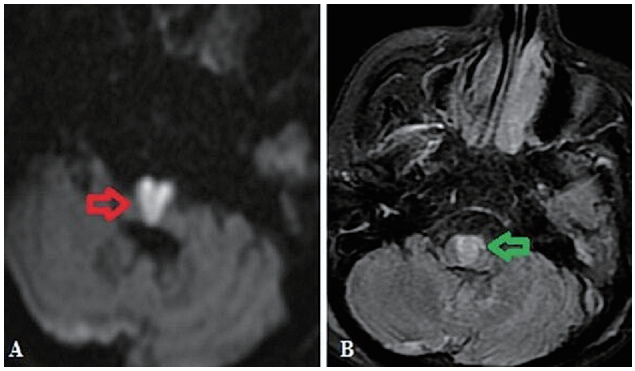

Fig. 1. (A) In DWI sequence, bilateral medial medullary area hyperintensity (red arrow) compatible with heart-shaped acute infarct (B) Fluid-attenuated inversion recovery imaging (FLAIR) also showed bilateral MMI resembling a heart (green arrow).

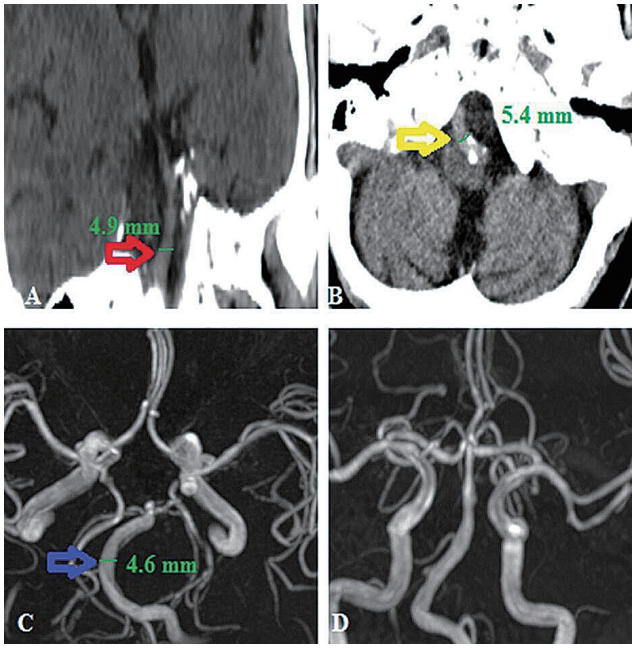

Fig. 2. (A) Coronal CT shows tortuous basilar artery and its diameter as 4.9 mm (red arrow) and the basilar artery bifurcation extends to the base of the 3rd ventricle. (B) Axial CT shows an enlarged calcified vertebral artery indenting from the left into the medulla oblongata (diameter 5.4 mm-yellow arrow). (C) Basilar artery diameter of 4.6 mm at the pons level in the 3D-TOF MRA image (blue arrow). (D) 3D-TOF MRA Vertebral artery appears to be bent to the right.

Written consent was obtained from the relatives of the patients for the publication of this case report.

DISCUSSION

MMI posterior circulation occurs in less than 1% of stroke patients, and bilateral MMI is much less common [7, 8]. Bilateral MMI is a rare subtype of stroke with the sudden onset of quadriparesis/quadriplegia, profound sensory loss, hypoglossal palsy, bulbar dysfunction, and respiratory failure (The anatomical structures and area affected in medial medullary syndrome are presented in figure 3). Limited information on clinical features, etiology, and prognosis is available [1]. The occurrence of acute flaccid tetraplegia and respiratory failure may confuse diagnosis with acute polyneuropathy, brainstem encephalitis, and neuromuscular junction diseases. Early diagnosis is important as the thrombolytic treatment window is limited [9]. Initially, DWI may not show a medullary infarct due to lesion location, imaging time, small size of lesions, inadequate signal-to-noise (S/N) ratio in the early stage of infarction, and artifacts. Therefore, the diagnosis may be delayed. Neurological signs and symptoms should be considered, and imaging studies should be repeated in patients with suspected medullary infarction [2,10]. In this case, the initial DWI examination was normal, which caused a delay in diagnosis. However, clinical findings still pointed to medullary infarction. In the DWI examination after 24 hours, bilateral medial medullary infarction with a heart-shaped appearance was detected in the medulla oblongata. The false-negative rate is not low in the DWI examination performed in the first 24 hours after the onset of stroke symptoms. The false negative rate may increase in small infarcts, especially in posterior circulation stroke cases. In cases of posterior circulation stroke, the false-negative rate of initial DWI has been reported to be approximately 19%. This rate is even higher in brainstem infarcts (31%) [11]. In the study of Fukuoka et al., false-negative initial DWI was found in 4 (13%) of 29 lateral medullary infarction cases and 3 (37%) of 8 MMI cases. Concerning this result, a higher false-negative DWI rate was reported in medial medullary infarction[2]. Therefore, a detailed neurological examination is still considered important for the detection of medullary infarction. These results suggest that detailed neurological examination is still important for the detection of medullary infarction.

In addition to clinical features, it is pathognomonic to observe a heart-shaped infarction extending along the ventral, medial, and dorsal medulla in DWI for bilateral MMI [1,3,9]. Studies have shown that bilateral MMI expands more frequently in the dorsal area than unilateral MMI [9,10,12,13]. At the DWI evaluation of the present case, a unique radiological appearance extending to the bilateral dorsal area and resembling the pathognomonic heart appearance for bilateral MMI was detected.

Causes of MMI include vertebral artery stenosis,

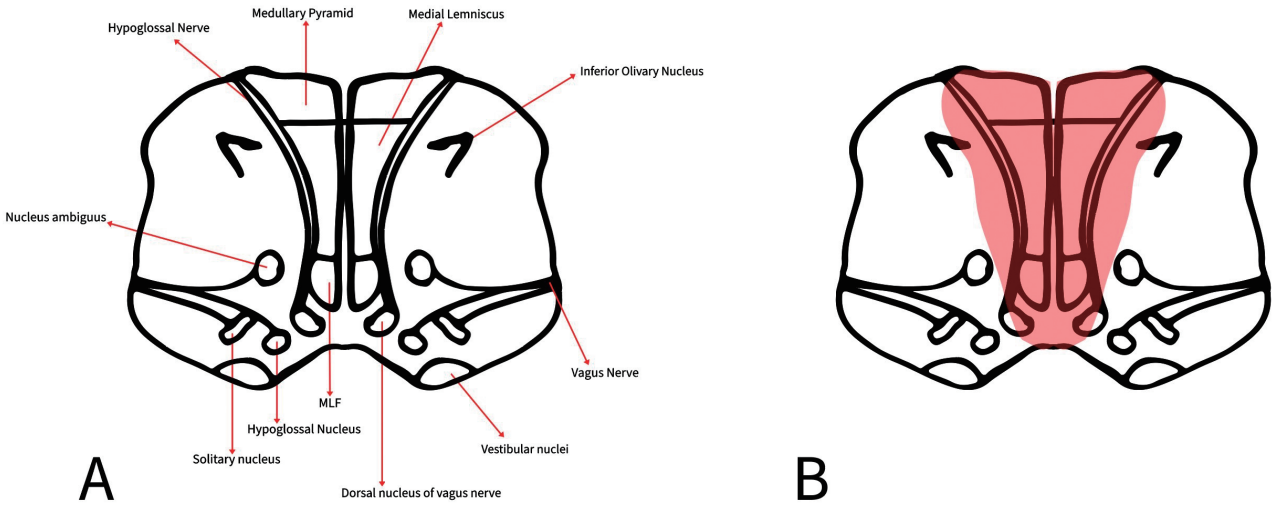

Fig. 3. (A) Schematic drawing of neuroanatomical structures in the medulla oblongata (transverse section of the level of the olivary nucleus). (B) Schematic drawing of affected areas in bilateral MMI (red shaded areas).

penetrator artery disease, dolichoectatic vertebrobasilar arteries, and cardiac embolism [7]. Bilateral MMI may be associated with the anatomical variability of the VBD or perforating branches supplying the medulla. VBD is now well known to cause posterior circulation infarctions independently of atherosclerotic diseases. There are several mechanisms by which VBD triggers posterior circulation ischemia. Blood flow can be bidirectional in dilated arteries, which contributes to the reduction of antegrade flow and thrombus formation. At the same time, the widening of the arteries deforms the perforating artery branches, resulting in reduced blood flow. Decreased blood flow causes thrombus formation. The resulting thrombus may cause occlusion of the perforating branches, leading to synchronized bilateral ischaemic pathologies [4].

In the present case, the brain CT showed an enlarged and elongated vertebral artery pressing on the medulla oblongata, and 3D-TOF MRA showed vertebrobasilar dolichoectasia. Digital subtraction angiography is not recommended due to the risk of stroke in advanced ectatic VBD [14]. CT angiography and magnetic resonance angiography are sufficient for diagnosis [5]. Radiologically, VBD is defined as a basilar artery diameter >4.5 mm and an intracranial vertebral artery (VA) diameter >4.0 mm [15]. Our patient's basilar artery diameter at the pons level and VA diameter measurements in the V4 segment on CT and 3D-TOF MRA met these values (Figure 2). In a previous study investigating the risk factors for posterior circulation infarction in VBD patients, hypertension and intracranial atherosclerosis in the posterior circulation were identified as risk factors for posterior circulation infarction in patients with VBD [15]. In this respect, the patient in the present case had hypertension as a risk factor. Other examinations of this patient regarding the etiology of stroke were normal, suggesting VBD as the cause of stroke.A case series analysis has observed isolated cranial nerve involvement in patients with elongated and tortuous but normal-sized basilar arteries, while multiple neurological deficits have been reported in patients with dilated basilar arteries [14]. The basilar artery of our patient was ecstatic and of normal size, and there were no signs of brain stem compression. Mortality is higher than expected in cases of VBD-induced stroke [4]. Bilateral MMI has a worse prognosis than unilateral MMI, and this rare stroke subtype has a higher mortality rate [9,16]. Old age and severe motor dysfunction at baseline have been reported as predictors of poor prognosis (mRS > 3) in cases of MMI [13]. In a systematic review study, in-hospital mortality of patients with bilateral MMI was reported as 23.8%. It has been reported that the dependency rate of the surviving patients was 61.9% [1]. Pneumonia is a major cause of death in stroke patients. The higher prevalence of pneumonia in dysphagic patients may lead to a poor prognosis [3]. This case also died due to sepsis secondary to pneumonia.

CONCLUSIONS

Vertebrobasilar dolichoectasia is a condition with various neurological complications, especially stroke in the posterior circulation. Bilateral medial medullary infarction is a rare subtype of stroke in the posterior region with a poor prognosis, and vertebrobasilar dolichoectasia may be a cause of this stroke. As in our case, although the heart-shaped appearance in bilateral medial medullary infarction is pathognomonic on imaging, the initial nontypical imaging findings may lead to misdiagnosis or late diagnosis. Therefore, patients may not be able to benefit from the intravenous thrombolysis treatment option, which provides a significant reduction in disability in the acute phase of stroke. In this respect, the early intuition of clinicians is important for early diagnosis of this rare subtype of stroke.

DISCLOSURE STATEMENT

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