

The Heart Shape-Sign in the Brainstem

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Abstract

Case Report

Bilateral medial medullary syndrome is a rare type of stroke, which results due to occlusion of the anterior spinal artery or vertebral artery or its branches. Clinical diagnosis without neuroimaging is very difficult; the characteristic brain MRI finding of “heart appearance” at DWI has been described in bilateral medial medullary infarct. We report the case of 53-year-old man with acute onset quadriparesis. MRI scan of brain, diffusion weighted (DW) imaging revealed heart shaped hyper-intensity areas in the bilateral medial medulla.

Keywords: Bilateral medial medullary syndrome, vertebral artery, Clinical diagnosis, diffusion weighted (DW).

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INTRODUCTION

Bilateral medial medullary infarction (MMI) is a rare stroke syndrome with only a few case reports. Typical signs of MMI include contralateral hemiparesis, bilateral sensory loss, ipsilateral tongue palsy, and contralateral pharyngeal palsy. Patients with bilateral MMI syndrome often present with quadriplegia, bilateral loss of depth sensation, dysphagia, and speech difficulties. The diagnosis of bilateral MMI has become easier with the advent of diffusion-weighted magnetic resonance imaging (DW-MRI) [1, 2]. We report the case of a patient diagnosed with bilateral MMI based on clinical and MRI features.

CASE REPORT

A 53-year-old man, known diabetic and hypertensive, presented to the emergency room with a sudden onset of vertigo, 2 episodes of vomiting, generalized body weakness, slurred speech. The

patient's symptoms worsen gradually over 6 hours with lethargic, severe bulbar symptoms of dysarthria, dysphagia, bilateral facial weakness, dyspnoea, and quadriparesis. A neurological examination revealed lethargic, highly slurred dysarthric speech, and bilateral facial muscle weakness. The strength of the upper and lower limbs was 1/5 (MRC grading). Pinprick and limb proprioception were not assessed due to poor patient awareness.

The patient underwent an MRI C-spine, diffusion weighted (DW) imaging revealed heart shaped hyper-intensity areas in the bilateral medial medulla as well as T2-weighted imaging suggestive of acute medial medullary infarct (Fig 1).

The patient was hospitalized in intensive care and he benefited from thrombolysis but without significant improvement.

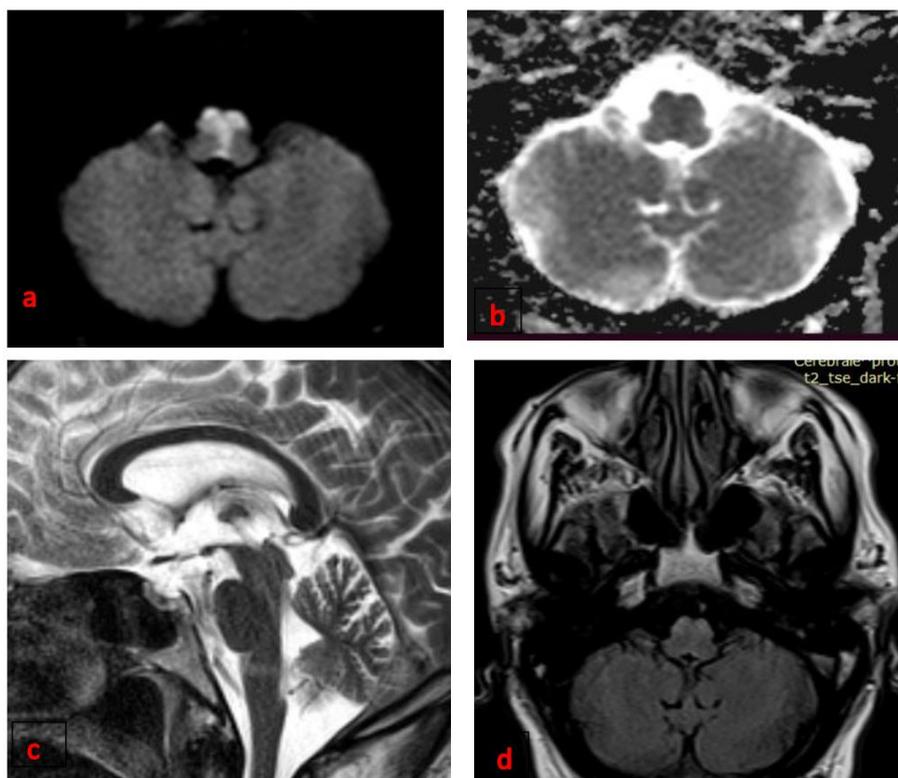


Fig 1: a) Brain diffusion-weighted image on admission showing a “heart-shaped” hyperintense area in the bilateral ventral medulla, characteristic of bilateral MMI. b) Corresponding apparent diffusion coefficient map, showing restricted diffusion within the heart-shaped lesion. c) Sagittal T2 weighted showing a linear hyperintense area, d) axial Flair showing a discrete hyperintense area in the bilateral ventral medulla

DISCUSSION

Medial medullary syndrome is a rare subtype of stroke characterized by paralysis of the ipsilateral hypoglossal nerve, contralateral hemiparesis and impaired deep sensation on the other side. It results from an infarction of the medial area of the medulla oblongata due to obstruction of the vertebral/ anterior spinal artery or their small branches. Medial medullary infarction is rare and accounts for <1% of posterior circulatory infarctions [3]. Bilateral MMI is an even rarer subtype of stroke [4]. The most common symptoms are dysarthria, hypoglossal palsy, flaccid, or spastic quadriplegia [5].

DW-MRI is currently the gold standard for diagnosing acute ischemic stroke and can detect ischemic changes within minutes of symptom onset. Minutes after ischemia, the infarct area appears as a hyperintense area on DWI, with a corresponding hyperintense area on an apparent diffusion coefficient (ADC) map consistent with restricted diffusion [6, 7]. Acute to sub-acute MMI can be differentiated accurately only by MRI. In acute MMI, there will be diffusion restriction with ADC reversal and no abnormality on T2/FLAIR sequences. Sub-acute MMI shows T2 shine through phenomena, no ADC reversal with hyper-intense signal changes on T2/FLAIR [8].

Intravenous/ intra-arterial thrombolysis using recombinant tissue plasminogen activator (r-TPA) may

be beneficial in cases with bilateral MMI. Pfefferkorn *et al.*, have demonstrated that a combination of intravenous thrombolysis with consecutive in endovascular mechanical thrombectomy may be an option in this difficult clinical situation [4].

CONCLUSION

Bilateral MMI is a rare stroke presenting with rapid onset quadriplegia with sensory loss and bulbar weakness which can also be seen in patients with no known co-morbidities, these clinical features should raise suspicion of bilateral MMI. MRI with diffusion-weighted imaging is an accurate way of diagnosing bilateral MMI in the acute setting leading to timely diagnosis and treatment and consequently decreasing the possibility of long-term disability.

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