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Case Report

Bilateral medial medullary stroke: "The heart sign"*,**

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ABSTRACT

Bilateral medial medullary stroke is a very rare condition. It is usually associated with severe motor deficits, dysarthria, sensory symptoms, nystagmus, and respiratory disturbances. The most common etiology is atherosclerotic disease of the vertebral and anterior spinal artery or its branches. We present the case and brain imaging of a 48-year-old man with a bilateral medial medullary stroke and its classic "Heart Sign" in the magnetic resonance imaging. This case highlights the anatomy of the rostral medulla, clinical presentation, etiology, and characteristic radiologic findings of this uncommon type of stroke.

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Introduction

Bilateral medial medullary stroke (BMMS) is a rare condition. Medial medullary structures, including medial lemniscus, medial longitudinal fasciculus, pyramids, and hypoglossal nucleus, are supplied by paramedian branches of the anterior spinal artery. The most common etiology is atherosclerotic disease of the paramedian branches of the anterior spinal artery, the vertebral artery, or the basilar artery. It is associated with severe motor deficits, dysarthria, sensory symptoms, nystagmus, and respiratory disturbances. The unique imaging findings of an acute BMMS is "heart appearance" on the axial view of the different magnetic

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resonance imaging (MRI) sequences, including diffusionweighted imaging (DWI) and apparent diffusion coefficient (ADC).

Case report

A 48-year-old man with history of hypertension and amphetamine use presented to the emergency department after being found disoriented and combative, with a blood pressure of 200/160 mmHg and a urine drug screen test positive for amphetamine. Initial physical examination was significant for disorientation, drowsiness with intermittent agitation, not following commands, but reportedly moving all 4 extremities spontaneously. Patient was started on sedative medications due to his behavior. Twenty-four hours later, after sedation was discontinued, he was noted to have worsening respiratory distress, dysarthria, and spastic quadriparesis, with normal sensory exam. At this point neurology department was consulted. Patient was alert, oriented, following commands, with normal pupils, extraocular movements, facial symmetry, and sensation. He had severe dysarthria, poor tongue movement, and poor oral secretion management. On his extremities, the motor power was 2/5 (per the Medical Research Council Manual Muscle Testing scale), with normal sensation. Based on neurological examination, stroke in the medulla was suspected, however the patient was already out the window for thrombolytic therapy. Brain MRI confirmed an established acute ischemic stroke in the bilateral medial medulla, depicted as the "heart sign" (Fig. 1). Computed tomography angiography of head and neck showed a tortuous right vertebral artery (Fig. 2), but no occlusion. Atherosclerotic disease was presumed to be the main etiology of the infarct given patient's risk factors and vertebral artery findings. Hypotension and cardio-embolism were ruled out, and vasospasm was also deemed to be less likely given the uncommon location of the infarct and angiographic findings. Patient was started on aspirin, atorvastatin, and aggressive physical therapy.

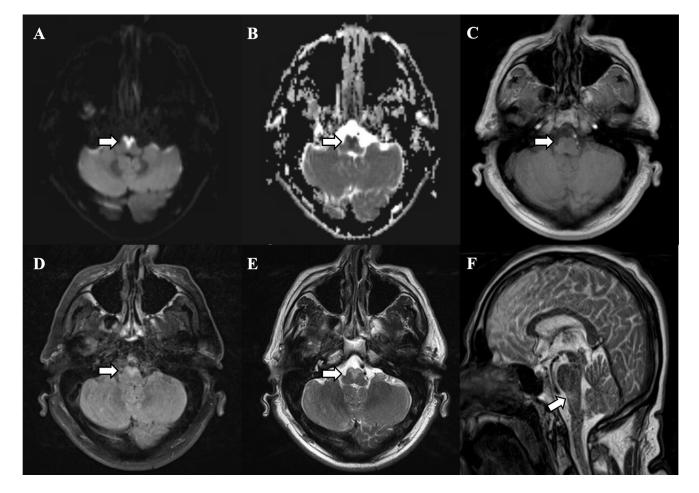


Fig. 1 – MRI of the brain at the level of the medulla, showing signal changes in the rostral medulla, including the pyramids and the medial region, bilaterally. These changes give rise to the classic "Heart Sign" in axial view (arrows A-E), characteristic of acute BMMS. (A) DWI; (B) ADC demonstrate restricted diffusion (arrow); (C) T1 sequence with faint hypointense signal (arrow); (D) fluid attenuated inversion recovery; (E) T2 sequences show hyperintense signal at the same level (arrow) and (F) Sagittal view of T2 sequence depicting hyperintense signal in the medulla (arrow).

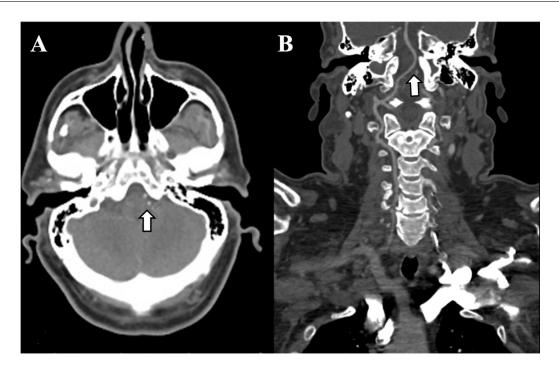


Fig. 2 – CTA of the head, axial view (A), and right vertebral artery curved planar reformatted (CPR), coronal view (B) showing the position of the right and left vertebral artery (arrow), at the level of the medullary stroke. A dominant tortuous right vertebral artery crosses the midline towards the left side at the level of the bilateral infarction (B).

Discussion

Bilateral medial medullary strokes are very rare; they are generally secondary to atherosclerotic disease involving the vertebral arteries, the basilar arteries, the spinal anterior artery, or its branches [1]. Like in our case, the radiological finding of vertebral artery dolichoectasia has also been associated to BMMS [1]. The "heart sign" is a classic radiological finding in bilateral medial medullary infarcts given the symmetric affection of the pyramids anteriorly [2,3]. Both pyramids, divided by the anterior median fissure, give the "heart appearance" in the axial view of the MRI (Figs. 1A-E). Signal changes can also be seen on the sagittal view right below the pontomedullary junction (Fig. 1F). Only two-thirds of patients with BMMS will present with the "heart sign" in the first 24 hours [4]. Restricted diffusion is seen on the DWI/ADC sequences (Figs. 1A and B). Depending on the timing of the MRI and onset of symptoms, some signal changes might be apparent also in the T1, T2, and fluid attenuated inversion recovery sequences (Figs. 1C-F). A typical MRI findings in BMMS have also been described, including faint or delayed DWI changes [5,6] or an incomplete heart sign in 2 consecutive axial sections [7]. Other names given to the "heart appearance" are "Airpod sign" [8] or "Letter Y sign" [9].

Ischemia of the rostral medulla, particularly in the pyramids, causes prominent motor deficits, but can also be associated with dysarthria and respiratory distress due to the proximity of the hypoglossal nerve and medullary respiratory center, respectively [2,3], such as in our patient. Sensory deficits and nystagmus have also been commonly documented, secondary to involvement of structures in the medial medulla, including the medial lemniscus and medial longitudinal fasciculus, respectively [2].

This report serves to raise awareness among healthcare providers about clinical-radiological correlation, and the typical imaging findings in a very rare type of stroke, BMMS, as prompt recognition and treatment might save a patient from long term neurologic sequela.

Patient consent

I confirm that I acquired written informed consent from the patient, for publication of his case, for academic purposes.

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