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RESEARCH ARTICLE

ACUTE QUADRIPARESIS: GBS OR BILATERAL MEDULLARY INFARCT – A NEUROLOGIST'S DILEMMA

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ABSTRACT

Bilateral medial medullary stroke is a very rare type of stroke, with catastrophic consequences. Early diagnosis is crucial. Here, we report a case presenting with acute vertigo, progressive generalized weakness of all 4 limbs, dysarthria, and decreased single breath count. Brain magnetic resonance imaging (MRI) that was done showed characteristic "heart appearance" shape at diffusion weighted (DWI), and confirmed bilateral medial medullary stroke. Other possibilities were excluded by lumbar puncture and MRI of cervical spine with screening of whole spine. Retrospectively, a vague-defined hyperintense linearDWI signal at midline was noted in the first brain MRI. Because of the symmetric and midline pattern of this abnormal signal and similarityto an artifact, some radiologists or neurologists may miss this type of stroke. Radiologists and neurologists must co relate clinical and MRI findings of this rare type of stroke, where early treatment could make a difference in patient outcome and therapy. The abnormal DWIsignal in early stages of this type of stroke may not be a typical "heart appearance" shape, and other variants such as small dot or linear DWI signal at midline must be recognized as early signs of stroke. Also, MRI of cervical spine may be helpful if there is suspicion of spinal pathology.

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INTRODUCTION

Bilateral medial medullary stroke is very rare, and clinical diagnosis without neuroimaging is very difficult (Kumral *et al*., 2002). Brainstem encephalitis and Guillain-Barre's syndrome (GBS) can present similarly (Ma *et al*., 2011). Despite a huge progress in MRI technology, still human factor and experience can determine correct interpretation. Here, we discuss a clinical case and MRI findings of a patient with this diagnosis.

Case Presentation

A 59-year-old diabetic male patient, right-handed, presented with acute vertigo dysarthria and quadriparesis. Past medical history was remarkable for hypertension & diabetes on treatment and moderate alcohol consumption.

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He was on regular medicationat home with adequate glycemic control. Initial examination in the hospital showed bilateral nystagmus.The first MRI of brain was done showed medullary infarct (Figure 1), with a faint linear signal at DWI at midline medulla.Over duration his condition deteriorated, and he presented to our hospital, with generalized weakness, falls, slurred speech, respiratory failure, and dysphagia.A repeat MRI Brain C spine & screening of whole spine showed a bilateral medullary infarct in the typical heart shaped infarct in DWI. Shortly after examination, he developed respiratory failure secondary to aspiration and was intubated. His evaluation after extubation showed severe dysarthria with normal comprehension. His cranial nerves revealed direction changing gaze evoked horizontal nystagmus.No facial palsy noted, and extraocular movement still was full. Decreased gag reflex was noted. Motor exam showed diffuse bilateral weakness of both upper more than lower extremities at the range of 2-3/5, along with decreased tone and absent deep tendon reflexes everywhere, resembling/ similar to an acute spinal cord shock state/areflexia of GBS.

Figure 1. First MRI done in acute phase showing a small linear abnormal DWI signal at midline

He had mute Babinski response bilaterally. Figure 1: MRI of brain and DWI at presentation. Abnormal signal at DWI, a midline dot-linear signal, was similar to an artifact. The differential diagnosis prior to imaging wereFor the possibility of GBS and brainstem encephalitis, lumbar puncture was performed which showed normal cell, protein, and glucose. Blood and CSF cultures were negative. Since GBS still could not be excluded, Nerve conduction study was done which showed polyradiculopathy which was secondary to the long term diabetes as presumed, he was started on antiplatelets RT feeding and physiotherapy keeping stroke as the diagnosis of the acute presentation. There was progressive weakness to the point of quadriplegic state in less than two days. Still he was able to communicate with eye movements and blinking and slurred speech. He was given vigorous physiotherapy and bed care and his condition improved but he developed an aspiration pneumonia which responded to antibiotics. In the ward he developed another episode respiratory distress and was shifted to ICU .In the ICU there was hypotension. The patient condition deteriorated and he was intubated & later taken up for tracheostomy. With regular physiotherapy & care he was taken off the ventilator; gradually the tracheostomy was also closed and his condition improved and was discharged.

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DISCUSSION

Bilateral medial medullary stroke is a very rare type of stroke. The most common symptoms are weakness, dysarthria, hypoglossal palsy, flaccid, or spastic quadriplegia. It has generally a poor prognosis (Kumral *et al*., 2002; Kleinert *et al*., 1998; Jagiella, 1989). The characteristic brain MRI finding of "heart appearance" at DWI has been described in multiple case reports (Kumral *et al*., 2002; Kleinert *et al*., 1998; Jagiella, 1989). In this case, the first MRI (Figure 1) done in acute phase did show a small linear abnormal DWI signal at midline .Considering interventions such as thrombolytic in the acute phase, it is important to diagnose any stroke as early as possible, MRI of cervical spine with whole spine screening was normal. Figure 2: Second brain MRI, DWI shows characteristic "heart appearance" sign suggestive of bilateral medial medullary infarct. Normal MRA of brain may suggest small vessel disease of one of the paramedianperforatings (from vertebralor anterior spinal arteries).

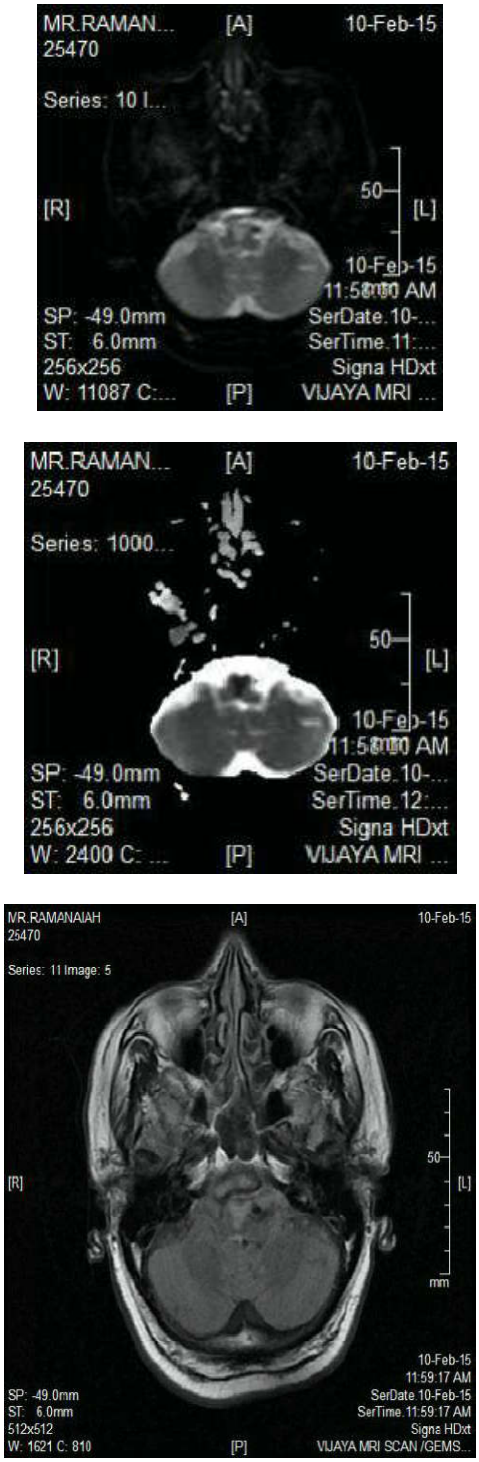

Figure 2. Second brain MRI, DWI shows characteristic "heart appearance" sign suggestive of bilateral medial medullary infarct

It is possible if one paramedian artery is supplying both pyramids in rare cases. Brainstem encephalitis is also very rare and has a broad range of differential diagnosis including infectious and autoimmune causes. It has features such as ophthalmoparesis, ataxia, weakness, and upper motor neuron signs. In this case, negative CSF, absence of ophthalmoplegia, and MRI findings are all against brainstem encephalitis. GBS similarly can mimic the presentation, particularly with diffuse areflexia and progressive weakness, and was excluded by MRI findings (Ma *et al*., 2011). MRI of spine ruled out spinal pathologies.Overall outcome of this type of stroke is not good withsevere morbidity and mortality, and early diagnosis based on combination of clinical and MRI findings by both neurologist and radiologist is critical.

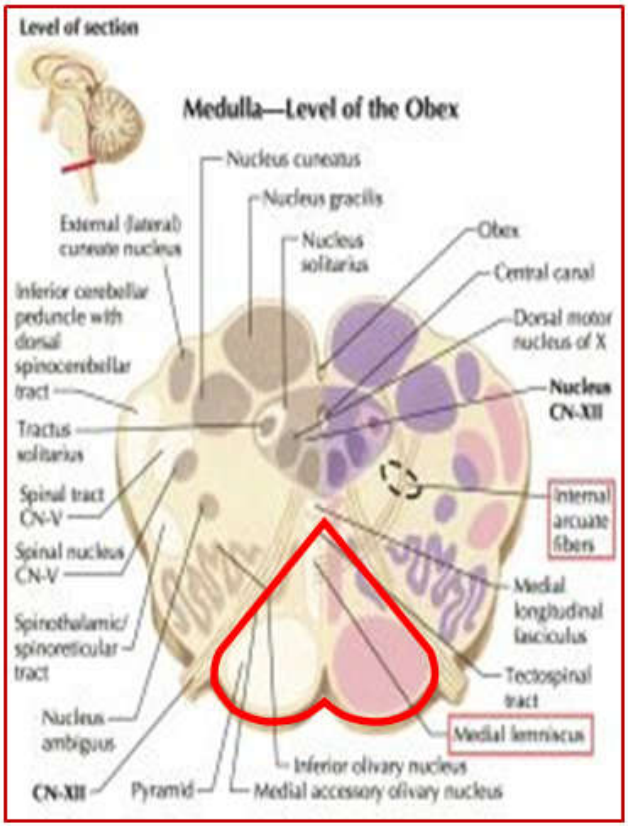

Figure 3. Shows the diagram of medulla and "heart appearance" sign suggestive of bilateral medial medullary infarct

MRI findings particularly in the first few hours may not be typical, and along with clinical presentation neurologists and radiologists should have a high index of suspicion. Considering the location of infarct and the confusing presentation we look at a short overview of anatomy of medulla the medulla oblongata (or medulla) is the part of the brainstem that is situated between the pons and the spinal cord.

The medulla contains the cardiac, respiratory, vomiting and vasomotor centers and therefore deals with the autonomic (involuntary) functions of breathing, heart rate and blood pressure.

The medulla can be thought of as being in two parts:

- an open part or superior part where the dorsal surface of the medulla is formed by thefourth ventricle.
- a closed part or inferior part where the fourth ventricle has narrowed at the obex in the caudal medulla, and surrounds part of the central canal.

External surfaces

The anterior median fissure contains a fold of pia mater, and extends along the length of the medulla oblongata. It ends at the lower border of the pons in a small triangular area, termed the foramen cecum. On either side of this fissure is a raised area termed the pyramid of medulla oblongata. The pyramids house the pyramidal tracts–the corticospinal and the corticobulbar tracts of the nervous system. At the caudal part of the medulla these tracts cross over in the decussation of the pyramids obscuring the fissure at this point. Some other fibers that originate from the anterior median fissure above the decussation of the pyramids and run laterally across the surface of the pons are known as the anterior external arcuate fibers. The region between the anterolateral and posterolateral sulcus in the upper part of the medulla is marked by a pair of swellings known as olivary bodies (also called *olives*). They are caused by the largest nuclei of the olivary bodies, the inferior olivary nuclei. The posterior part of the medulla between the posterior median sulcus and the posterolateral sulcus contains tracts that enter it from the posterior funiculus of the spinal cord. These are the gracile fasciculus, lying medially next to the midline, and the cuneate fasciculus, lying laterally. These fasciculi end in rounded elevations known as the gracile and the cuneate tubercles. They are caused by masses of gray matterknown as the gracile nucleus and the cuneate nucleus. The perikarya or cell bodies in these nuclei are the second-order neurons of the posterior column-medial lemniscus pathway, and their axons, called the internal arcuate fibers or fasciculi, decussate from one side of the medulla to the other to form the medial lemniscus. Just above the tubercles, the posterior aspect of the medulla is occupied by a triangular fossa, which forms the lower part of the floor of the fourth ventricle. The fossa is bounded on either side by the inferior cerebellar peduncle, which connects the medulla to the cerebellum. The lower part of the medulla, immediately lateral to the cuneate fasciculus, is marked by another longitudinal elevation known as the tuberculum cinereum. It is caused by an underlying collection of gray matter known as the spinal trigeminal nucleus. The gray matter of this nucleus is covered by a layer of nerve fibers that form the spinal tract of the trigeminal nerve. The base of the medulla is defined by the commissural fibers, crossing over from the ipsilateral side in the spinal cord to the contralateral side in the brain stem; below this is the spinal cord.

Blood supply

Blood to the medulla is supplied by a number of arteries.

- Anterior spinal artery: This supplies the whole medial part of the medulla oblongata.
- Posterior inferior cerebellar artery: This is a major branch of the vertebral artery, and supplies the posterolateral part of the medulla, where the main sensory tracts run and synapse. It also supplies part of the cerebellum.
- Direct branches of the vertebral artery: The vertebral artery supplies an area between the other two main arteries, including the solitary nucleus and other sensory nuclei and fibers.

Conflict of Interests

The author declare that there is no conflict of interest.

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