A rare case of bilateral medullary syndrome: Heart-shaped infarct in MRI brain

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WORD COUNT 1180
TIME SUBMITTED 20-MAY-2024 06:07PM
PAPER ID 109088252

Word Count – Words: 1,180

A rare case of bilateral medullary syndrome: Heart-shaped infarct in MRI brain

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ABSTRACT

Bilateral medullary syndrome (BMS) is an exceptionally rare neurological disorder characterized by a myriad of symptoms resulting from lesions affecting the dorsal medial part of the medulla oblongata. Here, we present the case of a 70-year-old male with systemic hypertension who developed weakness in his right upper and lower limbs, slurred speech, and swallowing difficulties. MRI brain imaging revealed a heart-shaped infarct indicative of bilateral medullary infarction, with subsequent progression to quadriplegia and respiratory distress. Despite medical intervention, including antiplatelets and statins, the patient's condition deteriorated, leading to his demise. Discussion revolves around the pathophysiology, clinical manifestations, and prognosis of BMS, emphasizing the importance of early recognition and management to mitigate associated complications and improve outcomes.

INTRODUCTION

Medullary syndrome is a rare neurological disorder with an incidence rate of 0.5% - 1.5%, and bilateral medullary syndrome is even rarer. This syndrome presents a complex array of symptoms, including horizontal gaze palsy, convergence-retraction nystagmus, dysphagia, dysarthria, hemiparesis, and hyperreflexia, among others (1,2). Understanding Medullary Syndrome involves a nuanced understanding of the neurological deficits resulting from lesions affecting the dorsal medial part of the medulla oblongata, which houses vital structures responsible for regulating autonomic functions such as respiratory and cardiovascular control (3). A hallmark of this syndrome is the V-shaped or heart-shaped appearance observed in MRI brain scans (4,5). Here we discuss a case of bilateral medullary syndrome.

CASE DETAILS

A 70-year-old male, previously diagnosed with systemic hypertension, presented with complaints of weakness in the right upper and lower limbs following an episode of dizziness. He also had a history of slurred speech and difficulty in swallowing. Upon arrival, he exhibited consciousness but lacked proper orientation. A Ryles tube was inserted as a precaution against aspiration. Subsequent MRI brain imaging revealed a distinct area of diffusion restriction with corresponding low ADC and T2/FLAIR hyperintensity affecting bilateral pyramids and the central medulla, indicative of a heart-shaped infarct in the medulla, thus pointing to bilateral medullary infarction (6,7). An MR angiogram indicated features suggestive of flow restriction in both vertebral arteries and the basilar artery (8,9).

Throughout the hospitalization period, the weakness progressed, affecting all four limbs and ultimately resulting in quadriplegia. Subsequently, due to ongoing microaspiration, the patient developed respiratory distress secondary to aspiration pneumonia, necessitating intubation. Treatment encompassed antiplatelets, statins, and other supportive measures, yet despite medical efforts, the patient's condition deteriorated, leading to his demise.

Figure 1,2: Diffusion-weighted image with apparent diffusion coefficient map showed restricted diffusion within the lesion.

Figure-1 **Figure-2**

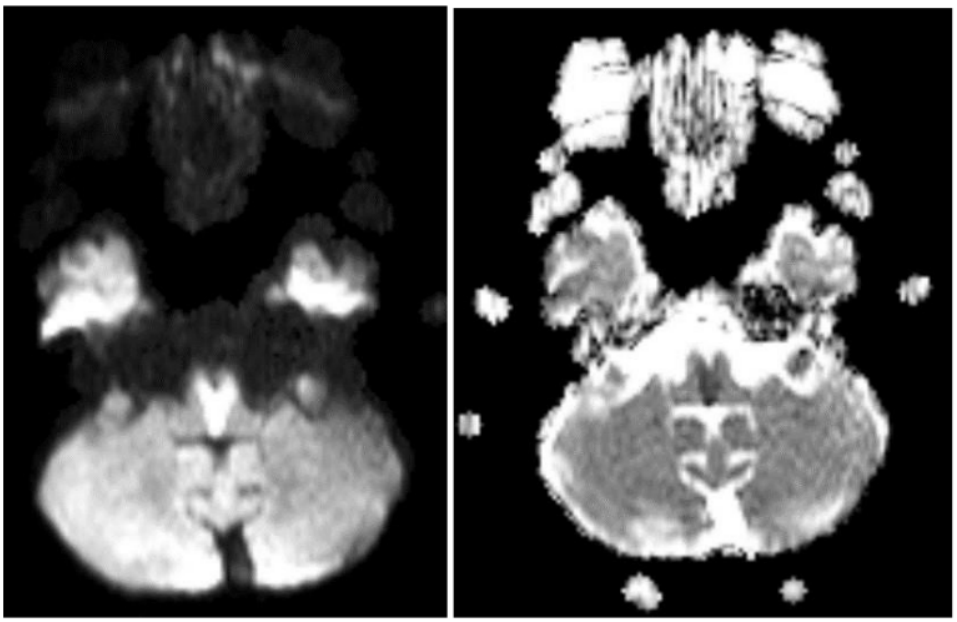


Figure 3: Axial FLAIR (fluid-attenuated inversion recovery) image showed bilateral hyperintensities in the medulla involving pyramids.

Figure-3

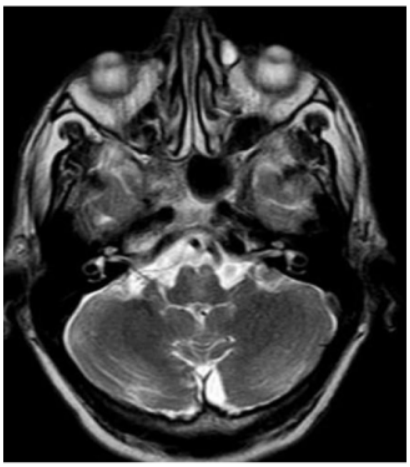
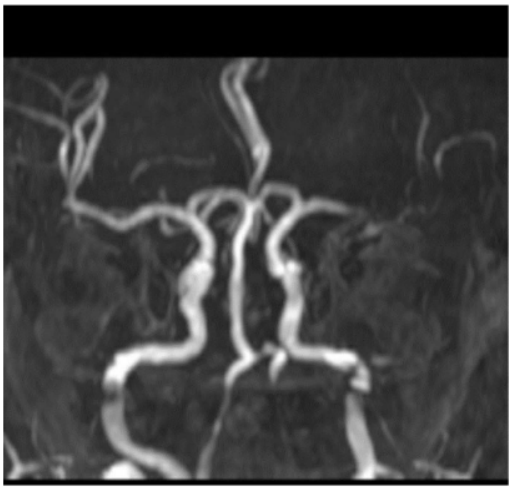


Figure 4: MR angiography time of flight image showed flow defect and irregularities of the bilateral vertebral arteries, basilar artery, and internal carotid arteries.

Figure-4



DISCUSSION

Medullary syndrome is a rare neurological disorder with an incidence rate of 0.5% - 1.5%, and bilateral medullary syndrome is even rarer (2). This syndrome presents a complex array of symptoms, including horizontal gaze palsy, convergence-retraction nystagmus, dysphagia, dysarthria, hemiparesis, and hyperreflexia, among others (3). Understanding Medullary Syndrome involves a nuanced understanding of the neurological deficits resulting from lesions affecting the dorsal medial part of the medulla oblongata, which houses vital structures responsible for regulating autonomic functions such as respiratory and cardiovascular control (4). A hallmark of this syndrome is the V-shaped or heart-shaped appearance observed in MRI brain scans (5). The medulla oblongata primarily receives its blood supply from the vertebral arteries, which merge to form the basilar artery. The basilar artery then branches into several vessels, including the anterior spinal artery, supplying the anterior portion of the medulla. Additionally, perforating branches from the vertebral and basilar arteries contribute to the medulla's blood supply (6,7). Bilateral Medial Medullary Syndrome (BMMS) typically results from bilateral infarction or ischemia affecting the paramedian branches of the vertebral arteries or the anterior spinal artery, leading to compromised blood flow to the medial medullary region (8,9).

BMMS presents with a spectrum of neurological deficits stemming from specific anatomical structures within the medulla. Common clinical features include contralateral hemiparesis or hemiplegia affecting the limbs and lower face due to corticospinal tract involvement, ipsilateral tongue weakness or deviation due to hypoglossal nucleus or nerve involvement, and contralateral loss of proprioception and vibration sense due to involvement of the medial lemniscus (10,11). Patients may also exhibit dysarthria, dysphagia, and respiratory compromise depending on the extent of medullary involvement. The prognosis of BMMS varies based on the severity of neurological deficit and the presence of associated complications such as aspiration pneumonia or respiratory failure. While some patients may experience partial recovery with rehabilitation, others may face long-term disability or mortality (12). The management of BMMS focuses on addressing acute management,

preventing complications, implementing rehabilitation therapies, and devising long-term management strategies.

CONCLUSION

Bilateral medullary syndrome (BMS) presents a complex and challenging clinical scenario, as illustrated by the case discussed herein. This neurological disorder, though exceedingly rare, manifests with a wide spectrum of symptoms resulting from lesions affecting vital structures within the medulla oblongata. Despite advancements in medical imaging and therapeutic interventions, the prognosis for BMS remains guarded, often leading to significant disability or mortality. The case underscores the importance of a multidisciplinary approach involving neurology, radiology, and critical care in the management of such cases. Furthermore, it highlights the imperative for heightened vigilance among clinicians to promptly recognize and address the myriad neurological deficits associated with BMS, thereby optimizing patient care and outcomes. Further research is warranted to elucidate the underlying mechanisms of BMS and explore novel therapeutic avenues aimed at improving prognosis and quality of life for affected individuals.

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References:

Lo YL, Teo M, Tjia HT. Bilateral medial medullary infarction: report of a patient with "heart appearance" on diffusion-weighted MRI. J Neurol Sci. 2004;219(1-2):117-20.

Kim JS. Medial medullary infarction: clinical, imaging, and outcome study in 86 consecutive patients. Stroke. 2003;34(4):1005-10.

Pula JH, Marill KA. Medullary infarction presenting with palatal myoclonus. West J Emerg Med. 2012;13(1):104-6.

Kumar S, Selim MH, Caplan LR. Medical complications after stroke. Lancet Neurol. 2010;9(1):105-18.

Milandre L, Joux J, Olindo S, Sainte-Rose C, Signate A, Cabre P. Bilateral medial medullary infarction: three cases. Cerebrovasc Dis. 2007;24(1):104-6.

Amarenco P, Caplan LR, Bogousslavsky J, Donnan GA, Hennerici MG. Classification of stroke subtypes. Cerebrovasc Dis. 2009;27(5):493-501.

Choi JY, Kang DW, Kim JS. Diffusion-weighted MRI in medial medullary infarction. J Neurol Sci. 2001;193(2):137-45.

Caplan LR. Posterior circulation ischemia: then, now, and tomorrow. Stroke. 2000;31(8):2011-23.

Marti-Fabregas J, Piles S, Escudero D, Santamarina E, Guardia E, Martí-Vilalta JL. Bilateral paramedian medullary infarction: a heart-shaped lesion. Arch Neurol. 1999;56(4):478-80.

Kim JS, Choi-Kwon S. Disabling complications of acute ischemic stroke. J Stroke. 2020;22(2):176-88.

Meyer MA. Medullary infarction. Neuroimaging Clin N Am. 2011;21(1):145-57.

Kang DW, Lee SH, Lim C, Yang Y, Kim JS. Medial medullary infarction: clinical, imaging, and outcome study. J Neurol. 2003;250(1):1341-8.