Heart in the Brain

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A 52-year-old man with a 5-year history of diabetes mellitus and chronic renal disease presented with sudden onset left upper limb weakness and numbness at 5 pm, which progressed to quadriplegia by the next day at 2:30 am. He had dysarthria at admission. There were no sensory symptoms in the lower limbs. There were no bladder symptoms. Examination showed bilateral tongue weakness and quadriplegia, with the left side more affected than the right. Reflexes were sluggish bilaterally. Plantars were extensor bilaterally. The sensory system was normal in all four limbs, including joint position sense and vibration. Diffusion-weighted magnetic resonance imaging (MRI) of the brain showed diffusion restriction in the bilateral medial medulla simulating a heart sign, suggestive of a bilateral medial medullary infarct (Figs 1 and 2). MR angiography showed left vertebral artery stenosis. He was treated as per stroke protocol and made partial recovery at follow-up.

Medial medullary infarction encompasses <1–1.5% of ischemic strokes, and bilateral medial medullary infarction (BMMI) is very rare. BMMI usually presents with tongue weakness, tetraplegia, and sensory loss, with or without respiratory failure. It can be confused with Guillain–Barré syndrome when there are no sensory symptoms or signs. The medullary pyramids, medial longitudinal fasciculus (MLF), medial lemniscus, and hypoglossal nucleus are nourished by paramedian branches of the anterior spinal artery, forming an airpod-shaped vascular territory. The most common etiology of BMMI is atherosclerosis affecting the vertebral and anterior spinal artery or its branches. The heart sign, airpod sign, or letter Y sign is classically seen in bilateral medial medullary infarct and is often due to type 2a anterior spinal artery occlusion, where a single anterior spinal artery comes off one vertebral artery, and occlusion causes bilateral medial medullary infarction.[1–4] The heart sign is seen in only two-thirds of patients with BMMI in the first 24 hours.

The heart appearance sign can also be seen in anteromedial infarction of the pons bilaterally and is attributed to thrombosis of the paramedian and short circumferential pontine arteries bilaterally, supplying the anteromedial pons.[5] It has also been reported in Wernekink's commissure syndrome due to infarct at the caudal paramedian midbrain tegmentum,[6] and in vitamin B_{12} deficiency at the rostral medulla in coronal MRI images.[7]

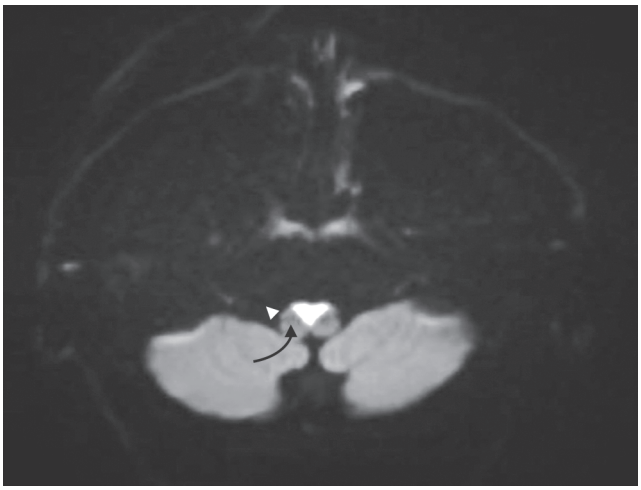

Fig. 1: Axial diffusion-weighted MRI of the brain showing heart-shaped diffusion restriction in the ventral medulla (heart sign)

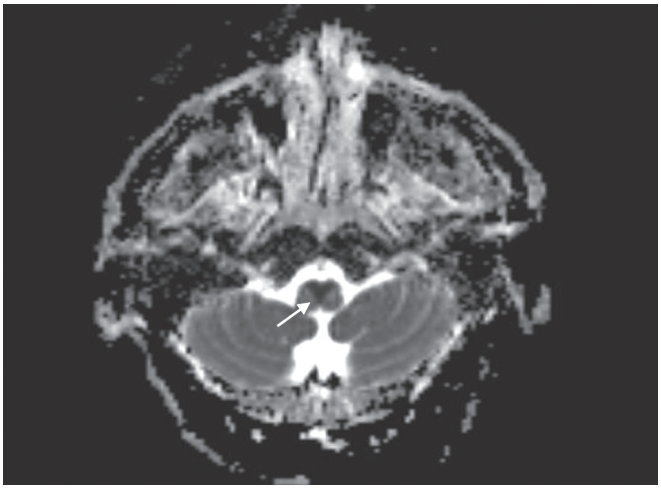

Fig. 2: Magnetic resonance imaging brain axial apparent diffusion coefficient image showing heart-shaped hypointensity in the ventral medulla

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References

1. Maeda M, Shimono T, Tsukahara H, et al. Acute bilateral medial medullary infarction: a unique 'heart appearance' sign by diffusion-weighted imaging. Eur Neurol 2004;51(4):236–237.
2. Agarwal S, Raz E, Yaghi S. Bilateral medial medullary infarction: the airpod sign. Stroke 2023;54(4):e157–e158.

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3. Al Tijani Mohammed M, Jalali A, Al Balushi A. et al. Bilateral medial medullary infarction resembling the letter "Y." Acta Neurol Belg 2023.
4. Er U, Fraser K, Lanzino G. The anterior spinal artery origin: a microanatomical study. Spinal Cord 2008;46:45–49.
5. Zhou Z, Wu Y, Wu W, et al. Giant "heart appearance-like sign" on MRI in bilateral ponto-medullary junction infraction: case report. BMC Neurol 2020;20(1):107.
6. Zhou C, He Y, Chao Z, et al. "The "heart appearance" sign on MRI of Wernekink's commissure syndrome caused by bilateral caudal paramedian midbrain infarction. Neurol Sci 2018;39:587–589.
7. Madhusudanan M, Muthukumarasamy B. "Heart appearance sign" in vitamin B12 deficiency. Neurol India 2023;71(3):600–601.