A Rare but Disabling Stroke

A 59-year-old woman presented with recurrent vomitting, problem with speech and swallowing difficulty for 2 days with sudden onset of bilateral upper and lower limb weakness. She did not have dizziness, vertigo, headache or neck pain. There was no recent history of head and neck trauma. She had an ischaemic stroke in 2016 with weakness in the left face, left upper limb and left lower limb. She was diagnosed with lacunar stroke but had full neurological recovery. She had a history of hypertension and diabetes mellitus with poor glycaemic control.

On examination, she was fully conscious but had dysphonia, absent gag reflex and weak tongue movement. Her bilateral upper limb power was 3/5 proximally and 4/5 distally while her bilateral lower limb power was 1/5 proximally and 2/5 distally. Tone was normal bilaterally. She had bilateral extensor plantar reflexes with no sensory loss, ophthalmoplegia or facial asymmetry.

What subtype of stroke did this patient have and what was the anatomical variant present in magnetic resonance angiography (Fig. 1)?

- A. Lateral medullary infarct with hypoplastic left vertebral artery
- B. Lateral medullary infarct with hypoplastic right vertebral artery
- C. Medial medullary infarct with hypoplastic left vertebral artery
- D. Right medial medullary infarct with hypoplastic right vertebral artery
- E. Bilateral medial medullary infarct with hypoplastic right vertebral artery



Fig. 1. Magnetic resonance angiography of artery.

Discussion

Option E is correct because brain magnetic resonance image (MRI) showed bilateral medial medullary infarct. Additionally, magnetic resonance angiography showed hypoplastic right vertebral artery that terminated as posterior inferior cerebellar artery (PICA).

Options A and B are incorrect because brain MRI showed medial medullary infarct and not lateral medullary infarct. Option C is also incorrect because the patient had hypoplastic right vertebral artery and not hypoplastic left vertebral artery. Option D is incorrect since the patient had bilateral medial medullary infarct but no right medial medullary infarct.

Medial medullary infarct (MMI) comprises the clinical triad of contralateral hemiparesis, contralateral loss of deep sensation and ipsilateral lingual palsy (Dejerine's triad).^{1,2} It is a rare subtype of stroke with an incidence of 0.5% to 1.5%.^{1,2} Bilateral MMI is even rarer.^{1,2}

Patients with bilateral MMI usually present with acute onset of quadriparesis or quadriplegia, bulbar palsy, lingual palsy and bilateral sensory loss with or without respiratory failure.³ In patients with lateral medullary syndrome, they present with ipsilateral Horner's syndrome, ipsilateral facial sensory loss and contralateral hemisensory loss. This patient did not have dizziness, vertigo, headache, neck pain or trauma to the neck and head which suggested arterial dissection. With a diversity of clinical symptoms, diagnosis is often delayed and patients are often treated for other more common diseases.

With advancements in brain neuroimaging techniques such as MRI, bilateral MMI can be diagnosed earlier and more easily. In acute bilateral MMI, MRI shows a characteristic "heart-shaped appearance" in the ventral medulla (Fig. 2).³ The probable stroke mechanism in this patient was occlusion of perforators (from vertebral artery or basilar artery) or atherosclerosis of vertebral artery. Another possible stroke mechanism described in the literature is anterior spinal artery occlusion.³ The patient had bilateral MMI but not unilateral MMI because the anatomical variation of hypoplastic right vertebral artery terminated as PICA. It was not occlusion because the right vertebral artery can be visualised but it was small with a diameter of \leq 2 mm. Additionally, the larger left vertebral artery was the dominant artery.

Answer: E



Fig. 2. A) MRI of heart-shaped appearance (arrow) in ventral medulla in diffusion-weighted image; B) Apparent diffusion coefficient sequence; and C) T2-weighted sequence. MRI: Magnetic resonance image.

PICA termination of vertebral artery (PICA-VA) happens when the vertebral artery does not communicate directly with the basilar artery and ends in PICA instead (Fig. 3).⁴ PICA-VA is a variant of vertebral artery hypoplasia and is present in 2% of population.⁴ In a recent study, the prevalence of PICA-VA in patients with posterior circulation stroke was shown to be significantly higher than normal individuals.⁴ Sarah and colleagues reported a patient who had aplasia of left vertebral artery and left PICA that originated from the right vertebral artery and crossed the midline and supplied the contralateral side of the cerebellar hemisphere.⁵ Figure 4 illustrates the arteries and PICA-VA.

Patients with bilateral MMI have a poor prognosis and high in-hospital mortality.² Most of them are dependent on follow-up.³ Knowledge of stroke presenting with features



Fig. 3. A) Right PICA (white arrow) seen in brain MRI and B) MRA. MRA: Magnetic resonance angiography; MRI: Magnetic resonance image; PICA: Posterior inferior cerebellar artery.



Fig. 4. PICA termination of vertebral artery. PICA: Posterior inferior cerebellar artery.

such as MMI is associated with early health-seeking practice.⁶ In conclusion, there should be a high index of suspicion in patients with bilateral MMI presenting with quadriparesis and bulbar palsy since this condition is treatable and morbidity or mortality can be avoided.

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