

CASE REPORT

An unexpected Holter monitor result: multiple sinus arrests in a patient with lateral medullary syndrome

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SUMMARY

A 52-year-old man presented with vomiting, dysphagia, left-sided ataxia and dissociated sensory loss. Diffusion-weighted MRI showed evidence of acute infarct in the left lateral medulla and left medial cerebellar hemisphere, probably secondary to thromboembolism from left vertebral artery dissection. While making an uneventful recovery as an inpatient, a routine 24 h ECG was performed 2 weeks after the stroke to investigate possible paroxysmal atrial fibrillation. The recording instead revealed 56 asymptomatic episodes of sinus arrest, necessitating implantation of a permanent pacemaker to prevent sudden cardiac death. The medulla contains key structures involved in autonomic regulation, including the dorsal vagal nucleus and the nucleus tractus solitarius. Acute infarction may disrupt cardiac autonomic regulation pathways, resulting in altered parasympathetic and sympathetic outflow to the sinoatrial and atrioventricular nodes, with potentially life-threatening effects.

BACKGROUND

The complex structure of the brainstem leads to various clinical syndromes when particular sites are affected. Longitudinal lateral brainstem structures include (1) the spinothalamic pathway, responsible for pain and temperature sensation in the body contralateral to the site of the brainstem lesion, (2) the sensory trigeminal nerve nucleus, responsible for sensation for the ipsilateral face, (3) the spino-cerebellar tract, causing ipsilateral cerebellar signs when damaged and (4) the sympathetic pathway, which gives rise to an ipsilateral Horner's syndrome when affected. The medulla contains the lower cranial nerves IX, X and XII, which when damaged, may result in hoarseness, dysarthria, dysphagia and impaired throat sensation and palatal elevation.¹ In relation to cardiac autonomic control, the medulla contains key structures involved in both sympathetic and parasympathetic outflow, including the nucleus tractus solitarius (NTS), the dorsal vagal nucleus, the nucleus ambiguus and the intermediate reticular zone.²

This case of a patient with classical features of lateral medullary syndrome (LMS) and his subsequent cardiac investigations serve to highlight the fact that arrhythmias not only lead to stroke but can result from cerebral pathology itself, through an insult to cerebral autonomic cardiac innervation.

CASE PRESENTATION

A 52-year-old Afro-Caribbean man presented with sudden onset of dizziness, reporting the impression

of objects 'dancing in front of his eyes', vomiting, slurred speech, unsteadiness and falling towards the left, associated with left-sided headache and neck pain. His symptoms had commenced the night before, but the patient thought they were due to gastroenteritis and only sought medical attention when they persisted the following morning. He had a background of hypertension and type II diabetes, was previously independent and worked as a cleaner.

Neurological examination revealed left-sided ptosis and miosis, with horizontal nystagmus in all directions of gaze, most apparent on left lateral gaze. He had dysarthria, dysphonia, left-sided tongue deviation, impaired palatal elevation and difficulty swallowing with nasal regurgitation. There was dysidiadochokinesia and dysmetria on the left side, with severe truncal ataxia and a tendency to fall towards the left. Sensory examination revealed reduced temperature and pinprick sensation on the left side of his face and the right side of his body.

INVESTIGATIONS

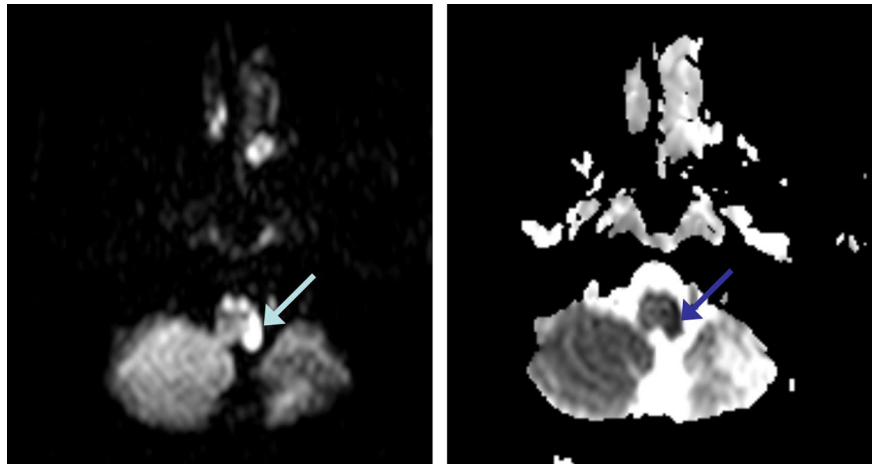
CT scan of the head at the time of presentation showed evidence of hypertensive arteriopathy and a subacute left cerebellar infarct, with a bright left vertebral artery. CT angiogram showed the absence of flow in left vertebral artery below the level of the posterior-inferior cerebral artery.

A subsequent MRI scan revealed restricted diffusion, in keeping with acute infarction, in the left lateral medulla oblongata and left medial cerebellar hemisphere (figure 1). There was T2-weighted/fluid-attenuated inversion recovery hyperintense change, in keeping with subacute infarction, in the lateral left cerebellar hemisphere (figure 2). A fat-suppressed MRI neck scan showed high signal in the entire left vertebral artery, from lowest level imaged at the carotid bifurcation to the vertebral basilar junction. The abnormal signal could have been consistent with slow flow, thrombus or dissection (figure 3). Given the history of neck pain at the time of the event, the patient was treated as having a spontaneous vertebral dissection. He was commenced on anticoagulation, initially in the form of subcutaneous low-molecular-weight heparin, and then warfarin, with target international normalised ratio 2–3, planned for a total of 6 months. This was followed up with lifelong clopidogrel (75 mg/day).

His baseline ECG showed normal sinus rhythm. A routine 24 h Holter monitor was performed 2 weeks after his stroke to look for possible underlying arrhythmias such as paroxysmal atrial

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Figure 1 Diffusion-weighted MRI (left panel) and apparent diffusion coefficient map (right panel) showing diffusion restriction in left lateral medulla.



fibrillation. The recording revealed multiple bradycardic and asystolic episodes, with a total of 56 significant asymptomatic episodes of sinus arrest longer than 3 s. The longest pause lasted 11 s (figure 4).

Transthoracic echocardiogram (ECHO) revealed a non-dilated left ventricle, with moderate wall thickness, normal systolic function and no evidence of intracardiac thrombus or masses.

DIFFERENTIAL DIAGNOSIS

The bradyarrhythmia and sinus arrests could have been due to inherent sinoatrial node disease, present prior to the stroke. Like atrial fibrillation, a more common arrhythmia, dysrhythmia could have predisposed the patient to stroke through cardiac thromboembolism.

However, the patient did not have any cardiac history, had a normal ECG at presentation and a normal ECHO. ECG recording over a 24 h period while an inpatient on the stroke ward had captured 56 episodes of sinus arrest. If frequent significant sinus arrests had been present for years or months prior to the stroke, we might have expected him to have had presented earlier with cardiac or syncopal symptoms.

The location of the patient's stroke in the lateral medulla raises the possibility that the ischaemic event could have affected key centres for cardiac autonomic regulation, resulting in potentially life-threatening sinus pauses.

TREATMENT

A permanent atrial pacemaker was inserted due to the risk of sudden cardiac death. While recovering from the procedure, the patient was found to have runs of 2:1 heart block on cardiac telemetry, with stimulated P waves from his newly inserted

pacemaker and narrow QRS complexes at a rate of 35–40 beats/min. Hence, he received a second procedure for insertion of a ventricular pacemaker lead.

OUTCOME AND FOLLOW-UP

While an inpatient, he demonstrated marked improvement in his speech and swallowing, but continued to suffer from significant ataxia. He was transferred to a rehabilitation unit for further physiotherapy after 4 weeks. Modified Rankin Scale (mRS) score was 3.

The patient was reviewed in the clinic, 8 months after his stroke. He reported he was doing well, but had developed mild left-sided facial pain, worse at night. He was independent in all activities of daily living and mRS score was one. He had nystagmus, and left-sided limbs showed mildly reduced co-ordination. He mobilised with the aid of one stick. ECG showed a paced rhythm.

DISCUSSION

A number of reports have described patients with LMS or medullary pathology who developed sinus arrests, or even fatal cardiorespiratory arrest. Von Heinemann *et al*³ describe a 45-year-old with right LMS secondary to right vertebral dissection who developed recurrent episodes of self-limiting asystole with loss of consciousness, each lasting approximately 10 s. Four episodes occurred within 48 h of admission, requiring insertion of a temporary transjugular pacemaker that was removed after 7 days. Kushner and Peters⁴ describe a patient with left cerebellar and brainstem infarct on CT scan who developed episodes of bradycardia, then a prolonged 27 s sinus arrest with a generalised seizure, which was terminated with atropine. He had a temporary pacemaker inserted, was intermittently pacemaker dependent for approximately 24 h and then normal sinus rhythm resumed. The authors postulate increased parasympathetic tone or decreased sympathetic tone following the stroke could have been responsible for the bradycardia and sinus arrests, supported by the fact that an anticholinergic agent was successful in terminating the arrest. Both patients described did not have a history of cardiac disease before their strokes.

Duberkar *et al* describe a 39-year-old with LMS and right vertebral artery thrombus, with extension into the proximal basilar artery, who suffered an unexpected and fatal cardiorespiratory arrest while receiving thrombolysis. The authors acknowledge the possibility that damage to the central

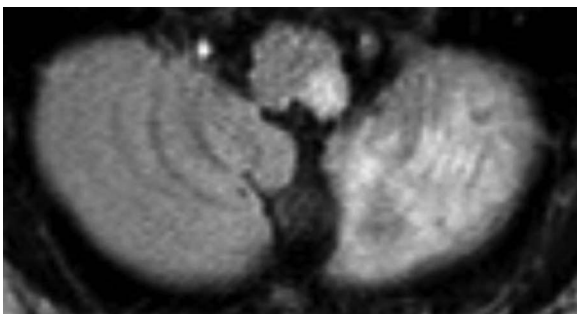


Figure 2 Fluid-attenuated inversion recovery MRI showing high signal in left lateral medulla and left cerebellar hemisphere.

Figure 3 Fat-suppressed MRI of the neck at two different levels showing high signal in left vertebral artery. There is low signal in the corresponding right vertebral artery, which appears patent.

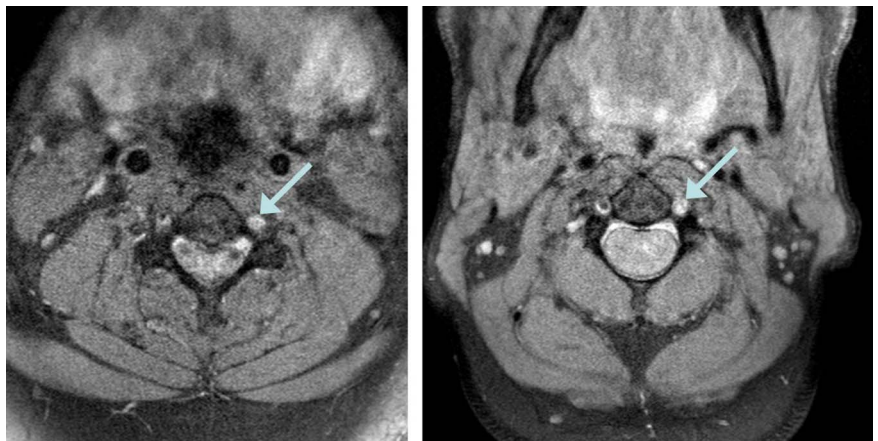
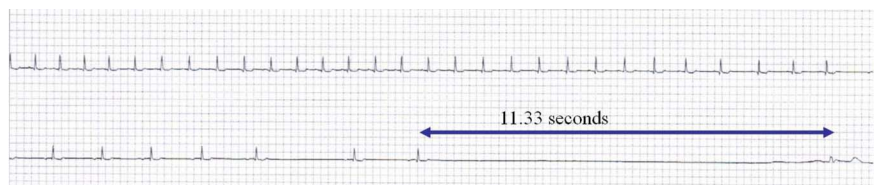


Figure 4 Excerpt from 24 h tape showing the longest episode of sinus arrest, lasting 11 s, preceded by a period of bradycardia.



cardiorespiratory centres following medullary infarction may have caused death.⁵

Other authors have described a 20-year-old with repeated episodes of sinus bradycardia and sinus arrest in a patient with a glioma in the medulla, which resolved quickly following targeted radiotherapy.⁶ Shibasaki *et al* describe a 61-year-old with multiple sclerosis (MS) who presented with intractable hiccups, nausea and syncope, with 52 sinus arrests on a 24 h tape. The patient was found to have a T2 hyperintense lesion in the medulla and diagnosed with a relapse of MS. The symptoms resolved after administration of high-dose intravenous methylprednisolone.⁷

A neuropathological study of five patients with acute heart failure who failed to resume independent cardiorespiratory function despite life support demonstrated bilateral changes in the medulla at the site of the NTS. The authors suggest abrupt decrease in cardiac output and cerebral blood flow may have caused irreversible ischaemic damage to the medullary nuclei responsible for cardiorespiratory regulation, resulting in failure to regain cardiorespiratory function.² Rodent studies have shown that stimulation of the P2X purinoceptors of the NTS produce rapid bradycardia and hypotension, through a combination of reduction in sympathetic outflow and an increase in parasympathetic outflow.⁸ Previous authors have suggested medullary infarction may disrupt central sympathetic inhibition of the NTS, resulting in reduced sympathetic outflow and increased vagal tone, thereby causing bradycardia and asystole.³ A small study comparing heart rate variability and plasma catecholamines, as a measure of autonomic activity, in 14 patients with either medullary or non-medullary brainstem stroke demonstrated acute dysfunction in the autonomic nervous system in those who suffered from medullary strokes.⁹

It appears that cardiac autonomic dysfunction is an uncommon but potentially fatal complication of medullary stroke and, indeed, other medullary pathology. We suggest extended cardiac monitoring should be considered for patients with medullary strokes.

Learning points

- ▶ Strokes do not always result in weakness. It is important to be aware of and recognise the lateral medullary syndrome with its constellation of clinical findings. A patient presenting with these symptoms could potentially be a candidate for thrombolysis.
- ▶ Cardiac dysrhythmia is an uncommon but potentially fatal complication of medullary pathology.
- ▶ Extended cardiac monitoring should be considered for patients with medullary strokes.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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