Lateral Medullary Syndrome Presenting with Ataxia and Bradycardia

T.E. Gofton, O.R. Segal, A.C. Skanes, J.A. Fraser, H.C. Hyson

Can. J. Neurol. Sci. 2009; 36: 390-392

The following outlines the case of a 60-year-old man presenting with ataxia and bradycardia as a consequence of a lateral medullary syndrome.

A 60-year-old right-handed man presented with a witnessed sudden loss of right arm coordination followed by brief loss of consciousness. Prior to symptom onset, while eating with his wife, the patient was suddenly unable to coordinate his fork, held in his right hand, between his plate and his mouth. He felt faint, his vision clouded and sounds became distant. The patient lost consciousness for a few seconds and fell onto the floor. Immediately upon awakening, he was alert and oriented. There was no tongue biting or urinary or fecal incontinence.

On initial assessment by paramedics the patient was dysarthric, had right-sided clumsiness and was profoundly bradycardic but normotensive (heart rate: 26 bpm, Figure 1). The dysarthria and clumsiness continued and diplopia (worse on right lateral gaze) and oscillopsia developed.

Past medical history included remote tonsillectomy and measles as a child. There was no history of smoking, alcohol consumption, diabetes mellitus, ischemic heart disease, hypertension or previous stroke. The patient was not taking any medications, alternative therapies, or illicit drugs. He did not have any known allergies.

On arrival in the emergency department, physical examination revealed a blood pressure of 140/74 mmHg, respiratory rate within normal limits and normal oxygen saturations. The heart rate was initially 26 beats per minute with a normal QRS complex and ST segment on electrocardiogram. Upon arrival in the emergency department, the electrocardiogram demonstrated a heart rate of 58 beats per minute with sinus rhythm. Recovery of heart rate and blood pressure occurred without medical intervention. Despite the bradyarrhythmia, heart sounds were normal with no added sounds or murmurs. Breath sounds were normal bilaterally and the abdomen was soft.

The patient was alert and oriented to person, place and time. Cranial nerve examination demonstrated full extraocular movements. Left beating horizontal nystagmus was present in primary position and in all directions of gaze. Right-sided partial ptosis was present, with a right pupil measuring 3 mm and a left pupil measuring 3.5 mm in bright light. In darker light, the right pupil measured 4 mm and the left pupil measured 5 mm. Both pupils were reactive to light. Funduscopy was not possible due to significant nystagmus. Left facial sensation was impaired. There was mild dysarthria. Motor examination was normal, there was no pronator drift and there were symmetrical reflexes with bilateral flexor plantar responses. Sensory examination revealed a left hemibody decrease in sharp touch and temperature sensation but normal joint position sense and vibration bilaterally. There was significant right-sided ataxia and dysdiadochokinesia and the patient had a tendency to fall to the right when sitting.

The clinical presentation was consistent with a right-sided lateral medullary infarction, confirmed on subsequent diffusion weighted magnetic resonance imaging (Figure 2). Magnetic resonance angiography demonstrated narrowing of the right vertebral artery likely secondary to atherosclerotic disease. There was no evidence of arterial dissection. Further investigations revealed: fasting glucose 8.4 mmol/L, hemoglobin A1c 0.054, 2h oral glucose tolerance test 10.4 mmol/L, fasting lipid profile with total cholesterol 4.3 mmol/L, triglycerides 0.7 mmol/L, high density lipoprotein (HDL) cholesterol 1.39 mmol/L, low density lipoprotein (LDL) cholesterol 2.6, cholesterol:HDL ratio 3.1. The patient was treated with aspirin 81 mg and atorvastatin 40 mg once daily. An echocardiogram revealed mild right ventricular dilation and hypokinesis.

The patient developed significant hiccups but no further arrhythmia on continuous cardiac telemetry over the following five days. The left-sided hemibody sensory deficit persisted while the right-sided ataxia improved steadily.

Lateral medullary syndrome or Wallenburg's syndrome is a well described syndrome resulting from infarction of the lateral aspect of the medulla posterior to the inferior olivary nucleus. Infarction of this area results in symptoms and signs secondary to both long tract and cranial nerve nuclei injury within the medulla. Clinically, a triad of ipsilateral ataxia, contralateral hypoalgesia and an ipsilateral Horner's syndrome is seen[1] among many other potential symptoms and signs. The ipsilateral ataxia is due to damage to olivocerebellar and spinocerebellar fibres as well as to the restiform body and inferior cerebellum. The loss of pain and temperature sensation over the contralateral trunk, arm and leg is explained by involvement of the spino-thalamic tract as it ascends through the brainstem. Ipsilateral Horner's syndrome occurs because of involvement of the descending

From the Department of Clinical Neurological Sciences (TEG, JAF, HCH), Department of Medicine, Division of Cardiology (ACS), London Health Sciences Centre, University of Western Ontario, London, Ontario, Canada; The Heart Hospital (ORS), University College London, London, UK.

Received March 5, 2008. Final Revisions Submitted January 16, 2009.

Correspondence to: Teneille E. Gofton, 7th Floor – University Hospital, Department of Clinical Neurological Sciences, London Health Sciences Centre, University of Western Ontario, 339 Windemere Road, London, Ontario, N6A 5A5, Canada.

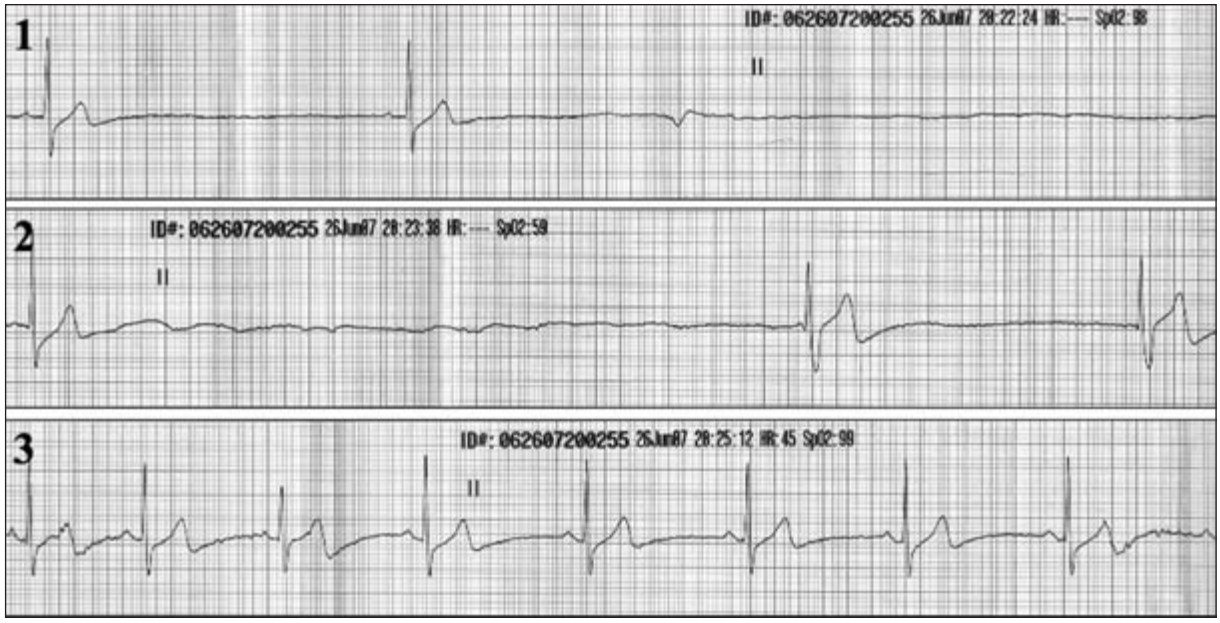

Figure 1: *Electrocardiogram strips demonstrating profound bradycardia (lead II only). Strip 1 shows sinus bradycardia with slowing of the sinus rate followed by asystole. The PR interval is within the normal range at 180ms. Strip 2 shows a P wave followed by a QRS complex with a shorter PR interval of 140ms. There is a long pause and then a P wave fused with a junctional escape beat. The 3rd QRS is another junctional escape beat. The 3rd strip shows sinus bradycardia with sinus arrhythmia and normal PR intervals throughout. AV nodal and interventricular conduction are normal in all 3 strips.*

sympathetic fibres in the lateral medulla.

The remarkable aspect of this case was the profound bradycardia at the onset of the stroke. From the history and the witness account, it is clear that there was definite right arm ataxia prior to the onset of syncope. Although autonomic dysregulation has been described in the context of lateral medullary infarction, it most often involves tachycardia or hypotension. While not captured by a formal twelve lead electrocardiogram, the lead II strips obtained by the paramedics demonstrate that the PR intervals, QRS duration and ST segments durations remained normal throughout the bradycardia (Figure 1). This demonstrates that intrinsic cardiac conduction was intact.

Sinus rate is controlled via sympathetic and parasympathetic input to the sinoatrial (SA) node. Slowing of the sinus rate can occur with a relative increase in parasympathetic tone over sympathetic tone, acting on muscarinic receptors of the SA node. In such a scenario, an escape rhythm may be seen from an alternate atrial site, the atrioventricular (AV) node, the His-Purkinje system or the ventricle. Sinus bradycardia may be due to sinus arrest (absence of impulse formation) or sinoatrial exit block (failure of exit of a sinus impulse)[2]. It is not possible to differentiate these causes from the surface electrocardiogram. In the present case, sinus bradycardia with a junctional escape rhythm was likely due to ischemic insult of the autonomic input, at the level of the lateral medulla, on the SA node resulting in a surge of parasympathetic tone.

Lateral medullary syndrome has been implicated as a neurological cause of hypotension, tachycardia or respiratory failure from involvement of the caudal and medial zone as well as the nucleus tractus solitarus[3]. There is more than one potential explanation for autonomic dysregulation occurring with a brainstem stroke including the sympathetic and parasympathetic systems. Experimental evidence in animal models suggests that neural influences from both the vestibular nuclei and the cerebellum on the reticular formation of the ventrolateral medulla may be important in the modulation of sympathetic outflow on the cardiovascular system[4,5]. Vestibular influences on the autonomic nervous system contribute to blood pressure and heart rate responses to orthostatic changes in cats[6,7]. This influence has been called the vestibulo-sympathetic reflex. Disconnection between the vestibular nuclei and to a lesser degree the cerebellum, as in the setting of a lateral medullary infarct, could impact the vestibulo-sympathetic reflex influence on the cardiovascular system resulting in decreased sympathetic tone. A transient duration of decreased sympathetic tone could result from unilateral lesioning of the vestibular nuclei by stroke. Homeostatic regulation of heart rate would potentially be restored once the intact contralateral vestibular nucleus compensates for its new situation.

Alternatively, an imbalance in parasympathetic tone could result in bradycardia. Within the parasympathetic system the nucleus of the solitary tract, the nucleus ambiguous and the dorsal motor nucleus of the vagus are implicated in cardiac rate control[8]. Vagal afferents from peripheral baroreceptors arrive in the brainstem from the periphery via the vagus nerve. Vagal efferents, providing heart rate inputs to the vagus nerve, partially

https://doi.org/10.1017/S0317167100007216 Published online by Cambridge University Press

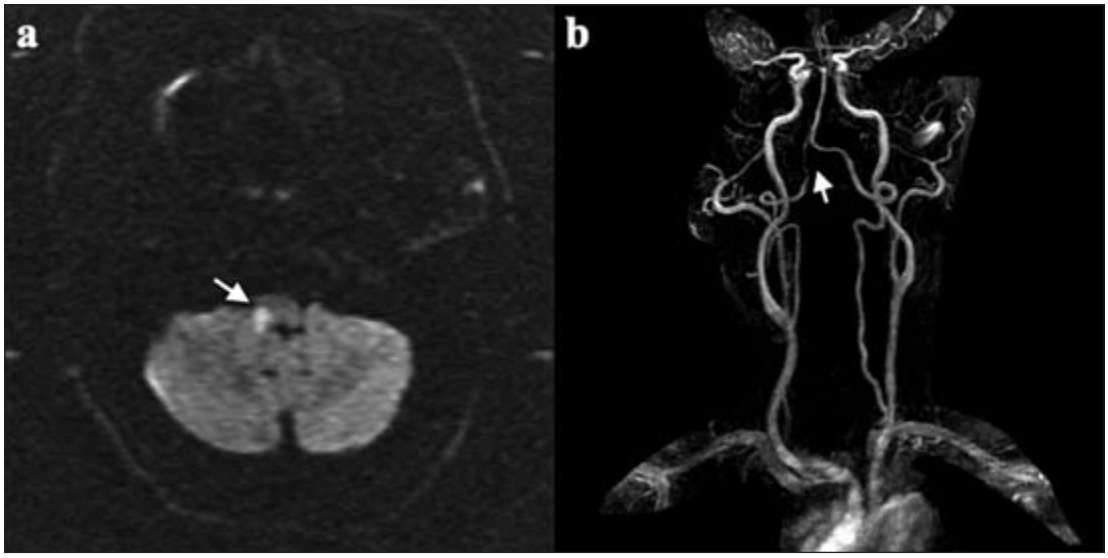

Figure 2: *MRI of the brain demonstrating a) diffusion restriction in the region of the right lateral medulla on diffusion weighted sequences and b) reduced caliber of the right vertebral artery (arrow) on MR angiography secondary to atherosclerotic disease.*

originate in the nucleus ambiguous[8]. Although rare, failure of the brainstem to receive baroreflex inputs or failure of adequate brainstem autonomic connections in the presence of intact parasympathetic efferents, as in the setting of brainstem stroke, could result in abnormal autonomic function[9]. Also, injury to the sympathetic efferent fibres with preservation of the parsympathetic efferent fibres could cause a surge in parasympathetic tone thereby causing bradycardia, hypotension and syncope[9]. Lesioning of the sympathetic system with an intact parasympathetic system in the setting of an acute stroke could also conceivably lead to transient bradycardia and hypotension by disrupting the balance of sympathetic and parasympathetic neural input on heart rate regulation.

In conclusion, this case was remarkable because of the striking symptomatic bradycardia that occurred in the context of a lateral medullary infarct, no doubt due to involvement of medullary autonomic control. Most likely, an acute and relative imbalance between sympathetic and parasympathetic control was the cause. In addition to the bradycardia, the clinical features in this patient were typical for a lateral medullary stroke syndrome, including the usual manifestation of sympathetic involvement (i.e. the Horner's Syndrome). Why bradycardia is not seen more frequently in the setting of a lateral medullary stroke is difficult to say. Whether the neural mechanisms responsible for bradycardia and hypotension in this case were due predominantly to loss of sympathetic tone or due to overactivity of the parasympathetic system cannot be determined. Nonetheless, it should be remembered that more serious symptoms can arise as a result of the lateral medullary stroke syndrome.

References

1. Currier RD GG, Dejong RN. Some comments on Wallenburg's lateral medullary syndrome. Neurology. 1961;11:778.
2. Waller BF, Gering LE, Branyas NA, Slack JD. Anatomy, histology, and pathology of the cardiac conduction system--Part V. Clin Cardiol. 1993;16(7):565-9.
3. Norrving B, Cronqvist S. Lateral medullary infarction: prognosis in an unselected series. Neurology. 1991;41(2 (Pt 1)):244-8.
4. Chen RV, Perlman J. Sudden cardiac arrest in an intubated premature infant with cerebellar and brainstem injury: is there a link? Pediatrics. 2006;117(5):1814-7.
5. Yates BJ. Vestibular influences on the autonomic nervous system. Ann N Y Acad Sci. 1996;781:458-73.
6. Fernandez C, Goldberg JM. Physiology of peripheral neurons innervating otolith organs of the squirrel monkey. III. Response dynamics. J Neurophysiol. 1976;39(5):996-1008.
7. Doba N, Reis DJ. Role of the cerebellum and the vestibular apparatus in regulation of orthostatic reflexes in the cat. Circ Res. 1974;40(4):9-18.
8. Oppenheimer S, Norris JW. Cardiac manifestations of acute neurological lesions. In: Aminoff MJ, editor. Neurology and general medicine. Philadelphia: Churchill Livingstone; 2001. p. 171-85.
9. Ketch T, Biaggioni I, Robertson R, Robertson D. Four faces of baroreflex failure: hypertensive crisis, volatile hypertension, orthostatic tachycardia, and malignant vagotonia. Circulation. 2002;105(21):2518-23.

https://doi.org/10.1017/S0317167100007216 Published online by Cambridge University Press