

Selected Topics: Neurological Emergencies

THE TROUBLE WITH SWALLOWING: DYSPHAGIA AS THE PRESENTING SYMPTOM IN LATERAL MEDULLARY SYNDROME

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Abstract—Background: Posterior circulation strokes account for approximately one-fourth of all ischemic strokes, but are frequently misdiagnosed by emergency providers. Current standard stroke screening tools such as the National Institutes of Health Stroke Scale and the Cincinnati Prehospital Stroke Scale are weighted toward anterior circulation stroke diagnosis. Lateral medullary syndrome, a type of posterior circulation stroke, can be particularly challenging to diagnose due to nonspecific presenting symptoms, such as dysphagia. **Case Report:** This report describes a 65-year-old man who presented with dysphagia, dizziness, and hoarseness. An initial neurological examination did not reveal any gross deficits, and imaging to evaluate for posterior circulation stroke was not obtained. The patient presented the following day with worsening symptoms, prompting imaging that revealed a large cerebellar ischemic infarction. **Why Should an Emergency Physician Be Aware of This?:** Although dysphagia frequently occurs in lateral medullary syndrome, it is rarely the presenting symptom in the emergency department. In patients with cerebrovascular risk factors who present with dysphagia, a complete neurological examination should be performed and noncontrast computed tomography (CT) of the head should be obtained if a neurological deficit is appreciated. Due to their poor sensitivity, CT scans can frequently miss posterior circulation strokes, therefore magnetic resonance imaging should be considered if provider suspicion remains high. Emergency providers are encouraged to have a high level of suspicion for this rare but debilitating stroke syndrome

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INTRODUCTION

Appropriate and timely recognition of posterior circulation strokes by emergency medicine providers is critical. Posterior circulation strokes account for 20–25% of all ischemic strokes (1). They are more than twice as likely to be misdiagnosed as anterior circulation strokes, with a misdiagnosis rate of up to 37% (2,3). Posterior circulation stroke symptoms are frequently misdiagnosed as dehydration, vestibular neuritis, benign positional vertigo, gastroenteritis, or various metabolic disorders (4). Standard stroke screening tools such as the National Institutes of Health Stroke Scale and Cincinnati Prehospital Stroke Scale are weighted toward symptoms commonly found in anterior circulation strokes (5,6).

Wallenberg syndrome, caused by infarction of the lateral medulla, is a posterior circulation stroke syndrome that typically arises due to occlusion of the vertebral artery or posterior inferior cerebellar artery (7). This syndrome can be difficult to differentiate among posterior circulation strokes. Key vestibulocerebellar features include dysphagia, hoarseness, gait abnormalities, dysmetria, dysarthria, dizziness, lethargy, headache, nausea,

and vomiting (2,8). Classic sensory signs and symptoms include loss of pain and temperature sensation to the ipsilateral face and contralateral trunk and limbs due to involvement of the spinothalamic tract (9). This report provides recommendations for emergency medicine providers as they approach high-risk patients presenting with nonspecific symptoms, specifically dysphagia, dizziness, and hoarseness, which are frequently associated with posterior circulation stroke syndromes such as lateral medullary syndrome.

CASE REPORT

ED Presentation #1

A 65-year-old white man presented to the Emergency Department (ED) with 2 days of “throat congestion and fullness,” cough, and dizziness. He reported similar episodes nearly every 6 months for the last 10 years, which typically lasted < 24 h and resolved with over-the-counter decongestants. He reported difficulty swallowing and voice changes. His dizziness was described as lightheadedness with standing, which he attributed to decreased oral intake secondary to dysphagia. He denied vertiginous symptoms. A comprehensive review of symptoms was otherwise noncontributory. He denied exposure to any reported allergens. Past medical history was significant for allergic rhinitis, migraines, and chronic back pain. He was a nonsmoker.

In the ED, vital signs were only significant for hypertension with a systolic blood pressure in the 160s (mm Hg), heart rate 70 beats/min, and oxygen saturation in the mid 90s. On examination he was alert and oriented, and in no acute distress. He had no asymmetry of the external face or neck, oral or labial swelling, tongue edema, or oral erythema. There was no appreciated asymmetry or erythema of the posterior oropharynx, and his uvula was midline. There was no palatal or submental fullness. The patient had nasal voice but no stridor or notable upper airway noises, and his respirations were unlabored. No gross cranial nerve (CN) deficits, dysarthria, or aphasia were appreciated on examination. There was no tracheal deviation, neck tenderness, or thyromegaly. He had no gross sensory or motor deficits, and his gait was grossly intact by observing him ambulate unassisted to the restroom in the ED. The patient received 1 L of intravenous (i.v.) lactated Ringer solution, i.v. diphenhydramine, i.v. solumedrol, and i.v. famotidine, with improvement in symptoms, and was able to tolerate oral intake. Renal panel demonstrated a glucose of 161 mg/dL, but was otherwise unremarkable. The resident and attending physician discussed performing a nasopharyngoscopy (NP scope) to rule out any significant pharyngeal edema, obstruction, or abnormality deep to

the soft palate. NP scope performed by the resident physician with attending supervision at bedside revealed diffuse mild edema of the posterior oropharynx with a thick white plaque. The uvula was midline, and epiglottis and vocal cords were normal in appearance. The patient was diagnosed with esophageal candidiasis, prescribed nystatin suspension, and instructed to follow up with an ear, nose, and throat specialist and his primary care physician.

ED Presentation #2 and Hospital Course

The patient returned to the ED the following day for persistent symptoms, with new-onset inability to tolerate oral intake and the prescribed nystatin due to dysphagia and odynophagia, which resulted in gagging upon attempted swallowing. He also endorsed continued dizziness with new-onset difficulty standing and assistance with ambulation. Vital signs on ED arrival showed he was hypertensive, afebrile, and without tachycardia or tachypnea. His examination was remarkable for bilateral submandibular gland swelling and cervical adenopathy, but no tonsillar erythema or exudate. Cardiac, pulmonary, and abdominal examinations were unremarkable. A brief neurological examination showed no gross motor or sensory deficits in his upper or lower extremities; however, CN and cerebellar functions were not initially tested. The patient received i.v. fluids, diphenhydramine, famotidine, guaifenesin, and sublingual atropine drops without improvement of symptoms. Laboratory work demonstrated a mild leukocytosis with a white blood cell count of 11.4 K/uL, but was otherwise unremarkable. At this time, the management plan was focused on identifying pharyngeal or laryngeal soft tissue edema, infection, or mass. A computed tomography angiography scan (CTA) of the neck was obtained to identify structural abnormalities, but incidentally revealed a large left cerebellar infarct with left vertebral artery occlusion at the level of C1–C2. Tissue plasminogen activator (tPA) was contraindicated given the time since onset of symptoms and established infarct present on CT. A comprehensive neurologic examination performed in the ED revealed left upper and lower extremity ataxia, left miosis, nystagmus on leftward gaze, left-sided facial weakness, and dysarthria. The patient was admitted to the intensive care unit for cerebellar infarct with lateral medullary syndrome. An urgent magnetic resonance imaging (MRI) scan was performed, which showed large-volume infarct in the left cerebellar hemisphere with extension into the posterolateral medulla (Figure 1). He was intubated for hypoxic respiratory failure and aspiration pneumonia. He received a tracheostomy due to prolonged intubation and failed extubation. A percutaneous endoscopic gastrostomy tube was placed for poor feeding due to significant

dysphagia. He was discharged to inpatient rehabilitation and has since been discharged home. To date, the patient does not eat by mouth due to persistent dysphagia and aspiration risk; his tracheostomy tube has been decannulated and capped, and his modified Rankin Score is 4.

DISCUSSION

Posterior circulation strokes frequently present with nonspecific symptoms, which contributes to their high misdiagnosis rate (10). Our patient was a 65-year-old man with acute lateral medullary infarction, or Wallenberg syndrome, who initially presented with a primary complaint of dysphagia, with associated hoarseness and nonvertiginous dizziness. Dysphagia is common in lateral medullary syndrome due to impaired swallowing function, and may be present in up to 94% of lateral medullary strokes (8,11). However, dysphagia is a presenting symptom in < 15% of all posterior strokes and is frequently transient or mild (11,12). Although several prior case reports have discussed cases of lateral medullary syndrome presenting with dysphagia, this report is unique because the authors outline specific diagnostic considerations for the emergency physician to mitigate future misdiagnosis (13–15).

Brainstem strokes, such as the one our patient suffered, will typically present with numerous crossed symptoms due to the density of tracts within the brainstem that are affected by relatively small infarcts. However, posterior strokes can present with one to two isolated complaints and deficits (16). Dysphagia is typically affected in lateral medullary syndrome because swallowing is controlled by several anatomical regions in the medulla (17,18). These include the dorsal region, which contains the nucleus solitarius, and the middle and ventral

regions, which include the reticular formation and the nucleus ambiguus (17,18). In patients with cardiovascular risk factors presenting with dysphagia, it is crucial to maintain a high level of suspicion for cerebrovascular causes.

Patients with dysphagia should undergo a detailed neurological examination, including cranial nerve and cerebellar testing, to identify a potential posterior circulation stroke. When evaluating a patient with dysphagia, structural causes (e.g., masses, infection, lymphadenopathy, food boluses) must first be excluded. After these have been ruled out, the following physical examination maneuvers are recommended to evaluate for neurological causes: bedside swallow test; examination of the uvula, specifically looking for contralateral deviation (as CN 9 and 10 are affected in lateral medullary syndrome); hyposthesia of the face from CN 7 (although this is higher in the brainstem), loss of gag reflex, or tongue deviation to suggest CN 12 involvement. With regards to the patient's dizziness, a more detailed gait test (looking for specifically wide-based gait) as well as Romberg test could have differentiated a more concerning neurological cause of dizziness from a more benign orthostatic cause secondary to dehydration. The HINTS examination (Head-Impulse, Nystagmus, Test-of-Skew) has gained significant traction in both the emergency medicine and neurology literature as a tool to aide in the diagnosis of posterior strokes (19). The presence of normal horizontal head impulse test, direction-changing nystagmus in eccentric gaze, or skew deviation have been proven to be highly sensitive and specific for vertebrobasilar strokes (19). Failure to perform the above testing during the initial ED visits could have contributed to misdiagnosis, had the CTA of the neck not been ordered for other diagnostic purposes. If both dysphagia and a neurological deficit are

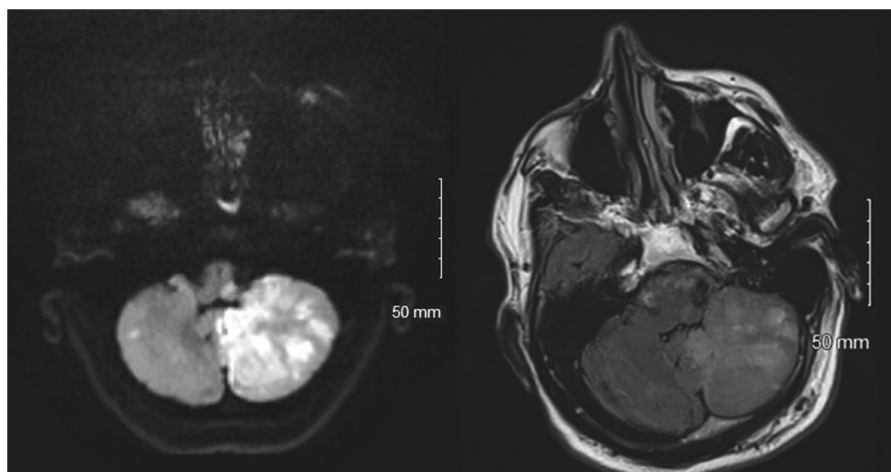


Figure 1. Diffusion-weighted imaging (left) and fluid-attenuated inversion recovery (right) magnetic resonance imaging of the brain during hospitalization illustrating large-volume infarct in the left cerebellar hemisphere with extension into the posterolateral medulla.

appreciated on examination, standard noncontrast CT imaging of the head should be obtained. Patients presenting with acute symptoms should be considered for thrombolysis, if eligible. However, CT scans frequently miss acute posterior strokes, and MRIs offer superior sensitivity when compared with CT scans (3,8,20).

Diffusion-weighted imaging (DWI) can also aid in early demonstration of infarction, if suspected (3). Despite the superiority of MRI DWI, there is still a considerable false-negative rate (anywhere from 6–12% quoted in the literature) of acute-onset posterior circulation strokes missed on MRI (3,19,21). Selected patients who present > 4.5 hours from when last seen well may be candidates for thrombolysis on the basis of DWI-fluid-attenuated inversion recovery mismatch on acute MRI, although this is not the standard of care for tPA and the decision should be made on a case-by-case basis in conjunction with Neurology (22). We recommend that patients with cerebrovascular risk factors who present with dysphagia and objective cranial nerve impairment receive urgent MRI despite a negative CT of the head if the level of suspicion is high. Additionally, emergency medicine providers can advocate for admission for repeated imaging and frequent neurological examinations despite initial negative imaging, if symptoms persist.

Posterior circulation strokes are diagnostically challenging to emergency medicine providers. They frequently present with nonspecific symptoms common in benign diagnoses. A detailed neurologic examination should accompany any patient with cardiovascular risk factors presenting with dysphagia to identify subtle neurologic findings. If standard stroke imaging is negative and provider concern remains high, evaluation of these patients may be further enhanced by early utilization of MRI. Overall, early diagnosis of posterior stroke requires a high level of suspicion among emergency physicians.

WHY SHOULD AN EMERGENCY PHYSICIAN BE AWARE OF THIS?

Although dysphagia frequently occurs in lateral medullary syndrome, it is rarely the presenting symptom in the ED. In patients with cerebrovascular risk factors who present with dysphagia, a complete neurological examination should be performed and noncontrast CT head should be obtained if a neurological deficit is appreciated. Due to their poor sensitivity, CT scans can frequently miss posterior circulation strokes, therefore MRI should be considered if provider suspicion remains high. Emergency providers are encouraged to have a high level of suspicion for this rare but debilitating stroke syndrome to avoid misdiagnosis and delayed care.

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