Sudden Bilateral Deafness

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About the Authors

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Basilar artery occlusion is associated with a high mortality rate and poor functional outcome in survivors. The most common prodromal symptoms are motor and oculomotor deficits. Hearing loss is not a major prodromal symptom.

Sudden deafness usually results from either circulatory disturbances or inflammation. Deafness of vascular etiology generally occurs unilaterally. Occlusion of basilar arterial origin suggests that thrombosis is the primary mechanism of stroke, especially in elderly patients. This case highlights the importance of hearing loss as either a main manifestation or a warning of impending brainstem ischemia. Clinicians should be aware of the possibility of vertebrobasilar ischemia in patients with bilateral sudden deafness, even when classic brainstem or cerebellar signs are mild or absent. This case also highlights the importance of prodromal signs and symptoms.

Case Report

A 65-year-old right-handed woman with long-standing hypertension presented to the emergency room with sudden bilateral deafness and difficulty communicating. Collateral history was obtained from her family, as the patient was unable to communicate because of her bilateral deafness. Twelve to 24 hours previously, the patient had had an acute onset of intense vertigo-associated vomiting and had become bedridden because of ongoing vertigo and dizziness, with rapid deterioration of her clinical status over the previous two hours.

She now had slurred speech and was unable to ambulate independently. The patient's family denied her having any convulsions, incontinence, or loss of consciousness. The patient's only medication was hydrochlorothiazide (25 mg daily). She had no cerebrovascular risk factors except hypertension. She was a nonsmoker and a nonalcohol user. Her family history was noncontributory. She had no known drug allergies.

On arrival at hospital, her Glasgow Coma Scale score was 9 (E2, M5, V2). She was afebrile and in sinus tachycardia with a heart rate of 115 beats per minute (BPM), her blood pressure was 177/85, and her oxygen saturation was 99% on 5 litres of oxygen. Her pupils were equal and reactive to light. Funduscopy revealed hypertensive eye changes but no papilledema. Cranial nerve VIII was compromised; the remaining cranial nerves could not be fully assessed as the patient was unable to follow commands. Her neurologic examination was limited due to her hearing impairment. The right upper and lower extremities were flaccid relative to the left, and plantar reflexes were bilaterally extensor. Cerebellar testing could not be performed, as the patient was not fully able to cooperate.

Her respiratory examination was unremarkable. There was no evidence of volume overload. Her jugular venous pulse was flat, and heart sounds were normal. She had a 2/6 holosystolic murmur at the apex that radiated to the axilla. Her abdomen was soft, and no obvious guarding was noted. Her level of consciousness was fluctuating, and she was intubated and mechanically ventilated for airway protection.

On admission her hemoglobin level was 87 g/L, with a mean corpuscular volume of 90 fL, white blood cell counts were 19,900

g/L with 92% neutrophils, 4% lymphocytes, 2.5% monocytes, and 0% eosinophils. Her blood film showed no evidence of hemolysis. Platelets were 150,000 g/L. Her serum sodium level was 136 mEq/L, serum potassium level was 3.2 mmol/L, and serum chloride level was 111 mEq/L. Serum creatinine was 99 µmol/L, urea was 33 mmol/L, and a random glucose level was 95 mg/dL. Her serum transaminases, amylase, and lipase were all within normal limits, but cholesterol values were extremely elevated, with low-density lipoprotein at 4.2 mmol/L and high-density lipoprotein at less than 1.0 mmol/L, total cholesterol/HDL greater than 5, and triglycerides at 2 mmol/L. Cardiac work-up and autoimmune and vasculitic screens were negative except for an elevated C-reactive protein and erythrocyte sedimentation rate. Her transthoracic echocardiogram revealed a normal ejection fraction and mild-to-moderate mitral regurgitation. Her electrocardiogram showed sinus tachycardia at 140 BPM, with evidence of left ventricular hypertrophy but no ST changes and a normal QT interval of 420 ms. Noncontrast brain computed tomography showed high attenuation of the basilar artery, consistent with a thrombus and bilateral cerebellar peduncle infarction, and T2-weighted magnetic resonance imaging of the brain confirmed hyperintensities involving both inferior cerebellar peduncles and pontomedullary junction. Magnetic resonance angiography of the brain revealed complete occlusion of the basilar artery at its origin and severe stenosis of the distal vertebral artery bilaterally.

Due to the delay in presentation, the patient was not eligible to receive intravenous thrombolysis. In fact, in order to be effective, patients should be treated with intravenous tissue plasminogen within 4.5 hours of the onset of stroke symptoms. Instead she was started on an antiplatelet agent, clopidogrel 75 mg daily and simvastatin.

The patient was admitted to the intensive care unit for further monitoring and care. During her stay, she remained dysarthric and unable to obey commands consistently. She had recovered very little of her motor function; her balance had mildly improved and she continued to be wheelchair dependent. Otoscopic examination confirmed sensorineural hearing loss (SHL). Urgent audiometric testing and brainstem auditory evoked potentials confirmed almost complete hearing loss bilaterally. Eventually the patient was extubated, and she was supplied with hearing aids and referred to a rehabilitation centre, where she participated in a graded exercise rehabilitation program. A follow-up pure tone audiogram confirmed 80% bilateral deafness, while her motor deficits (including

coordination and gait) improved steadily with rehabilitation over several weeks. At a three-month follow-up post-discharge, bilateral deafness persisted.

Discussion

Idiopathic sudden SHL has an incidence of 5 to 30 cases per 100,000 cases per year.¹ The hearing loss is unilateral in most cases; bilateral involvement is reported in less than 5% of cases.² Tinnitus occurs in about 80% of patients, and vertigo indicating an associated peripheral vestibular dysfunction occurs in about 30%.³ Up to 80% of patients report a feeling of ear “fullness.”^{3,4} In 2.8% of patients, sudden SHL is due to vascular or hematologic causes, particularly ischemia involving the cochlear or the ascending pathways. The cochlea is supplied by an end artery, and vascular occlusion has been postulated to be a cause for sudden SHL.⁵ In our case there was evidence of basilar artery infarction, suggesting that thrombosis was likely the cause. This is in keeping with previous studies that have shown that local atherothrombosis is the major etiology of vertebrobasilar occlusions in the elderly population.^{4,6}

Despite limited large-scale studies, the literature has shown that sudden SHL is associated with a significant increase in the hazard of stroke during the subsequent five years.^{7,8} From this we can conclude that patients with sudden hearing loss should be examined for additional brainstem symptoms, since this can be the presenting sign of a life-threatening basilar artery thrombosis.⁹

Conclusion

Vertebrobasilar or posterior circulation territory stroke accounts for 20% of all strokes.¹⁰ Basilar artery occlusion is generally associated with a high mortality rate and poor functional outcomes in survivors (i.e., major disabilities, including tetraplegia and coma).^{9,10} Sudden SHL can present as a warning signal of an acute basilar artery stroke. Therefore, an early diagnosis is essential if direct or supportive measures are to be of benefit.^{9,10}

This case highlights the importance of hearing loss as either a main manifestation or a warning of impending brainstem ischemia. Clinicians should be aware of the possibility of vertebrobasilar ischemia in patients with bilateral sudden deafness, particularly in older patients, even when classic brainstem or cerebellar signs are mild or absent.

References

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