Teaching NeuroImage: Sudden Bilateral Sensorineural Hearing Loss Due to Vertebrobasilar Ischemia

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A 56-year-old woman with history of uncontrolled hypertension, hyperlipidemia, and tobacco use developed sudden bilateral hearing loss preceded by one week of persistent vertigo. Physical examination demonstrated saccadic pursuit with spontaneous bilateral horizontal and vertical down-beating nystagmus, profound bilateral sensorineural hearing loss (bSNHL) confirmed by audiogram, and truncal ataxia. Neuroimaging showed occlusion of the right vertebral artery, basilar artery, and near-occlusion of bilateral anterior inferior cerebellar arteries (AICAs) (Figure 1) causing acute/subacute ischemic strokes (Figure 2). AICAs give rise to internal auditory artery, almost a sole blood supply to inner ear, and supply anterior part of cerebellum, middle cerebellar peduncle, and flocculus. Sudden bSNHL is a rare phenomenon representing < 5% of all acute SNHL cases dominated by unilateral presentation. In contrast to the latter, sudden bSNHL represents a medical emergency warranting immediate evaluation for life-threatening and/or reversible causes such as stroke, autoimmune disorders (e.g. Susac, Cogan syndromes, scleroderma), drug toxicities (e.g. gentamicin), etc. Occlusion/hypoperfusion of AICA in isolation or as part of vertebrobasilar ischemia can lead to sudden SNHL. AICAs give rise to internal auditory artery, almost a sole blood supply to inner ear, and supply anterior part of cerebellum, middle cerebellar peduncle, and flocculus. Sudden bSNHL is a rare phenomenon representing < 5% of all acute SNHL cases dominated by unilateral presentation. In contrast to the latter, sudden bSNHL represents a medical emergency warranting immediate evaluation for life-threatening and/or reversible causes such as stroke, autoimmune disorders (e.g. Susac, Cogan syndromes, scleroderma), drug toxicities (e.g. gentamicin), etc. Occlusion/hypoperfusion of AICA in isolation or as part of vertebrobasilar ischemia can lead to sudden SNHL.
Figure 1. CT angiography of head and neck (maximum intensity projections, [MIP]) with three-dimensional reconstruction. The following structures are indicated: occlusion of right V4 vertebral artery (black arrowhead), patent left V4 vertebral artery (white arrowhead), occlusion of basilar artery (white arrow), near-occlusion of bilateral AICAs (black arrows).

Figure 2. MRI brain without contrast showing subacute/acute areas of infarction. From right to left hyperintense signal on FLAIR, hyperintense areas of restricted diffusion on DWI with correlating hypointense areas on apparent diffusion coefficient maps in bilateral anterior cerebellum (panel A), middle cerebellar peduncle, flocculus and pons (panels B&C).
References


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