

CASE REPORT

A cardiac cause for deafness

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SUMMARY

We describe a case of a 49-year-old diabetic man with a history of myocardial infarction, presenting with deafness for 2 weeks. Initial assessment by otorhinolaryngologists was suggestive of sensorineural hearing loss. Subsequently, the cardiac evaluation showed an apical clot in the left ventricle. Careful neurological reassessment and a cranial MRI yielded a diagnosis of cortical deafness with auditory agnosia secondary to bilateral temporal infarcts. Doppler and MRI ruled out carotid artery thrombosis. The temporal infarcts were therefore considered to be cardioembolic in origin. Cardioembolic stroke is an important differential diagnosis for patients presenting with neurological symptoms on a background of cardiac disease.

BACKGROUND

Cardioembolic strokes account for up to 20% of all ischemic strokes, but often produce disproportionately greater disability than non-embolic strokes due to their potential for involving larger intracranial arteries.¹ Characteristic features of cardioembolic strokes include infarcts of multiple discrete vascular territories, and alternating right and left hemispherical involvement.² Recognition of the cardioembolic stroke is essential not only because of the morbidity associated with them, but also because of the significant reduction in the risk of stroke with appropriate antithrombotic therapy.¹ The patient described in this report developed bilateral temporal lobe infarcts after defaulting on standard antiatherosclerotic medications including low-dose aspirin. Although he attained only limited recovery of hearing loss, rapid institution of antiplatelet and oral anticoagulation therapy prevented further embolic events.

Isolated deafness is an uncommon manifestation of cardioembolic stroke; a meticulous neurological examination as well as a high index of suspicion is essential for diagnosis, especially in persons with a history of cardiac disease.

CASE PRESENTATION

A 49-year-old Asian-Indian diabetic man presented with the sudden onset of deafness in both ears for 2 weeks. There were no associated aural symptoms such as tinnitus, giddiness or ear pain or discharge and no other neurological symptoms. He had suffered a myocardial infarction 13 years before, but had since defaulted on antiatherosclerotic medications. He claimed to be obedient on antidiabetic drugs. He denied any history of smoking, alcohol consumption or other forms of substance misuse.

General physical examination was unremarkable. He was haemodynamically stable. Neurological

examination showed difficulty in comprehending spoken words, and distinguishing musical notes from environmental sounds. His speech was intact and comprehension of written words was unimpaired. There were no other focal neurological deficits. The cardiovascular examination was essentially normal.

INVESTIGATIONS

Routine laboratory tests revealed well-controlled glycaemia (fasting blood glucose 135 mg/dL; glycosylated haemoglobin 6%). Complete blood counts, erythrocyte sedimentation rate and liver and renal function tests were within normal limits. Electrocardiography showed pathological q waves in V₁₋₃ with loss of R waves, and T wave inversion, consistent with an old anterior wall myocardial infarction. The patient was in sinus rhythm. Transthoracic echocardiography demonstrated akinesia of the apical segment of the left ventricle with hypokinesia of the interventricular septum and anterior wall. A left ventricular apical clot was also visualised. Mild left ventricular systolic dysfunction was noted (ejection fraction 47%).

A cranial MRI showed a chronic infarct with encephalomalacia and gliosis involving the right temporal lobe, and a subacute infarct involving the left temporal lobe with mild effacement of adjacent sulci (figure 1). Intracranial flow voids were preserved. Carotid artery Doppler studies showed normal flow velocities and spectral wave forms; intimal plaques were not seen.

Audiometry revealed mild bilateral sensorineural hearing loss inconsistent with the degree of hearing impairment reported by the patient.

DIFFERENTIAL DIAGNOSIS

- ▶ Partial cortical deafness with auditory agnosia secondary to cardioembolic infarction of bilateral temporal lobes.
- ▶ Sensorineural hearing loss.

TREATMENT

Oral anticoagulation with warfarin was initiated for cardioembolic stroke, with a target international normalised ratio (INR) of 2.5. In addition, the patient was restarted on antiplatelet therapy, statins, ACE inhibitors and β -blockers for coronary artery disease. Speech therapy was administered for his hearing impairment.

OUTCOME AND FOLLOW-UP

Minimal subjective improvement in hearing loss was reported by the patient, following speech and pharmacotherapy. He was counselled regarding the nature of his illness and the need for compliance to



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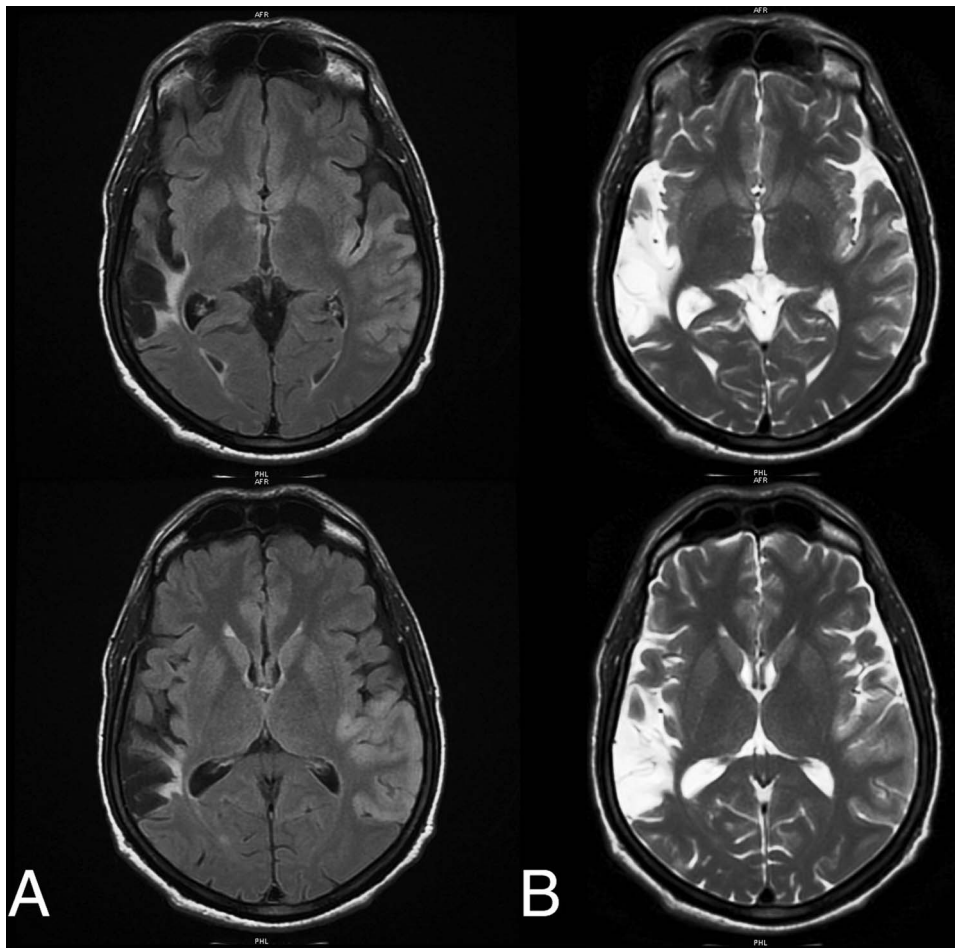


Figure 1 Cranial MRI showing a chronic infarct with encephalomalacia and gliosis involving the right temporal lobe, and a subacute infarct involving the left temporal lobe with mild effacement of adjacent sulci ((A) T1 sequence, axial section; (B) T2 sequence, axial section).

therapy, and was subsequently discharged. He continues to remain on follow-up at our hospital, with no further episodes of neurological deterioration.

DISCUSSION

Syndromes of central auditory disorders include cortical deafness, pure word deafness, auditory agnosia for environmental sounds and amusia.³ Of these syndromes, cortical deafness constitutes the most severe form, presenting with complete deafness and preserved startle reflexes to loud sounds. Conversely, auditory agnosias are characterised by impaired perception of certain classes of sounds. Examples of auditory agnosia thus include pure word deafness, wherein the patient cannot distinguish speech sounds but can lip-read and can comprehend written words, and agnosia for non-verbal environmental sounds, with preserved comprehension of spoken speech. Other specific forms of auditory agnosia include phonagnosia referring to an inability to recognise individuals from the sound of their voice⁴ and amusia indicating an impaired perception of music.⁵ The latter form of auditory agnosia was seen in our patient, who had difficulty in distinguishing musical notes from environmental sounds. Pertinently, amusia is frequently associated with other forms of auditory agnosia.⁵ Such was the case in this instance, as our patient combined word deafness with amusia.

Auditory agnosia typically results from damage to bilateral temporal lobes⁶ or specifically to the supratemporal gyri containing the primary auditory cortex, although there are isolated reports of auditory agnosia in patients with unilateral temporal

lobe disease and even subcortical lesions.⁷ Some reports have also demonstrated primarily apperceptive agnosia in patients with right hemispherical lesions as opposed to associative agnosia in those with left hemispherical lesions.⁸ Ischaemic insult resulting from stroke is a major cause of temporal lobe injury; other aetiologies include head trauma, congenital malformations, tumours, encephalitis and neurodegenerative disorders.⁹

By their very nature, auditory agnosias respond poorly to treatment. Therapeutic options including speech therapy and training in lip-reading can produce limited improvement in communication skills. Nevertheless, as this case report demonstrates, such agnosias can, albeit rarely, indicate an underlying source of emboli with definite therapeutic options.

Learning points

- ▶ Embolic stroke should be considered in patients with infarction of discrete bilateral cerebral arterial territories, especially on a background of cardiac disease.
- ▶ Bilateral temporal lobe infarction can present with cortical deafness, although unilateral temporal lobe involvement may be clinically silent.
- ▶ Careful neurological evaluation for auditory agnosia can provide critical clues for diagnosing cortical deafness.

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Competing interests None.

Patient consent Obtained.

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