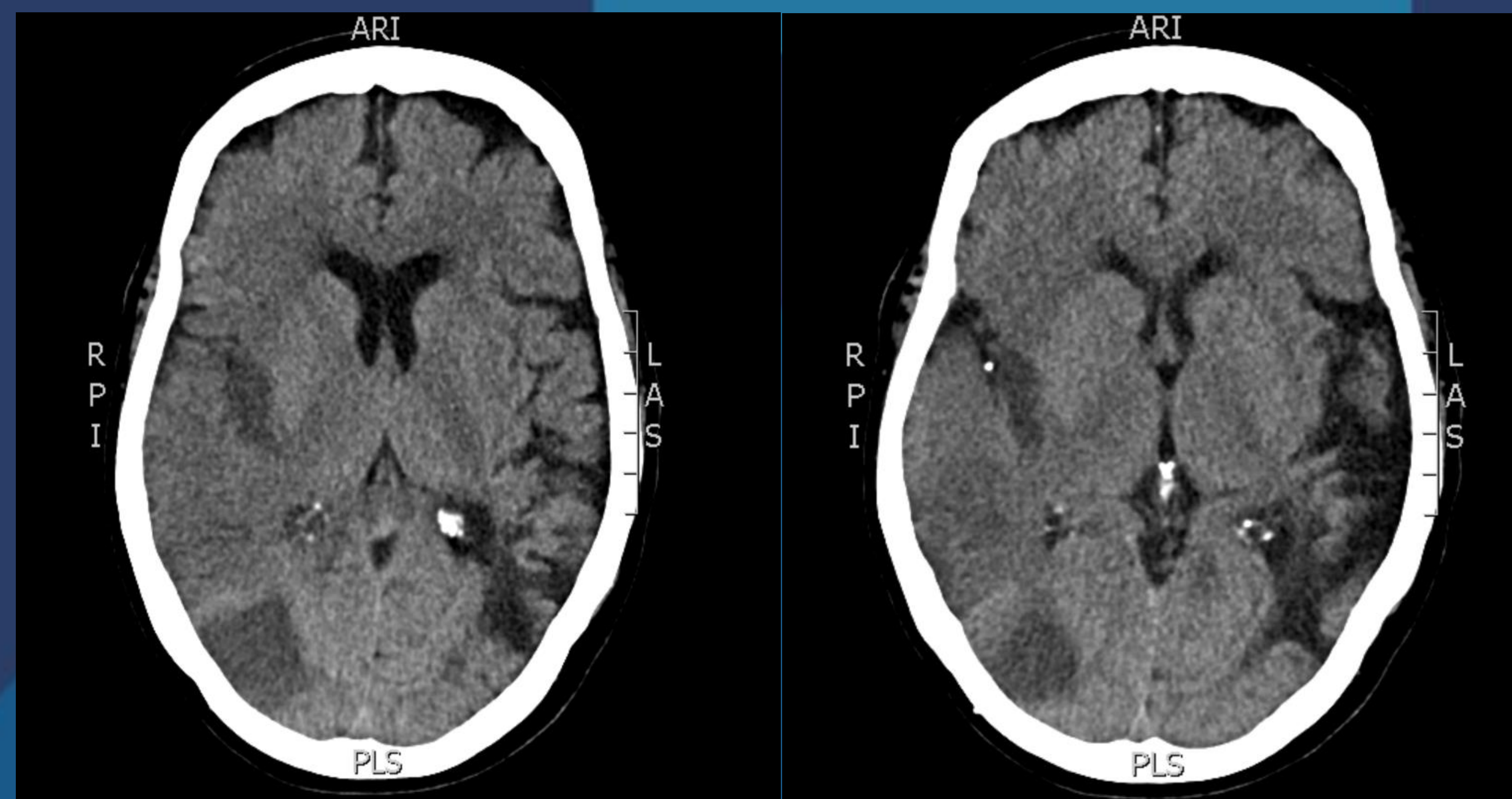


INTRODUCTION

Cortical deafness is a rare diagnosis and occurs secondary to damage to the bilateral auditory cortices. Recognizing and properly diagnosing this condition is challenging as it can be difficult to distinguish from many peripheral auditory disorders. With stroke being the most common cause, it is important to implement an interdisciplinary approach to rehabilitation.



DISCUSSION

Deafness of central auditory deficits can be divided into three variants: cortical deafness, auditory agnosia, or verbal auditory agnosia. Cortical deafness is a rare and more severe form of deafness where a patient is unresponsive to all types of sound. Interestingly, there are only twelve reported cases in the literature, which explains the dearth of medical knowledge and research. Typically, as seen in this case, cortical deafness is secondary to bilateral lesions of the Central Auditory Nervous System, specifically the primary auditory cortex region in the transverse temporal gyri of Heschl. Patients are unable to hear sounds, yet have no apparent damage to the inner ear. However, they may exhibit certain reflex responses, such as turning their head towards a loud sound.

RESULTS

After audiology evaluation, it was recommended to condition the patient for more reliable responses to sound. For example, speech therapy introduced response to a raised hand when hearing a sound. Eventually, the patient began responding to auditory startle and loud voices, and demonstrated brief attention towards structured questions with moderate cues. If needed, the patient was able to read questions presented on a whiteboard and respond accordingly. Recovery was gradual with simple and basic auditory function recovering first.

CASE DESCRIPTION

A 70-year-old female, with a past medical history of a left chronic MCA stroke, developed a right MCA infarct. On admission to rehabilitation, initial evaluation revealed an absent response to presented auditory stimuli. However, the patient could follow single unit directions with 100% accuracy in written form. Prior to admission, the patient had no hearing deficits. Following the most recent stroke, she developed a complete absence of hearing, particularly environmental or speech stimuli. Audiology evaluation revealed a deficit in central hearing, specifically retrocochlear, with intact peripheral hearing. Neuroimaging revealed infarcts in bilateral Heschl gyri. Ultimately, she was diagnosed with cortical deafness.

Frequency	Right Ear	Left Ear
1500 Hz	Present	Present
1800 Hz	Present	Present
2000 Hz	Present	Present
2500 Hz	Present	Present
3000 Hz	Present	Present
3500 Hz	Present	Present
4000 Hz	Present and Absent	Present
5000 Hz	Absent	Present
6000 Hz	Present	Absent and Borderline
7000 Hz	Present	Present and Borderline
8000 Hz	Absent	Present
10000 Hz	Absent	Present

Top: CT head revealing bilateral temporal lobe infarcts
Bottom: Table of bilateral otoacoustic emissions.

CONCLUSION

This case report makes a significant addition to the literature regarding this rare finding. Cortical deafness can act as a barrier to rehabilitation, as it can be confused for cognitive deficits. Due to early recognition with audiologic testing, therapy strategies were adjusted which ultimately led to functional improvement. It is vital for clinicians to keep this diagnosis on their hearing loss differential due to the potential for promising recovery.

REFERENCES

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