An unusual presentation of epilepsy

F J UDDIN, C NOGUEIRA, A SAMA

Department of Otorhinolaryngology, Queen's Medical Centre, Nottingham, UK

Abstract

Objective: To report a case of fluctuating hearing due to auditory aura, as an unusual presentation of temporal lobe epilepsy.

Methods: Case report and review of English language literature on temporal lobe epilepsy and auditory aura.

Results: A 31-year-old man presented with intermittent symptoms of bilateral fullness in the ears associated with deafness. He was subsequently diagnosed with temporal lobe epilepsy. Further enquiry revealed a family history of epilepsy.

Conclusion: Auditory aura as a presentation of temporal lobe epilepsy is rarely encountered in otolaryngological practice. This case highlights the importance of obtaining detailed information on epilepsy, including any family history of epilepsy, as a routine part of history-taking in patients presenting with fluctuating hearing loss.

Key words: Auditory Aura; Epilepsy; Deafness

Introduction

Epilepsy is a common neurological disorder; in developed countries, the incidence ranges from 24 to 53 per 100 000 person-years. Auditory auras are commonly associated with temporal lobe epilepsy; however, they are also a rare feature of partial seizures. Florindo *et al.* studied 8000 patients with epilepsy, only 121 of whom reported an auditory sensation at the onset of seizures. The relatively low incidence of auditory aura probably accounts for the limited literature on the subject.

We report the case of a 31-year-old man referred to our department with short episodes of bilateral deafness, which subsequently transpired to be auditory auras.

Case report

A 31-year-old man had originally presented to the out-patient department with occasional symptoms of bilateral fullness in the ears associated with deafness. These episodes lasted 30 seconds to one minute. There was no history of vertigo. The patient had normal otoscopic findings, and pure tone audiometry and tympanometry were within normal limits. His past medical history included allergic rhinitis and seasonal asthma, and anterior rhinoscopy was consistent with this. There was no abnormality of the nasopharynx.

The patient was diagnosed with probable eustachian tube dysfunction, and was given an explanation of his symptoms.

Eights months later, he presented to the neurology department with a history of further episodes of bilateral ear fullness followed by generalised seizures. At this point, his sensation of bilateral ear fullness was diagnosed as an auditory aura. Further enquiry into his family history revealed that his paternal uncle and a cousin also suffered from epilepsy.

Computed tomography and magnetic resonance imaging brain scans were undertaken but were unremarkable. An electroencephalogram showed intermittent slow waves in the left temporal region but no paroxysmal activity.

The patient was commenced on lamotrigine and his seizures ceased. His auditory auras continued for a further year but resolved with an increase in his medication dose.

Discussion

In 1876, Hughlings-Jackson reported two cases of epileptic auditory aura.³ The term aura has been defined as a 'qualitative disturbance undergone by the consciousness, a consequence of the onset of a partial epileptic seizure'.⁴ Epileptic auditory auras may be elementary or complex. Elementary auditory auras are simple noises such as ringing, buzzing, or hissing, and more commonly arise from the temporal lobe, presumably in or around Heschl's gyrus (the primary auditory cortex).^{2,5} Auditory distortions (hyper- or hypoacusis) and (rarely) deafness can also occur.^{2,6} Complex auditory hallucinations are characterised by the presence of hallucinations or illusions of more complex sounds, such as music or voices.^{2,7}

An increasing body of evidence suggests that genetic factors play an important role in temporal lobe epilepsy. Auditory aura may be a manifestation of autosomal dominant partial epilepsy with auditory features, which in some families is linked to mutations in the leucine-rich glioma inactivated 1 gene. ^{8,9}

Our patient had initially been diagnosed with eustachian tube dysfunction due to his symptoms of ear fullness and deafness. This case highlights a rare presentation of temporal lobe epilepsy with short-lived, episodic auditory symptoms.

We recommend that enquiries regarding previous epilepsy, and any family history of epilepsy, should be a

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routine part of history-taking for patients presenting with tinnitus or fluctuating hearing loss.

- Auditory aura is a rare presentation of temporal lobe epilepsy
- Auditory aura may be a manifestation of autosomal dominant partial epilepsy with auditory features
- The reported patient presented with auditory symptoms due to temporal lobe epilepsy
- All patients presenting with fluctuating hearing loss should be questioned about previous epilepsy and any family history of epilepsy

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Address for correspondence: Miss C Nogueira, Department of Otorhinolaryngology, Queen's Medical Centre, Nottingham NG7 2UH, UK

E-mail: claudia.nogueira@nuh.nhs.uk

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