Unpleasant auditory illusions and related avoidance behaviour in a child

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ABSTRACT – Auditory aura is a rare symptom in focal epilepsy. It has been described in autosomal dominant partial epilepsy with auditory features, but is, in general, poorly documented. We report on a 7-year-old, right-handed boy, who suffered seizures characterized by positive auditory illusions with verbal and gestural automatisms and noticeable attempts at covering his ears. Clinical evaluation and video-recording of the seizures, confirmed that most of the ictal behavior was deliberately directed at trying to prevent the unpleasant sensations reaching his ears. [Published with video sequences]

Key words: auditory illusions, temporal lobe, partial seizures, automatisms, aura

Auditory aura is considered a symptom of temporal lobe epilepsy. However, probably due to its low incidence (1.3% of 222 surgical patients in the series of Penfield and Kristiansen, 1951) and its doubtful lateralizing value (Florindo et al. 2006), few authors have focused on this topic. Interest in auditory aura has increased considerably since it was recognized as a manifestation of autosomal dominant partial epilepsy with auditory features (ADPEAF) (Ottman et al. 1995, Flex et al. 2005), which, in some families, is linked to mutations in the leucine-rich glioma inactivated (LGI1) gene (Berkovic et al. 2004).

Recently, sporadic cases lacking these mutations have been described, suggesting that auditory aura might be a more frequent and complex symptom than previously recognized. In the series described by Bisulli et al. (2004), in many patients diagnosis had been delayed by months or years, and a misdiagnosis of idiopathic generalized epilepsy had been formulated in 11% of cases.

We report on a 7-year-old boy, who presented with seizures characterized by positive auditory illusions, accompanied by reactive verbal and gestural behaviour. A video-recording of one such seizure is supplied.

Case study

A 7-year-old child with no family history of epilepsy and normal development was referred to the Pediatric Neurology Unit because of recent onset (3 weeks), daily episodes of short duration (10-20 seconds), both diurnal and nocturnal, characterized by positive auditory sensations of hearing simple and complex sounds from his surroundings (usually unfamiliar voices), which became louder and louder, causing such obvious discomfort that the child put his hands over
his ears. He then covered his eyes, and his facial expression indicated he was in pain. Sometimes screaming, crying and complex gestural automatisms followed. A previous diagnosis of ‘behavioural disturbances’ had been made elsewhere.

We performed prolonged scalp video-EEG monitoring and recorded more than thirty episodes, diurnal and nocturnal. Interictal EEG (figure 1) disclosed a well-structured background activity with sporadic, left fronto-temporal spikes and spike-wave complexes. Some seizures were very brief, consisting simply of auditory illusions during which the child put his hands over his ears, while the EEG demonstrated only muscular artefacts. On other occasions (see video sequence), seizures lasted one or two minutes, and after an initial auditory phase, screaming and gestural automatisms ensued. Ictal EEG (figures 2 and 3) showed an artefact when the child put his hands over his ears, which was followed by diffuse voltage attenuation, bilateral anterior rhythmic delta activity and then by spike-wave activity with left predominance. The ictal discharge usually stopped when the child quietened down, with immediate recovery of contact and the capacity to follow commands and correctly answer questions.

Brain MRI was normal. Genetic analysis for LG11 mutations gave negative results. Carbamazepine treatment led to progressive seizure improvement over a one month period, with only two seizures having occurred in the last two months.

Discussion

Auditory aura is a rare feature in partial seizures and has been rarely documented by video-EEG recordings. In a recent study (Florindo et al. 2006), among 8,000 patients with epilepsy, 121 reported an auditory sensation at seizure-onset and only 14 of these described positive illusions. In Florindo’s series, the auditory aura was often dynamic, evolving in character and intensity; sounds were very similar to a true sensory experience; complex hallucinations and illusions were commonly characterized by speaking, singing, or crying and were described as familiar. Likewise the symptoms described by the child reported

![Figure 1. Interictal EEG showing a well-structured background activity with sporadic left fronto-temporal spikes and spike-wave complexes.](image-url)
Figure 2. Ictal EEG showing an artefact when the child puts his hands over his ears, followed by a diffuse voltage attenuation and then, by bilateral anterior rhythmic delta activity, replaced by spike-wave discharge, with left predominance.

Figure 3. Ictal EEG showing an artefact when the child puts his hands over his ears, followed by a diffuse voltage attenuation and then, by bilateral anterior rhythmic delta activity, replaced by spike-wave discharge, with left predominance.
here, resembled true auditory experiences but were unpleasantly intense. Auditory illusions in children are reportedly bilateral, and we found it difficult to lateralize seizure activity, as also reported in the literature (Loddenkemper and Kotagal, 2005; Florindo et al. 2006). Conversely, ictal auditory illusions do seem to have localizing value and point to the lateral, rather than mesial, temporal lobe. Recently, Gupta et al. (2006) described an adult patient with auditory auras, who was seizure-free after invasive recordings had prompted resection of the left mid-superior temporal gyrus.

Bancaud (1987) hypothesized that auditory illusions occurred preferentially when epileptic discharges were widely extended over the superior temporal gyrus, while auditory hallucinations were likely to occur when discharges were more limited in extent. According to this hypothesis, more recently, auditory illusions have been associated with so-called “temporal plus seizures” (Barba et al. 2007), i.e. seizures involving a complex epileptogenic network including the temporal lobe, as well as immediately surrounding structures (Ryvlin and Kahane, 2005) and in particular to temporo-parieto-occipital seizures. In our patient, the ictal discharge remained localized to the anterior temporal leads, making temporo-parieto-occipital involvement unlikely.

Auditory aura is a symptom of ADPEAF (autosomal dominant partial epilepsy with auditory features) (Berkovic et al. 2004, Flex et al. 2005), which, in some families, is associated with mutations in the leucine-rich glioma inactivated (LGI1) gene. However, in the sporadic patients, such as the boy reported here, analysis of LGI1/epitempin-coding exons failed to disclose any mutation (Bisulli et al. 2004).

An additional reason to report our patient is that he had been initially diagnosed as being affected by behavioural disturbances as a consequence of the recurrent avoidance behavior that accompanied the auditory symptoms. A recent study (Askenazy et al. 2007), demonstrated a high prevalence of auditory hallucinations in pre-pubertal children attending psychiatric clinics and related these manifestations to both anxiety and psychosis. However, a detailed analysis of ictal semiology by epileptologists and, where appropriate, by video-EEG monitoring, may be particularly helpful for differential diagnosis.

Legend for video sequence
The child is lying on the bed, eyes open and relaxed. Suddenly, he puts his hands over his ears, with a pained expression on his face; he then calms down, but after a few seconds, he once again displays facial expressions of pain. He closes and covers his eyes with his hands, moaning more and more loudly; finally, he suddenly sits down, screaming and bending his arms. Then he quietens down and when his father asks him what he had been hearing, he replies “TV”. A star at the left upper corner indicates video sequences corresponding to the ictal EEG displayed in figures 2 and 3.

References


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