Ictal SPECT in a case of pure musicogenic epilepsy

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ABSTRACT − A 39-year-old, right-handed woman had seizures for two years which were always triggered by exposure to various types of music: the first occurred while she listened to a tune she particularly liked, Con Te Partiro, by Andrea Bocelli. Other triggering factors were various types of music such as supermarket background music and polyphonic singing or instrumental music played by family members. The seizures had a stereotyped course: she felt anxious, tearful, then occurred slight obtundation, during which she smacked her lips and moved restlessly. There was no complete loss of consciousness, but some degree of amnesia. She never experienced a generalized tonic-clonic seizure, but reported rare spontaneous feelings of déjà-vu that had begun at the same time as the induced seizures. There were no other spontaneous attacks; only one seizure was apparently provoked, not by music but by a loud background noise in her office. She was a music lover and a singer. Interictal EEG showed independent slow waves over the temporal regions. Several seizures with EEG localisation over the right temporal region were elicited after several minutes of exposure to music. Monoauricular stimulation with the same music produced a seizure when applied to the left ear but was ineffective when applied to the right ear. Ictal SPECT demonstrated right temporal hyperperfusion. MRI was normal. On high dose of carbamazepine, seizure frequency decreased. The addition of topiramate resulted in full seizure control. Musicogenic epilepsy is a rare form of reflex epilepsy. Pure cases, when patients do not experience unprovoked seizures, are exceptional. Our report confirms the implication of the right temporal lobe in this epilepsy. [Published with video sequence].

KEY WORDS: musicogenic epilepsy, temporal lobe epilepsy, reflex seizures, SPECT

Observation

A 39 year-old, right-handed woman was referred for the assessment of drug-resistant epilepsy. She complained of seizures which had begun two years earlier, persisted under treatment and had a very stereotyped and reproducible course. The first at-
A first seizure was elicited by a few minutes exposure to theta slow waves or slow spikes over the temporal regions. Interictal EEG demonstrated the presence of independent having a seizure. Unable to listen to music for more than a short time without deprivation of music in her home. Two typical seizures are presented on the video. She was born at term to healthy, non-consanguinous parents with no family history of epilepsy or other neurological disorder. Her medical history revealed that she had been treated for an auto-immune thyroiditis with low-dose thyroxin for the past 5 years. At the age of 37, several months before seizure onset, she had a sudden, bilateral hypoacusia predominating in the right ear, for which she consulted a doctor, but for which she was given no precise data with regard to the onset of musically-triggered seizures. Excluding 33 cases for whom the stimulus is represented by the excessive loudness and the resulting seizures are wild running followed by convulsions [2]: these models do not have much in common with musicogenic epilepsy. In humans, the most common example of sound-induced seizures is found in startle epilepsy, a condition characterized by seizures induced by sudden and unexpected stimuli, such as contact or noise, usually in patients with extensive brain damage and hemiplegia [3]; the auditory stimulus is non-specific, surprise or suddenness being the critical element. Other effective auditory stimuli include voice, tone, frequency or sound-induced seizures such as a whirring or ringing machine. Musicogenic epilepsy is a particularly rare form of complex reflex epilepsy identified in 1937 by Critchley [1]. Wieser et al. (1997) reported, from the literature and their personal experience, 83 patients with musicogenic or tone/sound-induced seizure [4]. Of the 83 patients, 78% reported music was the only seizure-provoking stimulus, but most of these patients also had spontaneously occurring seizures. Four percent reported tone as the provoking stimulus. In 4%, it was sound and in 14% “all kinds of music and sounds”. Excluding 33 cases for whom precise data with regard to the onset of musically-triggered seizures were missing, 36 subjects had both musicogenic and unprovoked seizures, while only 14 of the 83 patients (17%) had seizures exclusively triggered by music. Musical triggers are often variable but may be very specific for a patient: a discrete frequency band of church bells [5], brass wind instrument playing low notes [6], hillbilly
organ [7], the Marseillaise [8], String of Pearls by Benny Goodman [9], voice of a TV personality [10]. According to Jallon et al. (1989), seizures can be precipitated by the type of music (classical, lyric, religious, military, jazz, musical comedy), the type of instrument (piano, violin), the kind of music (Cheerful, sad, sentimental) or by a specific composer (Wagner, Beethoven) [8]. Sometimes, but very rarely, thinking about a type of music is enough to precipitate a seizure [8]. Affective components seem to be very significant in some cases. Wieser et al. (1997) reported a high prevalence of musicogenic epilepsy in music lovers. Among their 83 patients, the musical standard was relevant described in 35: four (11%) were professional musicians, 11 (31%) were amateur musicians, five (14%) were “music fans” and seven (20%) had an above average interest in music. Only eight patients declared that they were not especially interested in music.

From the study of Wieser et al. (1997), this epilepsy appears to be more frequent in women (females/
males = 54/46%). Seizure onset is in adulthood at a mean age of 27.7 years (12.5 years). Forty-four patients had a clear description of the localisation. Onset was found in the temporal lobe in 75% of cases, and seizure focus was more often right (61%) than left [4].

Ictal SPECT was performed in a patient by Wieser et al. (1997) and showed a right anteropolar temporal hyperperfusion [4]. Another ictal SPECT was reported by Genc et al. (2001) and demonstrated a right anterior and mesial temporal hyperperfusion [11]. Both author groups proposed the participation of the mesial temporal structures in the production of musicogenic epilepsy: this hypothesis is sustained by our observation, since our patient reported some spontaneous episodes of déjá-vu and had lip smacking and swallowing.

The physiopathology of seizures induced by music is unclear. The latency observed between the stimulus and the seizure is variable, from a few seconds to several minutes, longer than for simple reflex seizure. Musicogenic epilepsy appears to represent a peculiar form of complex reflex epilepsy [12]. Dichotic listening allows us to distinguish a main, crossed auditory pathway and a less important, ipsilateral auditory pathway. In our case, the monaural stimulation, controlateral to the epileptic focus, produced a simple partial seizure, whereas the ipsilateral stimulation was ineffective. This allows us to conclude that biauricular stimulation is not necessary to produce a seizure. To further validate the strongly lateralized sensitivity in this patient, a dichotic test should be performed. Our patient had right temporal lobe epilepsy. It is in accord with the preponderance of right-sided seizure origin, as reported in the literature [4] and that the right hemisphere is proved regularly superior to the left hemisphere in the identification of non verbal sounds in dichotic listening [13]. The only interesting history as regards our patient consisted of an unclear history of brief, spontaneously regressive right-sided hearing loss: this may point to the existence of inflammatory factors. The lack of clear EEG changes over 10-15 seconds at clinical seizure onset followed by high-voltage sharp 5-7 Hz theta rhythm over the anterior temporal region, is the most common pattern seen in medial temporal lobe seizures [14]. The high prevalence of significant emotional factors among patients with ME is also in favour of the implication the limbic system.

**Conclusion**

Musicogenic epilepsy is a rare entity among the large category of reflex epilepsies. Pure cases with no associated spontaneous seizures are exceptional. Our observation confirmed the implication of the right temporal lobe in this epilepsy. □
Sequence 1: Seizure triggered by listening to Verdi’s Nabucco. The clinical attack consisted of a conscious manifestation of anxiety and discomfort, followed quickly by lip smacking and swallowing. The EEG (see figure 1) was normal at clinical onset, with appearance, 20 seconds after the first symptoms, of a rhythmic theta discharge over the anterior right temporal leads. The patient did not remember the full duration of the seizure, and regained contact immediately after the cessation of the musical stimulation.

Sequence 2: Seizure triggered by listening to Conte Te Partiro with ictal HMPAO SPECT (see figure 2). The seizure occurred approximately 1.5 minutes after the beginning of the song. The first symptom is a change in mimic with a slight body movement, the patient signals seizure onset. There is no change on the EEG at this moment. Consciousness and contact appear normal. Early occurrence of lip smacking and oral automatism. About 20 seconds later, appearance of rhythmic slow wave activity over the right anterior temporal region. Oral automatisms persist, and there is a slight obtundation, later confirmed by the patient. The injection is performed 25 seconds after the onset of the EEG changes. At the end of the seizure, the patient is immediately responsive.

References


