

Gelastic seizures with dancing arising from the anterior prefrontal cortex

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ABSTRACT – *Aim.* This case report provides insight into the function of the anterior prefrontal cortex (aPFC), specifically Brodmann Area 10 (BA10), and its interconnectivity. *Method.* We present a 10-year-old patient with lesional epilepsy and ictal onset, localised to BA10 in the aPFC. *Results.* Thirty-four seizures were recorded. All seizures involved a demonstration of elation with laughter that was associated with a variety of different patterns of complex motor behaviour that included performing specific celebratory movements and acting out a Michael Jackson dance move. Electrographically, the seizures were all stereotyped and arose from the right frontal region, followed by a distinct left temporal ictal rhythm that corresponded with the onset of the behaviours. The lesion in the right aPFC was identified as a mixed lesion with both dysembryoplastic neuroepithelial tumour cells and type II cortical dysplasia. *Conclusion.* The electrographic analysis and unique seizure semiology suggest a connection between the aPFC and the contralateral temporal lobe. This neural pathway appears to be involved in the activation of previously formed procedural memories, creating an intensely positive emotional experience.

Key words: temporal lobe epilepsy, gelastic seizures, anterior prefrontal cortex, Brodmann area 10, neural networks

The anterior prefrontal cortex (aPFC), Brodmann Area 10 (BA10), is one of the least understood areas of the brain, and its connectivity to other cortical regions has yet to be fully elucidated. We present a patient with focal epilepsy arising from a neoplasm in the right aPFC. The patient's seizure semiology is unique as it consisted of specific

sets of patterns of complex motor behaviours that included laughter. This semiology exhibited components comparable to gelastic and ecstatic seizures. The electrographic seizure itself provides insight into neural circuitry involved in generating the emotional state and the retrieval of related specific memories and actions.

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Case study

A 10-year-old, right-handed boy presented with episodes described by his parents as “goofyz behaviour and a decline in academic performance over the preceding year.

During the “goofy” behaviour, the patient would fidget and laugh while distracted by his surroundings, becoming “inattentive”. The patient was amnesic of the events. During some episodes, he would emulate a Michael Jackson dance move in which he grabbed his groin and sang out loud, leading to reprimands at school. Our patient was very familiar with Jackson’s music and dance moves and had practiced them many times previously. His “goofy” behaviours increased and school performance declined, despite treatment with stimulant therapy and antiepileptic drugs (carbamazepine and clobazam). Neuropsychological deficits were identified for left-hand fine-motor skills, perception of faces, figural memory, and executive functions for planning and initiation of problem solving. Brain imaging revealed a right frontopolar lesion (*figures 1A and 1B*).

During three days of continuous video-EEG monitoring, 34 seizures were recorded, far in excess of what the parents had noticed prior to admission. The onset of each event was characterised by fidgeting or repositioning of the body, followed by laughter that appeared mirthful in the absence of external stimuli (the patient could not recount his feelings during the

seizure event as he was amnesic). Then, different patterns of complex behaviours, related to a positive personal experience, would occur. For example, he would imitate a famous Michael Jackson pose (as above), or would clasp his hands and rotate them in a circular motion and say “oh yeah” while laughing. Events were 7-12 seconds in duration and all exhibited a distinctive pattern of stereotyped movements, which were acted out in a specific sequence. Electrographically, seizure onset was marked by a generalised spike that was maximal in the frontal region bilaterally and lasted approximately one second. On review of this generalised spike on an expanded time scale, there was a consistent lead in from the right frontal region with a maximal negativity at F4 (*figures 2A and 2B*). Afterwards, there was a three-second period of diffuse attenuation of activity, followed by 2.5-Hz rhythmic spiking, originating from the left temporal lobe which was maximal at T3 (*figures 2A and 2B*). The onset of the clinical behaviour was synchronous with the onset of this left temporal ictal pattern. The left temporal activity and the clinical event would last up to 10 seconds.

Due to the intractable nature of his epilepsy, the patient underwent a lesionectomy. Pathological analysis revealed the lesion to be a dysembryoplastic neuroepithelial tumour (DNET) mixed with cortical dysplasia. The patient has remained seizure-free since the surgery, a time of approximately 18 months.

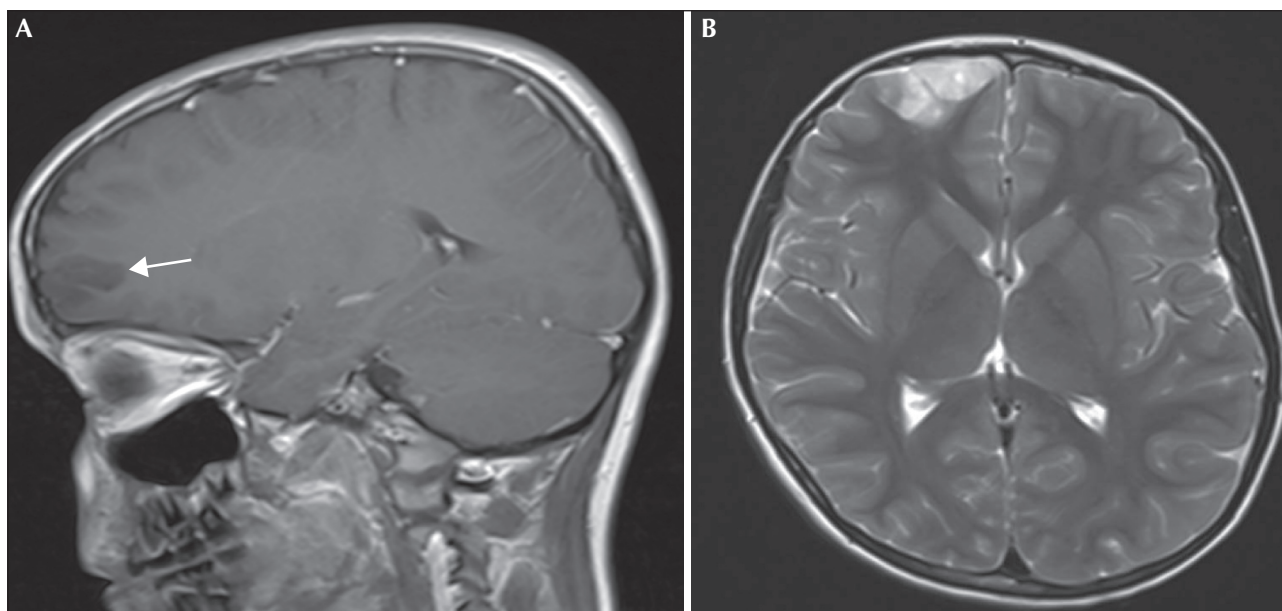


Figure 1. (A) T1-weighted sagittal MRI showing a hypodense cortical lesion in the right medial frontal lobe (white arrow designates the posterior edge of the lesion). (B) T2-weighted axial MRI showing the same lesion as a hyperintense cortical lesion in the right medial frontal lobe, corresponding to Brodmann Area 10 in the aPFC.

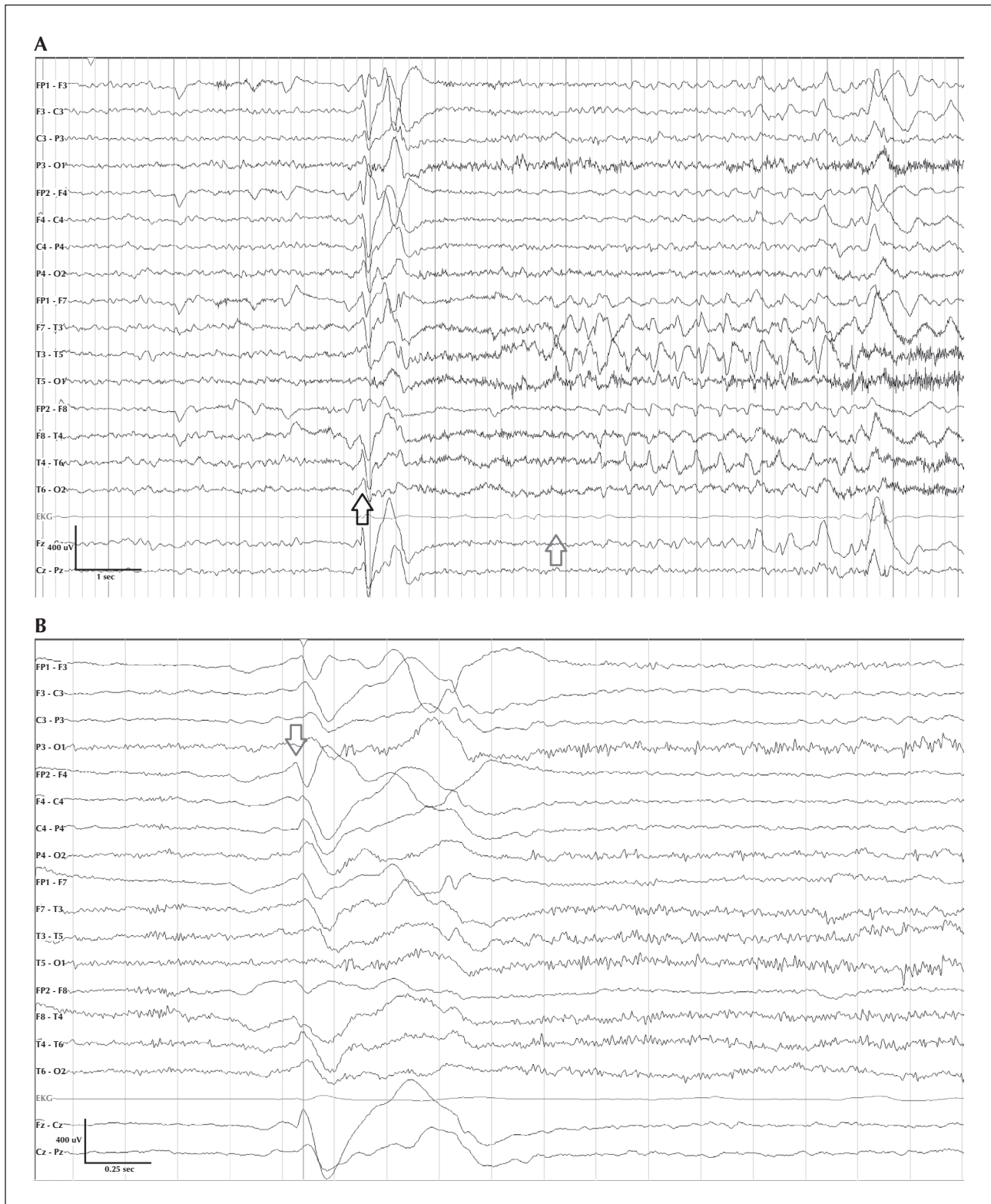


Figure 2. (A) Standard scalp EEGs, longitudinal montage. Electrographic seizure onset (black vertical arrow) is represented by generalised discharges with a slight right frontal lead-in (see *figure 2B* for expanded timeframe), followed by rhythmic 2.5-Hz left temporal spiking with phase reversal at T3 at +3 seconds from onset. Clinical onset is marked by vertical white arrow, which is time-locked with temporal (T3) spiking. (B) Standard scalp EEGs; longitudinal montage with expanded timeframe (see scale). In this image, the generalised discharge is marked with a white vertical line. Slightly preceding this discharge is a frontopolar lead with maximal negativity at electrode F4 (white vertical arrow), representing the first electrophysiological change and ictal onset.

Discussion

Although the patient's seizures contained elements of gelastic and ecstatic seizures, they defy current semiological classification. Key features of all of his seizures were laughter and displays of happiness and joy, moreover, the actual behaviours with each seizure varied from a re-enactment of a Michael Jackson dance move to other celebratory gestures, such as nodding and clapping hands. These behaviours appear as learned motor patterns based on procedural memory, one of the types of memory that the aPFC cortex is involved in generating (Uylings *et al.*, 1990). The clinical behaviours occurred after the spread of the ictal pattern to the left temporal region (figures 2A and 2B). This pattern and the unique seizure semiology suggest the function of the aPFC and how it may interconnect with various cortical regions.

The aPFC is hypothesized to be a centre of higher cognitive functioning, but a lack of "hard" neurological deficits makes precise and specific functions difficult to establish (Spiegel *et al.*, 2009). The "elusive" function of BA10 may be based on the supramodal integration of multiple higher cognitive functions, and, as such, connection with areas such as memory would be crucial for its actions (Ramnami and Owen, 2004). In addition, the proposed "gateway theory" implicates BA10 and the aPFC in distinguishing between one's experiences and thoughts (Burgess *et al.*, 2005). Impairment in this region thus causes one to react to one's thoughts or memories as though they were real experiences. The aPFC has been linked to generating positive emotions in association with reward and regulating the intensity of those feelings. This is most intense when comparing one's self to friends or famous people (Morgan, 1990; Benoit *et al.*, 2010), as with the patient and Michael Jackson. Given these hypothesized functions, his ictal pattern suggests that the interconnection between the aPFC and the left temporal lobe is critical in the production of positive emotional responses and recall of specific memories.

While the surface EEG suggests the primary symptomatogenic zone is the left temporal lobe, elements of our patient's seizures make this assertion overly simplistic. The aPFC controls aspects of inhibition and appropriateness of sexual behaviour through the orbitofrontal pathway. Lesions in this pathway (particularly in the right hemisphere) yield symptoms including disinhibition, sexual inappropriateness, and emotional lability (Spiegel *et al.*, 2009). The dance move performed by our patient may therefore reflect an ictal release of inhibition through disruption of this pathway and play a significant role in generation of his unique semiology.

Ictal laughter is the essential component of gelastic seizures (Daly and Mulder, 1957). It is classically

caused by hypothalamic hamartomas, although several other pathologies and locations have been reported to be implicated in causing gelastic seizures (Daly and Mulder, 1957; Akman *et al.*, 2002; Cheung *et al.*, 2007; Chen *et al.*, 2011; Cook and Joshi, 2011), moreover, separate neuronal pathways for voluntary and involuntary laughter have been theorized (Poeck, 1985). Ictal laughter that is without mirth and unmotivated is more often reported in gelastic seizures arising from the frontal lobe, electrographically involving mesiofrontal structures, the cingulate gyrus, and the supplementary motor area (SMA), whereas emotional, *mirthful* gelastic seizures involve structures of the temporal lobe: the amygdala, parahippocampal gyrus, fusiform gyrus, and so on (Kovac *et al.*, 2009). As our patient was amnesic of his seizures, he could not confirm any correlation between his mood and events during the seizures, but there appears to be an emotional quality to his seizures that may reflect the left temporal spread of his ictal EEG pattern. Additionally, the pathway for perception of laughter, hypothesized to originate in BA10 and descending through the brainstem with involvement of temporal lobes bilaterally, reflects, in part, the ictal circuit observed in our patient that generates ictal laughter with apparent mirth, without retained memory of the seizure (Wild *et al.*, 2003).

Finally, ictal dancing is extremely rare and has previously been reported with temporal lobe epilepsy (Bagla *et al.*, 2009). However, the complex motor behaviour required to generate a purposeful dance suggests the involvement of frontal lobe structures and pathways. Our patient's ictal circuitry, connecting the right aPFC and the left temporal lobe, reflects the complex connections that may be required to generate such a unique semiology. □

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None of the authors have any conflicts of interests to declare.

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