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Chess-playing epilepsy: a case report with video-EEG and back averaging

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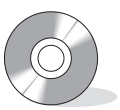
ABSTRACT – A patient suffering from juvenile myoclonic epilepsy experienced myoclonic jerks, fairly regularly, while playing chess. The myoclonus appeared particularly when he had to plan his strategy, to choose between two solutions or while raising the arm to move a chess figure. Video-EEG-polygraphy was performed, with back averaging of the myoclonus registered during a chess match and during neuropsychological testing with Kohs cubes. The EEG spike wave complexes were localised in the fronto-central region. [Published with video sequences]

KEYWORDS: juvenile myoclonic epilepsy, chess-playing, reflex seizures, praxis-induced seizures, back averaging

A 29-year-old, right-handed Caucasian man with no personal or family history of epilepsy, and with a normal neurological examination and a normal CT scan, started, at age 16, to present with brief, arrythmic, asymmetric myoclonus, mainly of the upper limbs. Playing video games clearly promoted myoclonus. One year later, a first, generalized tonic clonic seizure (GTCS) occurred on awakening. Some months later, a second GTCS was seemingly triggered by repetitive light. Valproate monotherapy was started, and the patient went into remission. Valproate was stopped at age 28 and myoclonus reappeared, aggravated by playing chess. It is interesting to note that myoclonus occurred not at the beginning of a chess match, when fairly standardised moves are made, but later, when he had to plan his strategy, especially when he had to choose between alternatives.

The baseline standard EEG in this drug-free patient showed bilateral, diffuse spike wave and polyspike wave discharges. No photoconvulsive response was obtained. While playing chess, numerous spike wave complexes were recorded, either with no clinical manifestations or with myoclonus involving mainly the proximal part of the upper limbs, asymmetrically. Valproate monotherapy was restarted and myoclonus stopped.

Two EEG-polygraphies were then carried out while playing chess, after 36 and 48 hours without valproate respectively. In addition, during the first of the two EEG-polygraphies, a series of neuropsychological tests were performed (Rey's figure on copy and by memory followed by WechslerMemoryScale-R: figurative memory, Kohs cubes and Raven Standard Progressive Matrices). The EEG-polygraphy consisted of a simultaneous recording of a video-EEG and



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electromyographic recording of the right and left deltoid muscle or the brachioradialis muscle. The patient only experienced myoclonus while manipulating Kohs cubes, accompanied by spike and wave discharges on the EEG, on test and retest *(video sequence 2)*.

During EEG-polygraphy, myoclonus was often registered at the very moment when he started to lift his arm in order to seize a figure on the chessboard and make a move *(figure 1* and *video sequence 1)*. For both types of this stimulation paradigm, i.e. chess playing and manipulating Kohs cubes, the patient reported that after each myoclonus he loses his mental preparation, he feels "initialised", and obliged to restart his problem-solving strategy from zero.

Twenty one myoclonic jerks, clinically observed and registered on the EMG, triggered the back averaging of cortical, time-locked spikes. This back averaging showed bilateral cortical spikes in the fronto-central cortical area *(figure 2)*. In our case, the amplitude of the cortical spike was almost symmetrical, even if the myoclonus was clinically observed most often on one side. Bilateral myoclonus was observed following cortical spikes of higher amplitude.

The visual pattern of the chessboard does not seem to be a provocative factor by itself. No EEG activation was observed in our patient under the following conditions: looking at the wooden chessboard or at the one on the computer screen, intermittent light stimulation, or visually-evoked potentials using an alternating checkerboard pattern.

Discussion

This patient fulfils the diagnostic criteria for juvenile myoclonic epilepsy (JME). Playing chess and manipulating Kohs cubes could trigger EEG discharges and myoclonus. His epilepsy is well-controlled by 500 mg of valproate daily.

Inoue *et al.* [3] reported on 21 cases with "praxis-induced" seizures, selected by a history of seizures induced by non-verbal, higher brain activities.

A systematic, prospective investigation of seizures, induced by higher brain activities has been carried out by Matsuoka *et al.* [4]. They found, in a series of 480 consecutive patients with epilepsies of various types, 38 pa-

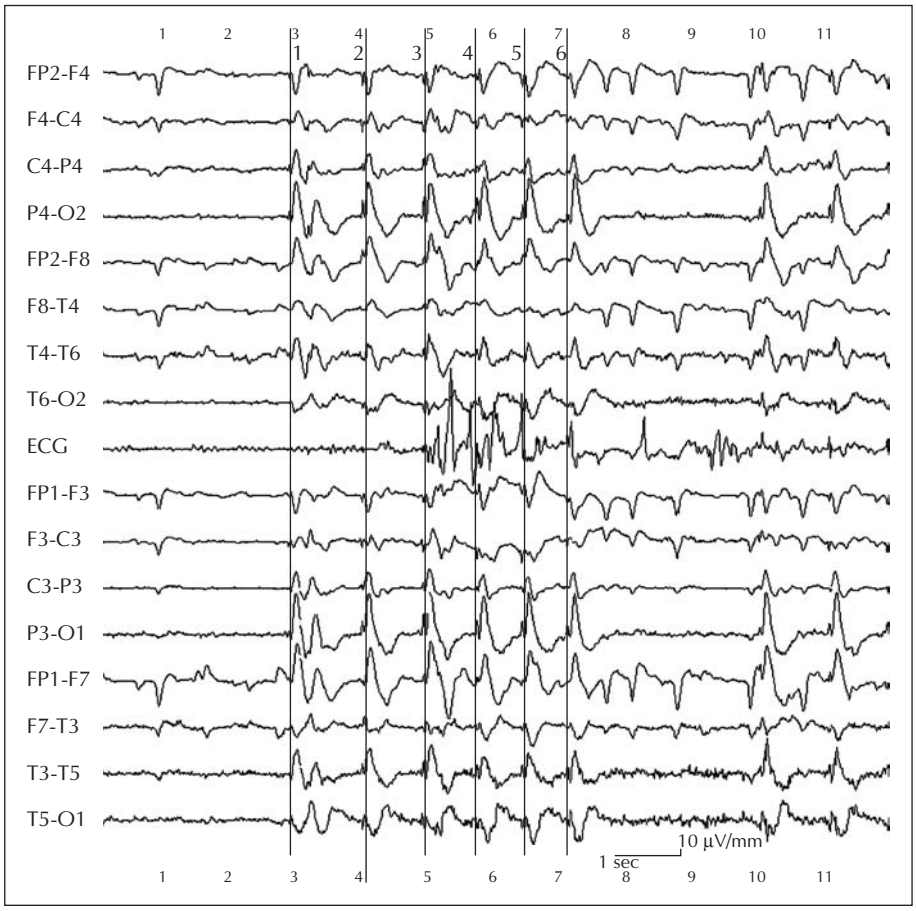

Figure 1. *Standard EEG during a chess game (amplitude: 10 microvolts/mm, time constant 0.1, 15 Hz filter): repetitive spike-wave discharges are maximum on fronto-central areas on both sides.*

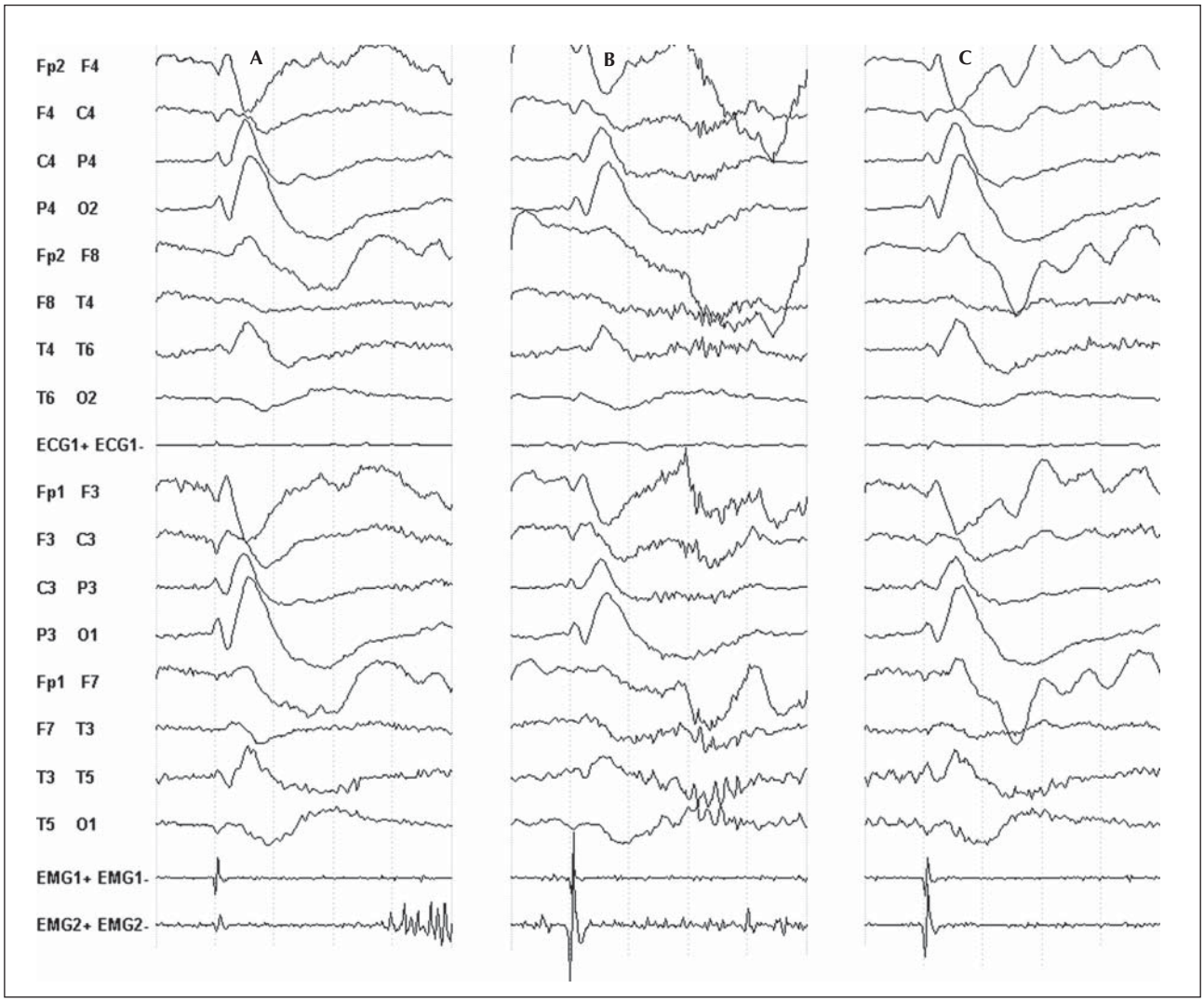

Figure 2. *A) Back averaging of n = 11 myoclonia of the right upper limb: the cortical spike is located under the electrodes C3 and C4 (phase reversal between F4-C4 and C4-P4 and between F3-C3 and C3-P3; B) Back averaging of n =three myoclonia of the left upper limb: the cortical spike is located under electrode C3 and C4 (phase reversal between F3-C3 and C3-P3 on the left hemisphere, and between F4-C4 and C4-P4 on the right hemisphere); C) Back averaging of n = seven bilateral myoclonia of the upper limbs: the cortical spike is located under electrode C4 (phase reversal between F4-C4 and C4-P4, spike on the opposite hemisphere partially hidden by the noise).*

tients (7.9 %) displaying "neuropsychological EEG activation" (NPA activation) i.e. neuropsychological tasks provoking epileptic discharges without (14/38 cases) or with (24/38) seizures, mainly of the myoclonic (15/38 cases) or absence (8/38 cases) type. So, "NPA activation" in patients with epilepsy is a rare phenomenon if all subtypes of epilepsy are considered together. Conversely, in IGE, in particular in JME, this phenomenon is far from exceptional: in the same study 46.7% of the 45 subjects with IGE displayed NPA activation. So cognitive tasks are mostly excitatory in idiopathic generalised epilepsies (IGE), whereas they are usually inhibitory in other types of epilepsies: 131 out of 208 patients (62,9 %) with epilepsies other than IGE and displaying spontaneous paroxysms on the awake EEG, showed a reduction of paroxysmal activity during NPA [4].

Seizures not induced by reflex activation are present in most patients with NPA activation. Various seizure types can be observed: myoclonus, absences and generalised tonic-clonic seizures. Higher brain activities most frequently induce myoclonus, sometimes absences with or without myoclonus and rarely GTCS. Patients with myoclonic seizures (myoclonus alone, myoclonus with absences or GTCS) had, in Matsuoka's study, the highest rate of NPA effect. The response to treatment was considered "generally good" by Goossens [2] and poor in Inoue's study [3]: 69 % of 19 patients with JME and predominantly

praxis-induced seizures were refractory to treatment, an unusually high figure in JME.

Chess, calculation, calculation using a "soroban" (a traditional Japanese calculator), thinking, writing, spatial tasks, card or board games etc. have been reported to be seizure triggers. There might be two forms of IGE seizure induced by higher mental activity: seizures induced by thinking and spatial tasks and seizures induced by writing, written calculation or drawing requiring action-programming activity. It is not clear, whether these two forms show a distinct mechanism or represent two ends of a pathophysiological continuum [4].

Neuropsychological EEG activation is fascinating in that it investigates the link between thinking and acting. Playing chess might be a particularly efficient stimulus since it implies action programming and a subtle motor action. As a matter of fact, a neuropsychological test having similar characteristics, i.e. manipulating Kohs cubes, clearly provoked myoclonus in our patient and proved in Matsuoka's study to be the most efficient neuropsychological stimulation paradigm. Inoue *et al.* [3] found that myoclonic jerks could well be predominant or localized in the part of the body concurrently moved or intended to be moved. They concluded that the process of transcoding thinking into voluntary or intentional acts seems to be essential, and therefore they coined the term "praxis-induced seizures". This leads to the hypothesis that it is the co-activation of circuits implicated in action programming and motor activity that trigger myoclonus.

The EEG recording at the time of myoclonus showed, in Rey's case [5], a spike-wave complex localised in the left rolandic area (phase reversal under the electrode C3 and Cz). In Matsuoka's study (2000), the "epileptic discharges in IGE induced by NPA were quite similar to those that appeared spontaneously. They consisted of diffuse and symmetric spike-wave or polyspike-wave complexes that predominated over the central electrode site both with and without lateral asymmetry". Matsuoka [4] cites an interesting case reported by Hasegawa: induced spikes predominated over the dominant central EEG site with letter writing, and over the non-dominant parietocentral EEG site with spatial construction tasks, suggesting that the type of functional cognitive circuits activated may be determinant for the site of consecutive spike generation. In our patient, back averaged spikes are localised bilaterally, quite symmetrically, on the central areas, while myoclonia predominate most often on one side. Transcallosal propagation and/or subcortical neuronal circuits may underlie this fairly symmetrical spike localisation. Back averaging of the cortical spikes is in favour of a stable spike generator, localised next to the central electrodes.

With respect to the localisation of spikes in the central area, it is interesting to note that in skilled chess players, memory circuits seem to be activated in the frontal and parietal neocortex [1]. A working hypothesis might be that this type of memory activation contributes to mechanisms that lead to cortical spike generation and ultimately to myoclonia. Further contribution to our paradigm comes from a combination of a visuoconstructive task, action planning and activation of sensorimotor circuits, elicited by chess playing and Kohs cubes manipulation. □

Legends for video sequences

Video sequence 1 (chess game): (from 9h58′ 55″ to 9h59′15″; 20″ duration) Several bilateral, asymmetric, synchronous myoclonia - abduction of both upper limbs – at the initiation of a movement of the left upper limb in order to make a move on the chessboard.

Video sequence 2 (Kohs cubes): a) 1 isolated myoclonus of the left biceps brachialis (during zoom) b) three myoclonia of the left biceps brachialis muscle without any movement of the arm, while manipulating Kohs cubes.

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