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Differential diagnosis of a paroxysmal neurological event: Do neurologists know how to clinically recognize it?

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ABSTRACT

Purpose: To investigate ability to recognize paroxysmal neurological events (PNE) based on video-recorded events alone in a group of physicians treating prevalent neurological conditions.
Methods: Total of 12 patients' videos (6 epileptic seizures (ES), 4 psychogenic nonepileptic seizures (PNES), 2 other nonepileptic seizures (oNES)) were selected. Videos were displayed once to physicians blind to clinical data and final diagnosis. Physicians determined their clinical choice: ES, PNES, oNES, and I don't know (IDK). When ES was chosen, subjects determined type of ES: focal ES, secondary generalized tonic-clonic seizure (GTCS), primary GTCS, and IDK.
Results: In total 145 physicians (62% female, mean age 46.2 ± 9 years) (neurologists 58.6%, neuropsychiatrists 25.5%, psychiatrists 5%, and neurology residents 10.3%) were enrolled. Physician's exposure to patients with epilepsy per week was diverse: ≤1 patient (43.7%); 1–7 patients (37.2%); >7 patients (14.5%). Reported frequency of observation of PNE was as follows: frequent (21.4%), sometimes (47.6%); rarely (26.9%); never (2.1%). Majority of subjects were not EEG readers (60.7%). Median percentage (Mdn%) of correct answers (CA) was 75% (range 25–100). Predictor of better PNE recognition was higher frequency of clinical exposure to PNE (OR 1.65; CI95% 1.11–2.45; $p = 0.013$). Mdn% of ES CA was 83.3%, (range 33.3–100), and of PNES CA was 50% (range 0–100). Physicians were more accurate in ES than PNES identification ($p < 0{,}001$). Mdn% of type of ES CA was 50%, (range 0–100).
Conclusions: We demonstrate the need for education about clinical features of PNE across subgroups of physicians who deliver neurological service, with emphasis on PNES and ES type classification.

1. Introduction

The gold standard for differential diagnosis of paroxysmal neurological events that may resemble epilepsy is generally considered to be long-term video-EEG monitoring [1]. Nonetheless, in the real clinical world diagnosis relies on the accuracy of seizure description by witnesses [2,3] or on clinical judgment based on direct observation of the events. It has been reported anecdotally that a wrong clinical decision determined by insufficient competence in paroxysmal neurological events may cause not only iatrogenic injury, but also an exposure of patients to the risk of death through inappropriate medical intervention [4]. In addition, it was recently shown that several "putative" psychogenic nonepileptic seizures (PNES) signs and epileptic seizure (ES) signs were neither sensitive nor specific for seizure type [5].

Overall interrater reliability for the diagnosis of ES, PNES, and physiological nonepileptic episodes was moderate among 22 board-certified neurologists and practicing epileptologists at epilepsy centers when exposed to video-EEG data only [6]. Based on growing evidence that video recordings of the events alone could represent a useful clinical tool in neurologically trained medical personnel [7,8], a recent feasibility study showed that in about one-third of unselected cases (23 video footages of sequentially recorded ES, PNES, and other nonepileptic seizures (NES)) correct diagnosis of PNES/ES but not NES can be established on clinical grounds when showed to five neurologists actively practicing in epilepsy centers [9]. Furthermore, it was demonstrated that training of emergency room staff based on a specifically developed, bedside 6-sign diagnostic tool designed for motor PNES identification can significantly improve seizure differentiation [10].

Nevertheless, general neurologists are leading practitioners in the routine clinical work with patients with epilepsy. In a web-based

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epilepsy community survey, it was reported that only 20% of patients were seen by an epileptologist regularly and most (70%) were seen by a general neurologist [11]. Our aim was to explore capability of correct diagnostic prediction of the most frequent paroxysmal neurological events (PNE) based on video examination alone in a group of physicians treating all prevalent neurological conditions.

2. Material and methods

Patient samples were collected at the Epilepsy Center, Neurology Clinic, Medical School in Belgrade, and consisted of 12 patients who underwent long-term video-EEG monitoring. Selection of patients was based on the following criteria: consecutively recorded patients with regards to etiology (ES, PNES, other nonepileptic seizure (oNES)), good quality of video and motor behavior during the PNE. In the case of multiple PNE recorded, videos with the EEG technician standing at the bedside conducting periictal testing were preferred. Final diagnosis was established by the clinical team after reviewing clinical data, video-EEG data, high-resolution brain MRI, and neuropsychological testing. Gold-standard diagnostic criteria were long-term video-EEG monitoring data for ES and PNES, and positive tilt-test for vasovagal syncope. In total, 6 ES, 4 PNES, and 2 oNES were selected (Table 1), based on average etiology distribution across a year at our Epilepsy Center. Each patient video included a minimum of 10 s before and after PNE, including periictal testing when present.

Table 1
Description of paroxysmal neurological events.

Pt. number (Diagnosis)	Video duration/event duration (min:sec)	Description
Pt. 1 (fES) Semiological classification: complex motor (hypermotor) seizure	1:32/0:49	(Male, 14 years of age). Sleeping. After an eye opening right arm tonically elevates in few seconds. This elevation is preceding nonintegrated motor behavior consisting of rhythmic repetitive hyperkinetic movements of trunk associated with bilateral arm tonic elevation. Prolonged postictal aphasia is evident on postictal testing. He does not report aura. (FCD left posterior temporal)
Pt. 2 (PNES)	0:37/0:08	(Female, 21 years of age). Awake. Sudden onset of violent arrhythmic and asymmetric muscle contractions of all limbs in discontinuous pattern and side-to-side head movements that produce body bouncing while eyes are closed. Obeys verbal command immediately after PNES.
Pt. 3 (PNES)	2:18/1:50	(Male, 47 years of age). Awake. After pressing the button to register presentiment, he stops communicating with EEG technician. Bilateral hand behavior that resembles fighting and defense with eyes firmly closed precedes moaning and bilateral asynchronous motor jerking with turn on the left side and then on belly. PNES stops with asymmetrical jerking of both arms with postictal shallow breathing. Does not obey commands after PNES.
Pt. 4 (PNES)	2:15/1:47	(Female, 45 years of age). Awake. Gradual onset of motor behavior of both legs while eyes firmly closed is followed by opisthotonic movements and shaking of both bedrails. PNES continues with very violent pelvic thrusting, body bouncing and loud ictal scream which intensifies with EEG technician's verbal commands (stuttering pattern). Does not obey commands after PNES.
Pt. 5 (sGTCS) Semiological classification: bilateral asymmetric tonic seizure → GTCS	1:27/1:45	(Female, 19 years of age). Sleeping. Sudden onset of right arm tonic elevation while left arm is flexed, head deviates to the left (fencing posture) and eyes are wide open; THIS is followed by bilateral tonic extension of both arms and legs, ictal scream and body tonic flexion that continues to bilateral and symmetric clonic jerking of all limbs which stops synchronously. After sGTCS patient is in postictal coma with heavy breathing. (FCD left mesial frontal)
Pt. 6 (oNES)	1:35/0:44	(Female 21 years of age). Awake. EEG technician is performing venopuncture while patient is sitting. oNES starts with head drop and body inclination to left side (prevented to fall by EEG technician). Both legs are elevated by EEG technician as body is floppy and eyes are closed. She is fully alert immediately after the oNES and obeys verbal commands.
Pt. 7 (fES) Semiological classification: automotor seizure	2:03/1:26	(Female 28 years of age). Awake. Lip smacking movements were almost immediately followed by left hand dystonia. After she takes sitting position as her facial expression changed from neutral to surprise, she becomes able to simply communicate with EEG technician. Postictal cough. Obeys all verbal commands immediately after fES. (HS right)
Pt. 8 (oNES)	1:33/0:54	(Male 23 years of age). Awake. After watching video footage of brain surgery on laptop he starts slightly to stoop on his right side while reporting lightheadedness to EEG technician. Immediately afterwards he takes supine position and obeys all verbal commands.
Pt. 9 (fES) Semiological classification: autonomic seizure → automotor seizure	2:24/2:01	(Male 51 years of age). Awake. Sudden onset of retching (several times) and face and décolleté flushing associated with loss of contact with EEG technician. Minor left arm distal automatisms, right postictal nose wiping and short lasting postictal aphasia are evident in the second half of the fES. Obeys commands and nominates correctly. (HS right)
Pt. 10 (PNES)	2:08/1:31	(Female 52 years of age) Awake. Gradual onset of moaning and bilateral arm elevation associated with asynchronous tremor of all limbs. Eyes are closed. After tremor cessation and shallow breathing right arm elevation and extension is evident. She is not responding to verbal commands given by EEG technician. This is followed by incorrect activity after EEG technician's verbal command.
Pt. 11 (fES) Semiological classification: clonic seizure (left face) → complex motor seizure	1:38/1:01	(Female 27 years of age). Awake. Left face clonic jerks associated with minor left head deviation. Not responsive, but visually follows EEG technician. Rhythmic repetitive trunk movements associated with moaning and fear face appearance are evident in the second half of the fES. Immediately after the cessation of motor activity she obeys verbal commands and reports a "seizure". (Perinatal injury right frontocentral)
Pt. 12 (pGTCS) Semiological classification: myoclonic seizure → GTCS	1:20/0:59	(Female 35 years of age) Awake. While laughing in social context sudden onset of mimical change and symmetric jerking of both arms associated with upward eyes deviation and head retroflexion. (It) THIS is followed by symmetric facial twitching and bilateral tonic extension of both arms that further develops to bilateral symmetrical jerking. Postictal coma. (electroclinical syndrome – JME)

Abbreviations: ES – Epileptic Seizure; PNES – Psychogenic Non-Epileptic Seizure; oNES – other Non-Epileptic Seizure; fES – focal Epileptic Seizure; sGTCS – secondary Generalized Tonic Clonic Seizure; pGTCS – primary Generalized Tonic Clonic Seizure; FCD – Focal Cortical Dysplasia; HS – Hippocampal Sclerosis; JME – Juvenile Myoclonic Epilepsy.