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Myoclonic status epilepticus with dystonia-like symptoms in patients with dementia: Report of two cases

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ABSTRACT

We report cases of two elderly women with dementia who presented with a new-onset seizure disorder characterized by subtle, rhythmic muscular contractions involving the buccolingual region and the four limbs, persistent jaw opening, and abnormal cervical posture that mimicked myoclonus-dystonia syndrome and oromandibular dystonia. The symptoms lasted several minutes to a few hours. Video-polygraphic recordings revealed an electromyographic (EMG) pattern of brief, shock-like muscular contractions consistent with myoclonus that correlated with a high-amplitude (70–90 μV), 11–14 Hertz, bilaterally symmetric electroencephalographic (EEG) rhythm over the frontocentral regions. A time-locked relationship between the frontocentral EEG activity and the EMG myoclonic potentials demonstrated the cortical origin of myoclonus and therefore the epileptic nature of the disorder, whereas the oromandibular and cervical dystonic-like postures suggested the pathogenic involvement of subcortical structures. The intravenous administration of diazepam suppressed the clinical symptoms and the EEG–EMG correlate of myoclonus. The clinical and neurophysiological findings illustrate a form of myoclonic status epilepticus (SE) with dystonia-like symptoms resulting from the functional involvement of cortical and subcortical structures. The manifestation of subtle, rhythmic myoclonus and dystonic-like postures in patients with atypical EEG patterns of SE may require challenging differential diagnoses with myoclonus-dystonia syndrome and oromandibular dystonia.

1. Introduction

Electroencephalographic (EEG) epileptiform discharges represent the neurophysiologic hallmark of status epilepticus (SE) [1]. Rhythmic nonepileptiform EEG abnormalities may also be observed in this condition, particularly with increasing duration of seizure activity. No ictal EEG pattern is currently regarded as specific to any type of SE [1]. While a myoclonic form of SE is reported in patients with Alzheimer-type dementia, dystonia-like symptoms are not explicitly included in the current classification of SE [1]. Myoclonus of cortical origin may be associated with evident EEG epileptic abnormalities [2] or subtle EEG events that can be demonstrated with back-averaging techniques [3,4]. Myoclonus may also originate in subcortical structures in association with dystonia (dystonic myoclonus, myoclonus–dystonia syndrome) [5,6] as the result of genetic defects [6] or acquired aetiologic factors [7]. Oromandibular dystonia is a focal dystonia characterized by sustained or intermittent involuntary, repetitive, patterned muscle contractions affecting the lower part of the face that may result in abnormal oromandibular postures [8,9]. This condition may occasionally include cervical dystonia and dystonic tremor of the upper limbs [9], with the potential for a challenging diagnosis. Time-locked EEG correlates are not reported in patients with oromandibular dystonia or myoclonus-dystonia syndrome [3,5,7,10,11]. We report cases of two elderly women with dementia who developed a form of SE with similar neurophysiological characteristics and clinical symptoms that mimicked myoclonus-dystonia syndrome and oromandibular dystonia.

2. Case reports

2.1. Patient 1

An 81-year-old woman with severe dementia presented to our hospital with continuous, rhythmic buccolingual muscular contractions associated with forced opening of the mouth that could not be corrected

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with passive mandibular movements. These symptoms lasted for a duration of approximately several minutes to a few hours at home and recurred in the hospital daily. The patient remained vigilant but unresponsive, while an occasional feeble guttural sound, a slight tongue protrusion, and a cervical dystonic posture were observed. Her previous clinical history included diabetes, HCV chronic hepatitis and a convulsive episode attributed to the use of metoclopramide two years earlier. The patient had not been thoroughly evaluated for dementia. A cortical degenerative aetiology was presumed on the basis of brain computed tomography (CT) findings. The patient's cognitive functions were severely impaired at the time of hospitalization, and her speech was limited to short phrases. Video-polygraphic recordings performed during buccolingual muscular contractions revealed the simultaneous occurrence of rhythmic electromyographic (EMG) activity in the orbicularis oris and a bilaterally symmetric, 11–14 Hertz, 70 μV, EEG rhythm over the frontocentral regions (Fig. 1). The frequency of these activities always corresponded and did not change significantly, nor did the morphology of the frontocentral EEG rhythm (generally arciform and notched), which never showed spatiotemporal evolution. The duration of the EMG signals was approximately 40 ms, which was consistent with brief, shock-like myoclonic contractions. The EEG-EMG polygraphic analysis demonstrated a constant interval of 17–20 ms between the peak of an EEG positive polarity over the Cz electrode and the onset of the EMG potentials, indicating the cortical origin of the buccal myoclonus (Fig. 2). Spectral analysis and topographic EEG mapping revealed the maximum absolute power of the frontocentral rhythm over the Cz electrode. Jerk-locked back-averaging was not performed. Intravenous administration of diazepam (10 mg) suppressed the buccolingual muscular contractions and EEG-EMG correlates (Fig. 3, 4) but induced sedation, with the polygraphic pattern subtly recurring during a phase of decreasing pharmacologic effects (Fig. 5). Even without pharmacologic sedation, the frontocentral EEG rhythm was never observed when buccolingual myoclonus was absent. The myoclonic movements appeared to occur spontaneously, with no sensitivity to somatosensory stimuli or action. Routine blood tests revealed abnormalities in liver function, including mild, transient hyperammonaemia (61 ng/dl; normal range 0–45), increased aspartate aminotransferase (45 U/l; normal range 15–37), hypoalbuminaemia (2.7 g/dl; normal range 3.4–5.0) and reduced platelet count ($38x10^3$/μl). A brain CT scan confirmed severe diffuse cortical atrophy. Despite the liver dysfunction, oral treatment with clonazepam 0.5 mg b.i.d. was prescribed with prompt control of the buccal myoclonus, but dosages had to be reduced because of excessive sedation, with consequent sporadic recurrence of the symptoms. The patient was lost to follow-up after a few weeks. Her death was reported a few months after her discharge from the hospital.

2.2. Patient 2

An 82-year-old bed-bound woman with dementia and iatrogenic parkinsonism was hospitalized for episodes of continuous, rhythmic movements of the four limbs, persistent jaw opening, and cervical dystonic posture. The disorder usually receded at home with the oral administration of lorazepam. The patient's relatives referred to these episodes as "seizures", as she was speechless and apparently unaware of the symptoms. Her previous clinical history included chronic psychosis and a mood disorder, which were ultimately treated with quetiapine 25 mg b.i.d. and lamotrigine 50 mg b.i.d. These drugs were discontinued soon after admission to the hospital without evident clinical consequences. The discontinuation of these drugs was initially decided, as their effects were considered potentially worsening the movement disorder. The patient's neurologic status was characterized by severe cognitive impairment, tetraparesis, a fixed dystonic posture of the right hand, speech deficit, hypomimia, muscular hypertonia, and increased tendon reflexes in the four limbs. Video recordings of the disorder revealed continuous, tremor-like movements in the four limbs and persistent jaw opening, whereas EEG-EMG polygraphy documented the simultaneous occurrence of rhythmic, shock-like muscular contractions consistent with myoclonus in the right wrist flexor and a bilaterally symmetric, 12–14 Hertz, 90 μV, EEG rhythm over the frontocentral regions (Fig. 6). Theta rhythm (5.5–6 Hertz) predominated in the posterior cerebral regions, indicating a moderate, nonspecific abnormality of the EEG background activity. The EMG signals had a duration of 20–30 ms and a frequency similar to that of the EEG frontocentral activity. The polygraphic analysis revealed a constant interval of 21–25 ms between the peak of positive EEG polarity over the C3 electrode and the onset of EMG potentials, indicating the cortical origin of myoclonus. Jerk-locked back-averaging was not performed. The EEG–EMG correlate of myoclonus and the abnormal mandibular posture could be suppressed with intravenous administration of diazepam (Fig. 7, 8), which also induced sedation. The patient reawakened after three hours without showing abnormal muscular contractions, while the polygraphic recording demonstrated the regression of the EEG–EMG correlate of myoclonus. Action-induced and stimulus-sensitive myoclonic movements were not observed. Routine blood tests revealed a moderate increase in urea nitrogen (41 mg/dl; normal range, 7–18 mg/dl) and creatinine (1.4 mg/dl; normal range, 0.6–1.3 mg/dl). A brain CT scan revealed diffuse cortical atrophy. The patient responded to the oral administration of clonazepam, which was gradually titrated in the hospital to 2 mg t.i.d. and then

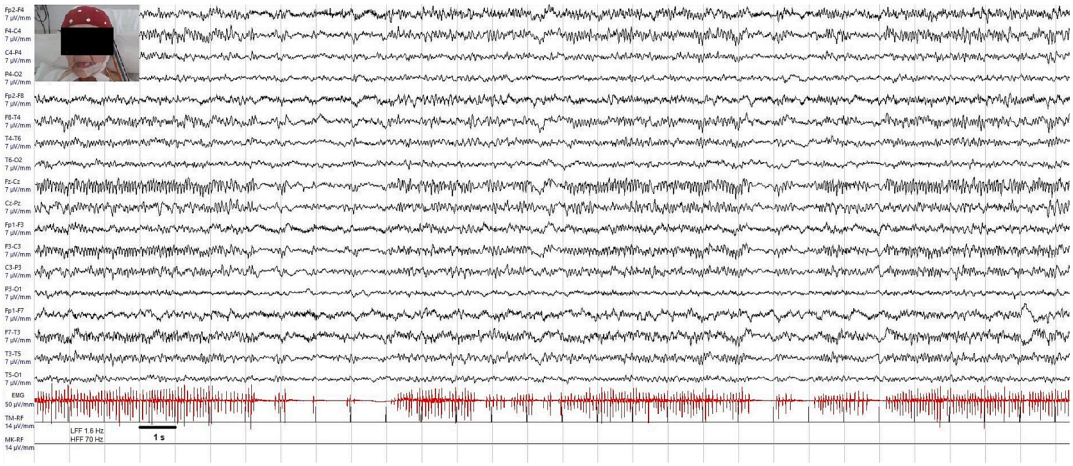

Fig. 1. (Patient 1) Video-EEG–EMG polygraphic recording of myoclonic potentials in the orbicularis oris and rhythmic EEG activity over the bilateral frontocentral regions. Note the similar rhythmicity and frequency of the EEG and EMG activities. The still frame from the video shows the concomitant jaw opening and slight tongue protrusion.

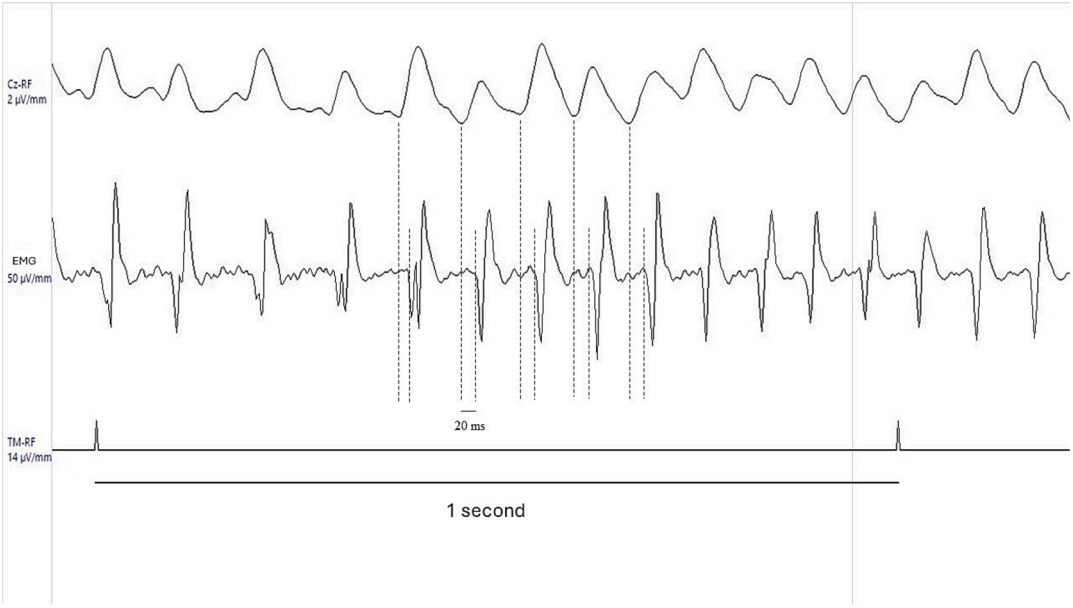

Fig. 2. (Patient 1) Time-locked EEG–EMG correlate of buccal myoclonus. The interval of 17–20 ms between the peak of positive EEG polarity over the Cz electrode and the onset of EMG potentials is consistent with the cortico-muscular conduction time, indicating the cortical origin of myoclonus.

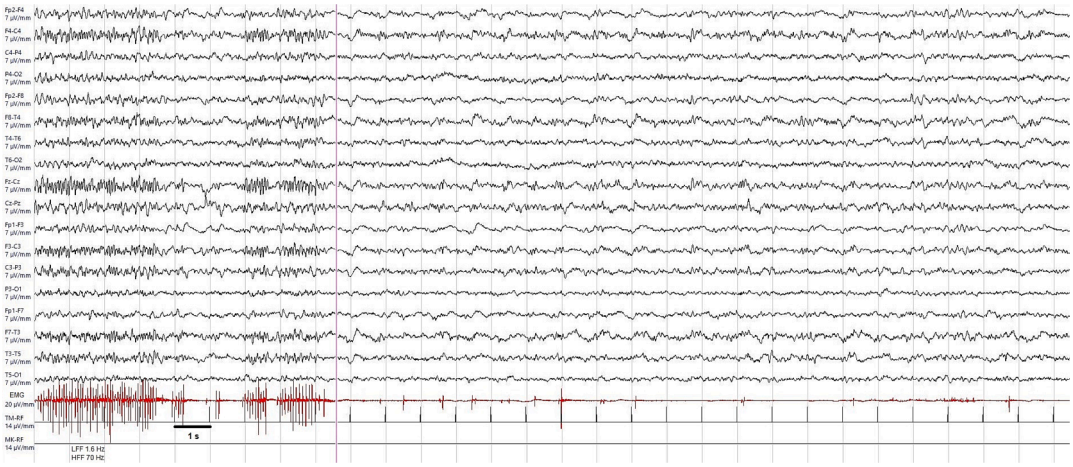

Fig. 3. (Patient 1) EEG-EMG polygraphy: the cursor marks the ending phase of the EEG-EMG correlate of myoclonus after administration of diazepam.

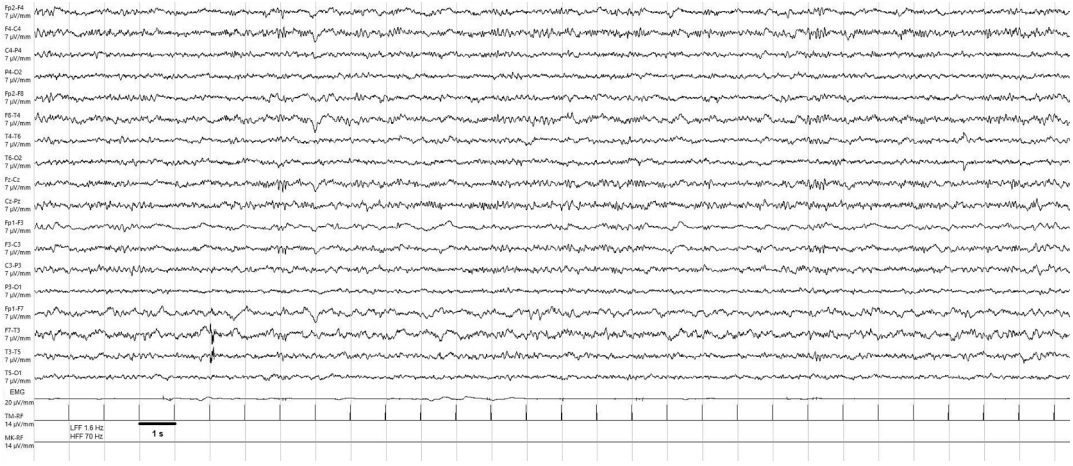

Fig. 4. (Patient 1) Disappearance of the EEG-EMG pattern of myoclonus after administration of diazepam.

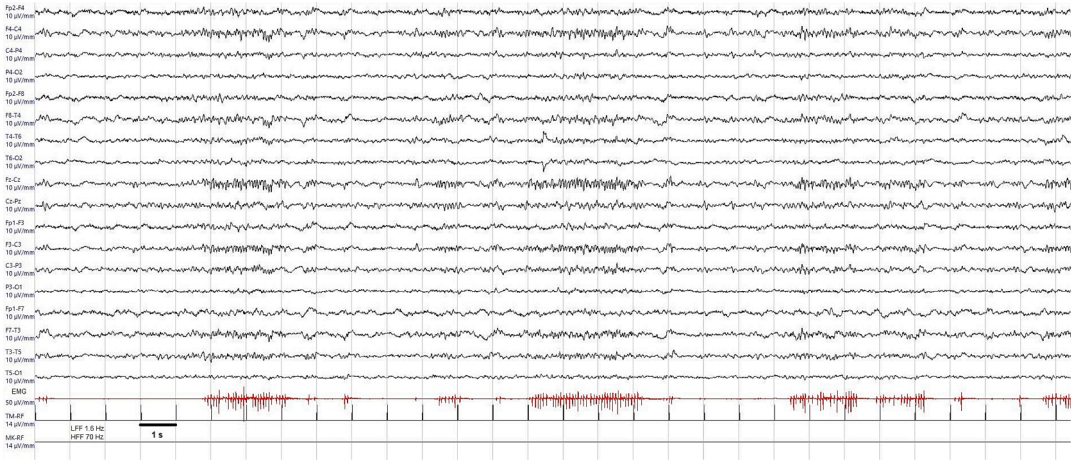

Fig. 5. (Patient 1) EEG-EMG polygraphy: recurrence of subtle EMG myoclonic potentials with EEG correlate during a phase of decreasing pharmacologic sedation.

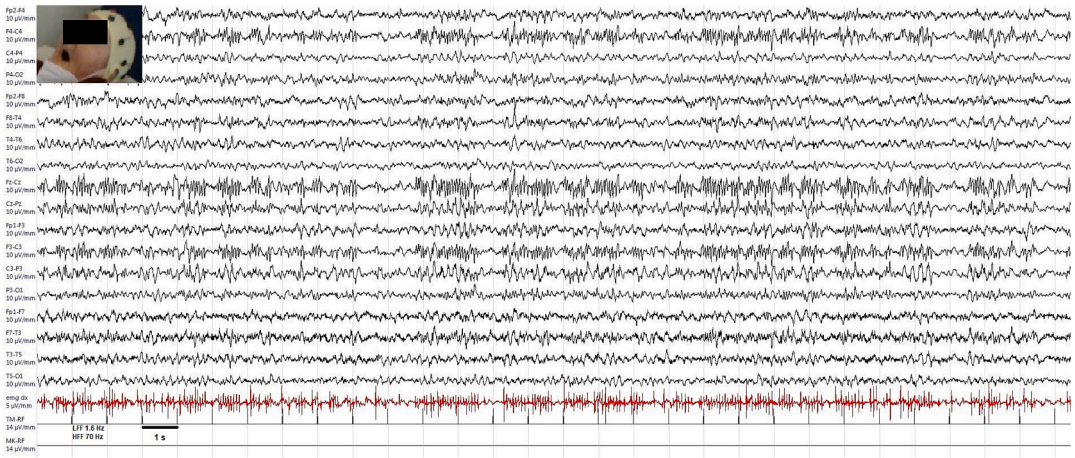

Fig. 6. (Patient 2) Video-EEG–EMG polygraphic recording of myoclonic potentials in the right wrist flexor temporally related to rhythmic EEG activity over the bilateral frontocentral regions. Note the similarity with the polygraphic pattern shown in Fig. 1. The still frame from the video shows the patient's abnormal opening of the mouth.

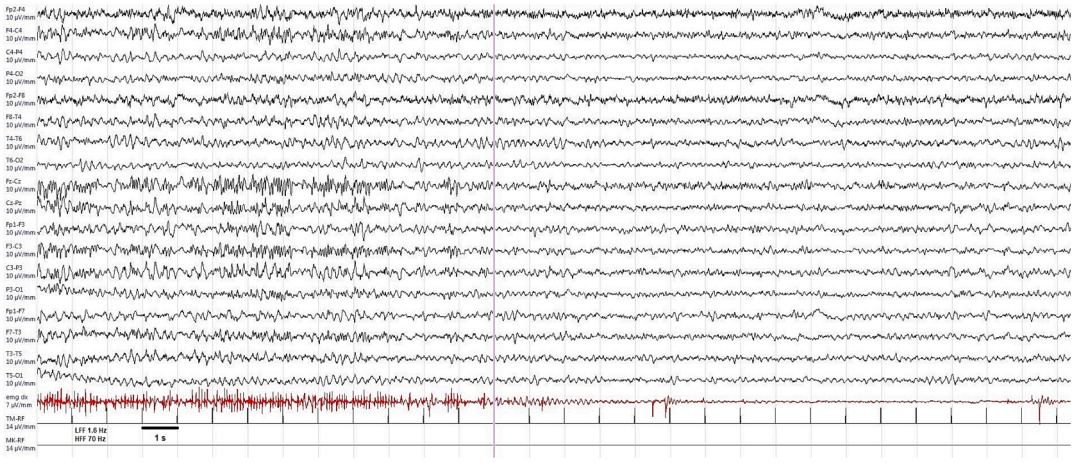

Fig. 7. (Patient 2) EEG-EMG polygraphy after intravenous diazepam: note the abrupt cessation of the EEG-EMG pattern.

reduced to 1 mg t.i.d. because of excessive sedation. An adjunctive oral treatment with levetiracetam 500 mg t.i.d. had incomplete beneficial effects after sporadic recurrence of the symptoms. The patient died for unknown reasons one year later.

3. Discussion

The patients presented with an episodic, complex movement disorder characterized by subtle, rhythmic muscular contractions involving

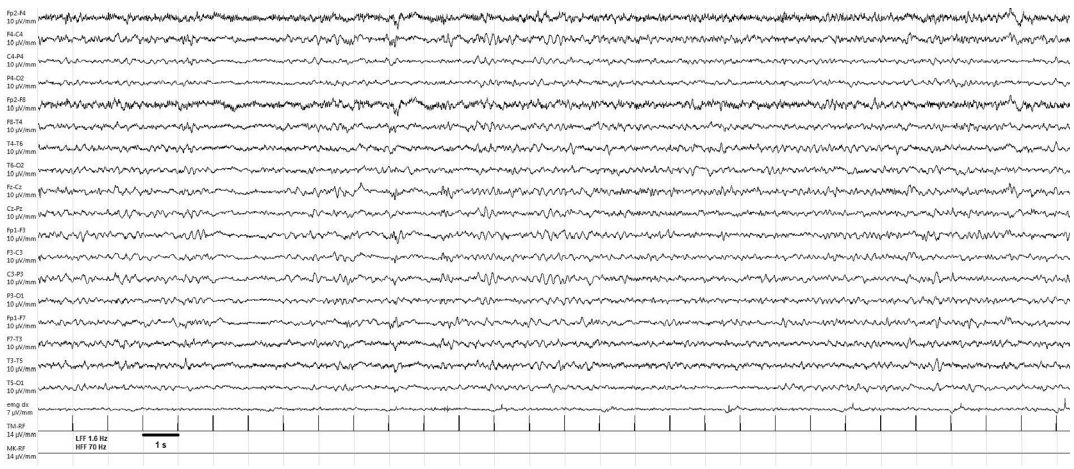

Fig. 8. (Patient 2) Disappearance of the EEG-EMG correlate of myoclonus after intravenous administration of diazepam.

the buccolingual region and the four limbs, persistent jaw opening, and abnormal cervical posture that required differential diagnoses of epileptic and nonepileptic disorders. SE could be considered on a clinical basis because of the abrupt onset and long duration of rhythmic muscular contractions, which typically ceased with intravenous administration of diazepam, despite the oromandibular and cervical dystonia-like symptoms that could be attributed to a nonepileptic movement disorder. Although the rhythmicity and low amplitude of movements resembled tremor, the shock-like muscular contractions and brief duration (20–40 ms) of the EMG signals could be more consistently interpreted as myoclonic movements [3,4]. While these subtle rhythmic movements, along with dystonic-like postures, mimicked a form of myoclonus-dystonia, EEG–EMG polygraphy demonstrated a correlation between muscular contractions and the bilateral frontocentral EEG rhythm, indicating epileptic pathophysiology. In fact, the rhythmicity and frequency of the EMG potentials corresponded exactly to those of the frontocentral EEG activity (Fig. 1, 6), which was observed only in conjunction with the myoclonic movements. Moreover, the constant interval (17–20 ms in Patient 1 and 21–25 ms in Patient 2) between the peak of positive EEG polarity (recorded over the Cz electrode in Patient 1 and C3 in Patient 2) and the onset of EMG potentials was consistent with the cortico-muscular conduction time and thus with the cortical origin of the myoclonic contractions [12,13]. These polygraphic findings clearly supported the diagnosis of cortical myoclonus, even if jerk-locked back-averaging was not performed. Myoclonus occurred with spontaneous, rhythmic muscular contractions (positive myoclonus), always in association with dystonia-like symptoms, indicating a prolonged seizure disorder with complex motor symptomatology. The bilateral synchronicity and symmetric distribution of the frontocentral EEG correlate, in addition to the dystonia-like symptoms, suggested the functional involvement of the basal ganglia and therefore a cortical–subcortical pathogenic mechanism for the overall symptomatology. In this regard, we believe that the acknowledged functional role of the thalamus in the synchronization of EEG activities [14,15] could explain the neurophysiologic pattern in our patients, as previously reported in cortical–subcortical epileptic myoclonus [16]. Notably, the EEG correlate of myoclonus could be easily identified by inspecting the tracings, even though the EEG findings were not characterized by classic epileptiform discharges. Indeed, myoclonus of cortical and cortical-subcortical epileptic origin is frequently associated with evident EEG abnormalities, such as spikes and waves or polyspikes and wave complexes, which typically show a time-locked relationship with muscular contractions [3,12]. These typical abnormalities were not observed in our patients, whereas myoclonus-related EEG activity presented a nonspecific morphology (generally arciform and notched in patient 1, spiky in patient 2) that remained unchanged throughout the manifestation of the disorder. Despite the nonspecific EEG morphology, a conclusive diagnosis of SE could be made consistently with evidence of a time-locked electroclinical pattern [17], which was typically interrupted with intravenous administration of diazepam. In fact, the EEG and clinical improvement after intravenous antiseizure medications is reported as one of the diagnostic criteria for nonconvulsive SE [18]. The neurophysiologic findings indicated massive pathogenic involvement of the motor cortices, as the evident EEG correlate of myoclonus was maximally distributed over the bilateral frontocentral regions. Notably, a similar ictal frontocentral EEG pattern has been recently reported in a patient with reflex seizures mimicking paroxysmal dystonic movements of the upper limb [19]. This previous report and the present findings indicate that paroxysmal dystonia-like symptoms may represent ictal epileptic phenomena correlated with a pattern of rhythmic EEG activity in the alpha and beta range over the frontocentral regions. Considering the potential aetiology of SE in our patients, the late onset of symptoms in the context of severe dementia, variably associated with diabetes, chronic hepatitis, or renal dysfunction, suggests a pathogenic role of neurodegenerative conditions and various metabolic abnormalities, the latter possibly acting as precipitating factors. A limitation of our case studies is that the patients' awareness could not be adequately assessed during the course of SE, because of the severe cognitive impairment. In addition, dystonia-like symptoms were evaluated only on clinical grounds, as the EMG was not concurrently recorded in any agonist–antagonist pair of muscles. Similar considerations could be made about the EMG diagnosis of myoclonus, which resulted from recordings of single muscle activities, even though EMG myoclonic potentials limited to individual muscles are described [4]. Regrettably, no information about the therapeutic effect of antiseizure medications other than clonazepam and levetiracetam could be obtained because of insufficient compliance and short follow-up. Considering the differential diagnosis of the overall symptomatology, while some clinical features resembled myoclonus-dystonia syndrome and oromandibular dystonia (the latter particularly in Patient 1) [8,9], the EEG-EMG polygraphic pattern reasonably excluded these conditions, as time-locked EEG correlates with evident neurophysiologic involvement of the motor cortices are not reported in these movement disorders [3,5,7,10,11]. Despite the limitations of our case studies and the confusing dystonia-like symptoms, the electroclinical pattern including subtle rhythmic myoclonus, oromandibular and cervical dystonic-like postures and apparent impairment of awareness in our patients could be reasonably attributed to a nonconvulsive form of SE, specifically to a myoclonic absence status [1].

4. Conclusions

The manifestation of subtle, rhythmic myoclonus and oromandibular and cervical dystonia-like symptoms in patients with atypical EEG patterns of SE may require challenging differential diagnoses with myoclonus-dystonia syndrome and oromandibular dystonia. EEG-EMG polygraphy may provide key diagnostic findings in these patients, demonstrating the epileptic nature of the disorder. The remarkable similarities in the EEG findings and clinical symptoms in our patients illustrate a distinctive electroclinical pattern of SE, resulting from the functional involvement of cortical and subcortical structures. Further reports are needed to confirm the specificity of these electroclinical features of SE and determine its aetiologic factors, pharmacosensitivity, and prognosis.

Ethical statement

The authors state that the study described in the paper complies with the publishing ethics of the journal.

CRediT authorship contribution statement

Rosario V. Rossi: Writing – review & editing, Writing – original draft, Validation, Methodology, Investigation, Formal analysis, Data curation, Conceptualization. **Rosanna Melis:** Visualization, Supervision, Resources, Data curation. **Noemi Murdeu:** Supervision, Resources, Investigation. **Sara Lizzos:** Visualization, Resources. **Maria Luigia Piras:** Visualization, Formal analysis. **Loretta Racis:** Visualization, Formal analysis. **Silvia Serusi:** Software, Resources, Formal analysis. **Maria Valeria Saddi:** Validation, Supervision, Formal analysis, Data curation.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

References

[1] Trinka E, Cock H, Hesdorffer D, Rossetti AO, Scheffer IE, Shinnar S, et al. A definition and classification of status epilepticus – Report of the ILAE Task Force on Classification of Status Epilepticus. Epilepsia 2015;56(10):1515–23. https://doi.org/10.1111/epi.13121.

[2] Riva A, D'Onofrio G, Ferlazzo E, Pascarella A, Pasini E, Franceschetti S, et al. Myoclonus: Differential diagnosis and current management. Epilepsia Open 2024;9(2):486–500. https://doi.org/10.1002/epi4.12917.

[3] Shibasaki H, Hallett M. Electrophysiologic studies of myoclonus. Muscle Nerve 2005;31:157–74. https://doi.org/10.1002/mus.20234.

[4] Wilkins DE, Hallett A, Berardelli A, Walshe T, Alvarez N. Physiologic analysis of the myoclonus of Alzheimer's disease. Neurology 1984;34:898–903. https://doi.org/10.1212/wnl.34.7.898.

[5] Obeso JA, Rothwell JC, Lang AE, Marsden CD. Myoclonic dystonia. Neurology 1983;33(7):825–30. https://doi.org/10.1212/wnl.33.7.825.

[6] Van der Veen S, Zutt R, Klein C, Marras C, Berkovic SF, Caviness JN, et al. Nomenclature of genetically determined myoclonus syndromes: recommendations of the international parkinson and movement disorder society task force. Mov Disord 2019;34(11):1602–13. https://doi.org/10.1002/mds.27828.

[7] Vingerhoets F, Russmann H, Carruzzo A, Combremont P, Ghika J. Mouvements anormaux (dystonie, athétose, chorée, ballisme). Neurologie, EMC (Elsevier SAS, Paris) 2003, 17-007-B-10.

[8] Sankhla C, Lai EC, Jankovic J. Peripherally induced oromandibular dystonia. J Neurol Neurosurg Psychiatry 1998;65:722–8. https://doi.org/10.1136/jnnp.65.5.722.

[9] Saraf U, Chandarana M, Divya MK, Krishnan S. Oromandibular Dystonia – A Systematic Review. Ann Indian Acad Neurol 2022;25(1):26–34. https://doi.org/10.4103/aian.aian_242_21.

[10] Li JY, Cunic DI, Paradiso G, Gunraj C, Pal PK, Lang AE, et al. Electrophysiological features of myoclonus-dystonia. Mov Disord 2008;23(14):2055–61. https://doi.org/10.1002/mds.22273.

[11] Marelli C, Canafoglia L, Zibordi F, Ciano C, Visani E, Zorzi G, et al. A neurophysiological study of myoclonus in patients with DYT11 myoclonus-dystonia syndrome. Mov Disord 2008;23(14):2041–8. https://doi.org/10.1002/mds.22256.

[12] Van der Veen S, Caviness JN, Dreissen YEM, Ganos C, Ibrahim A, Koelman JHTM, et al. Myoclonus and other jerky movement disorders. Clin Neurophysiol Pract 2022;7:285–316. https://doi.org/10.1016/j.cnp.2022.09.003.

[13] Donaldson IM, Marsden CD, Schneider SA, Kailash PB. Focal myoclonus, in: Marsden's Book of Movement Disorders. New York: Oxford University Press Inc.; 2012. p. 963.

[14] Amzica F, Lopes da Silva FH. Cellular substrates of brain rhythms, in: Schomer DL, Lopes da Silva FH, editors. Niedermeyer's Electroencephalography; basic principles, clinical applications and related fields, sixth edition. Philadelphia: Wolters Kluwer / Lippincott Williams & Wilkins; 2011, pp. 33-58.

[15] Hirsch E, Maton B, Kurtz D. Bases neurophysiologiques de l'électroencéphalographie clinique et principales indications. Neurologie, EMC (Elsevier SAS, Paris) 1995:17–031-A-10.

[16] Donaldson IM, Marsden CD, Schneider SA, Myoclonus KPB. Marsden's Book of Movement Disorders. New York: Oxford University Press Inc.; 2012. p. 941.

[17] Hirsch LJ, Fong MWK, Leitinger M, LaRoche SM, Beniczky S, Abend NS, et al. American Clinical Neurophysiology Society's Standardized Critical Care EEG Terminology: 2021 Version. J Clin Neurophysiol 2021;38(1):1–29. https://doi.org/10.1097/wnp.0000000000000806.

[18] Beniczky S, Hirsch LJ, Kaplan PW, Pressler R, Bauer G, Aurlien H, et al. Unified EEG terminology and criteria for nonconvulsive status epilepticus. Epilepsia 2013;54(Suppl. 6):28–9. https://doi.org/10.1111/epi.12270.

[19] Stabile A, Franceschetti S, Deleo F, Di Giacomo R, Didato G, Pastori C, et al. Teaching video neuroimage: reflex seizures mimicking paroxysmal dystonic movements in a patient with late-onset Rasmussen encephalitis. Neurology 2023;101:e1106–7. https://doi.org/10.1212/wnl.0000000000207412.