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Psychogenic axial myoclonus: Clinical features and long-term outcome

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ABSTRACT

Background: It has been increasingly recognized that the majority of patients with a diagnosis of idiopathic propriospinal myoclonus have either a subsequent clinical course or electrophysiological features indicating that the likely etiology is psychogenic. However, the clinical features of psychogenic axial myoclonus and the long-term outcome have not yet well characterized.
Patients and methods: Here we describe clinical findings with representative videos and long term outcomes of 76 patients with an electrophysiologically established diagnosis of psychogenic axial myoclonus.
Results: Thirty-seven patients were male. Mean age at onset of symptoms was 40.1 ± 15.1 years. Thirty-two patients (42.1%) presented with isolated axial myoclonus, while 44 patients (57.9%) presented additional features, including involvement of face or limb. In all patients but six (7.9%), the axial myoclonus was in flexion. In more than one-third of patients (42.1%), jerks were multifocal, meaning that there was no clear stereotyped pattern of jerks. Comparison between groups stratified according to the clinical outcome, revealed "delay of diagnosis" as the only predictor of worse outcome.
Discussion: We describe here the clinical features and long-term outcome on the largest series of patients with psychogenic axial myoclonus reported in the literature. The description of our series highlights a number of clinical features, which may help neurologists to reach a correct diagnosis on clinical grounds alone. Delay in diagnosis of a psychogenic disorder has a negative effect on long-term outcome.

1. Introduction

Myoclonic jerks thought to originate from the spinal cord may be subdivided into two broad types: spinal segmental myoclonus and propriospinal myoclonus (PSM) [1]. While spinal segmental myoclonus is often reported to be secondary to a spinal lesion, most reported cases of PSM are classed as idiopathic without identifiable spinal pathology [1]. Furthermore, it has been increasingly recognized that a number of patients with a diagnosis of idiopathic PSM have either a subsequent clinical course or electrophysiological features indicating that the likely etiology is psychogenic (functional) [2]. In fact, there is considerable uncertainty about the possibility of reaching a firm diagnosis of idiopathic PSM on a clinical basis alone. We have recently reported that all the patients referred over a 9-year period to our center with a clinical diagnosis of idiopathic PSM turned out to have a psychogenic disorder, based on an incongruent EMG pattern for PSM and/or the presence of a Bereitschaftspotential (BP, from German, "readiness potential", also called the premovement potential) prior the onset of the jerks [3]. Similar data have been reported from another centre where of 35 patients presumed to be affected with idiopathic PSM, 24 patients were diagnosed with psychogenic myoclonus on electrophysiological grounds and a further 10 patients were classed as psychogenic on clinical observation [4]. However, it is important to acknowledge that polymyography and BP recording are not widely available. Therefore, there is the need to accurately define the clinical entity of psychogenic (functional) axial myoclonus (PAM). This is especially important for as discussed above, PAM may be much more prevalent than organic idiopathic PSM and misdiagnosis of the two is a common problem [2–4]. In addition, despite the recognition of PAM, little is known about its clinical

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features (for instance, whether specific clinical clues are indicative of PAM) and little is known of its long-term outcome.

For the purpose of this paper, we have used the term PAM to describe those patients initially presumed to be affected with idiopathic PSM [5], whose axial jerks turned out to be psychogenic. While in our previous work we have only focused on the electrophysiological features, here we wish to highlight the clinical features of PAM. Moreover, we report here the long-term outcome of the 65 patients previously described by the authors [3], and we include 11 additional cases, seen in our center between May 2012 and February 2013.

2. Patients and methods

The study was approved by the University College London Hospitals, and written consent forms obtained by the patients (also concerning the publication of their video-recording). Inclusion and exclusion criteria have been extensively described elsewhere [3]. Briefly, the cohort described here includes patients initially referred to us with a diagnosis of idiopathic PSM between 2003 and 2013. They have been first re-assessed by a movement disorder expert, and the diagnosis revised to PAM in a number of them (approximately 50%). Finally, all patients underwent a multichannel video-EEG–EMG, as previously detailed [3]. On the basis of the electrophysiological findings, all patients had a diagnosed revised to PAM [3]. Beyond the electrophysiological recording, all patients had a MRI of the spine, excluding any spinal pathology. Moreover, in a number of them (66 out of 76 patients, 86.8%) a psychiatric assessment was performed within 6 months from the first clinical assessment at our center, using the Structured Clinical Interview for DSM-IV Axis I Disorders (SCID-I) [6]. At the end of this diagnostic work-up (Fig. 1), the diagnosis of PAM was communicated to all patients.

We retrospectively collected details on the following baseline parameters: age, age at disease onset and disease duration, precipitating factors at onset, prior treatments, concomitant conditions, alleviating/exacerbating factors, clinical examination, and psychiatric assessment, where available.

All patients were regularly followed-up and had at least 6 months of follow-up (range: 6–72 months) at the time of this study. Reviewing medical charts with regard of the follow-up period collected the following parameters: presence of fluctuations, treatments, and outcome. The assessment of outcome was based on clinician assessment at last visit.

To address whether these patients could be different in basic demographic and clinical parameters to those with other psychogenic movement disorders (PMDs), twenty consecutive patients with clinically established functional limb dystonia were selected as control group. Such control group was not meant to compare the clinical outcome, since we believe it can be strongly influenced by the specific phenotype [7]. Data are shown as mean ± standard deviation or as percentage. Comparisons between groups were done using *t*-test or chi2 test, as appropriate. The Kruskal Wallis test was used for more than 2 samples (see text) and $P < 0.05$ was considered statistically significant. Statistics were performed using STATA software, version 11.0 (StataCorp LP, USA).

3. Results

Demographic and clinical data of the patients included in this study are listed in Table 1. For all of the patients, the diagnosis of PAM was established on the basis of electrophysiological testing: the presence of a BP associated with the jerks or EMG findings not consistent for PSM (i.e., inconstant pattern of muscle activation, conduction velocity determined from the inter-burst interval between two different muscles >15 m/s, and EMG burst duration > 1000 ms), as previously reported [3].

Of the 76 patients with PAM, 37 were male, while in the functional dystonia group only 2 out of 20 were male (48.7% vs 10%, $p < 0.01$). Age at onset was higher in the PAM group compared to the functional dystonia group (40.1 ± 15.1 years vs 27.9 ± 2.7 years, respectively, $p < 0.01$). A precipitating event (most commonly a minor surgical procedure) was identifiable prior to the onset of myoclonus in a 36.8% of patients. Approximately 50% of patients reported other unexplained medical symptoms (mainly gastrointestinal, fatigue and

Table 1
Demographic and clinical characteristics of our patients (ys: years; AM: axial myoclonus; *: after a single injection of botulinum toxin).

	Psychogenic AM ($n = 76$)	Psychogenic dystonia ($n = 20$)	*p* Value
Gender	37 M; 39 F	2 M; 18 F	0.001
Age at onset	40.1 ± 15.1 (range 16–80 ys)	27.9 ± 2.7 (range: 22–31 ys)	0.0006
Disease duration	5.9 ± 5.7 (range 1–32 ys)	5.3 ± 5.5 (range 1–13 ys)	ns
Delay in diagnosis	3.7 ± 5.5 (range 0.5–30 ys)	0.5 ± 0.7 (range: 0.5–2 ys)	0.009
Precipitant	36.8%	41.3%	ns
Other somatizations	51.3%	39.7%	ns
Psychiatric co-morbidity	28.8% (19 out of 66 for which psychiatric assessment was available)	–	–
Number of medications	2.6 ± 1.6 (range: 1–8)	–	–
Alleviating/exacerbating factors	30.3%	–	–
Clinical fluctuations	30.3%	–	–
Last follow-up	2.2 ± 1.4 (range 0.5–6 ys)	–	–
Clinical outcome:		–	–
Recovery*	22.4%		
Better	15.8%		
Stable (without medications)	38.1%		
Stable (with medications)	6.6%		
Worse	17.1%		

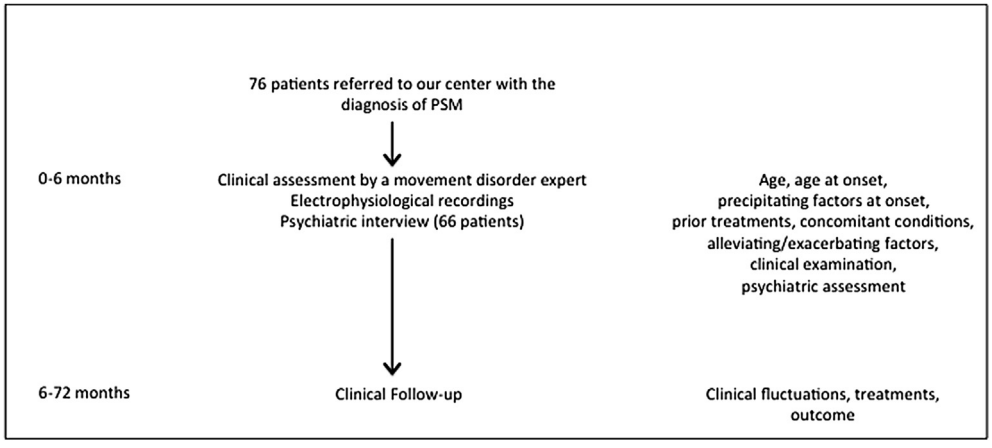

Fig. 1. Overview of the clinical work-up and data collection.