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Case Report

Lateralized, nonepileptic convulsions in an adult with cerebral palsy: Case report and review of the literature☆



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ABSTRACT

The authors report a case of unilateral functional neurological symptoms (nonepileptic convulsions) in a 38-year-old man with mild, motor-predominant cerebral palsy. His convulsions are all lateralized to the same side as his paretic limbs. His episodes significantly decreased after several months of weekly psychodynamic-oriented psychotherapy. Functional neurological disorders have been rarely reported in children or adults with cerebral palsy. Among patients with brain injury, right-hemispheric brain disease may be more helpful than either handedness or the side of symptoms in clinically profiling patients with suspected functional disorders. This case raises biomechanistic questions about brain injury, the development of functional disorders, and the lateralization of functional symptoms.

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1. Introduction

Here, we report a case of functional neurological symptoms (nonepileptic convulsions) in a 38-year-old man with mild, motor-predominant cerebral palsy (CP). His episodic, hypermotor convulsions are almost all right-sided, occurring in the same limbs affected by his CP.

Nonepileptic convulsions, often named “psychogenic nonepileptic seizures” (PNES) in current literature, have long been conceptualized as a form of “conversion disorder” [1]. Like similar “conversion” symptoms of paralysis, hypoesthesia, or blindness, PNES remain not fully explained by current models of neurologic or medical disease. A newer term called “functional neurological symptom disorder” (FNSD), intended to subsume many functional disorders under one umbrella, has replaced “conversion disorder” in the DSM-5 [2]. A motivation for this name change was to expand the disorder beyond its psychoanalytic origins (i.e., “conversion hysteria”, “conversion reaction”) and place it in a framework of neural system function/dysfunction [3]. We will use the term FNSD in this paper.

To our knowledge, functional neurological symptoms have rarely been reported in patients with CP. Two independent case reports exist for children, whose ages were 9 and 12 [4,5]. One of these children also had left-hemispheric focal epilepsy, with secondarily generalized seizures. Neither child had lateralized functional symptoms. In a separate, population-based study of adults over a 5-year period at one

video-EEG monitoring center, there was one patient out of 23 with functional symptoms who also had CP [6]. That patient's functional symptoms were not described in any detail.

In general, it has been suggested that functional symptoms occur bilaterally in about 2/3 of patients and unilaterally in the other 1/3 [7]. Our patient has chronic, right-sided motor deficits from cerebral palsy and then later developed episodes of right-sided, functional convulsions in his 30s. Because his symptoms are all right-sided, our case raises biomechanistic questions about lateralization in functional disorders and the temporal sequence of neural events before and during active symptoms. This case also raises the notion that there may be a predilection for aberrant motor control of neural substrates that have been previously damaged.

Here, we will address the issue of whether the side of symptoms or the presence of preexisting brain disease helps in improving the detection of suspected, functional symptoms. We point the reader to a more extensive recent review [8] and a neurobiological-based conceptual model [9] of FNSD for more theoretical discussions. We will address the unique aspects of this case and how they might fit into current models of the disease.

2. Case report: description and methods

KJ is a high-functioning 38-year-old, left-handed man, with mild, spastic hemiparetic type of CP (SHCP). He experienced early, hypoxic brain injury on day 2 of life, secondary to bronchopulmonary dysplasia. As a child, KJ had predominantly gross motor delays, with only mild cognitive delays. Reports indicate that he had difficulty sustaining attention in school and that he was treated with methylphenidate with some benefit. His motor system deficits have always been right-sided: he has a mild right hemiparesis of the face, arm, and leg; dystonia in the right hand; and overall increased right-sided motor tone.

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Throughout adolescence and early adulthood, KJ experienced depression and anxiety, with self-described low self-esteem, social anxiety, and social ostracism. He has mild obesity and describes being teased and bullied through his school years. He also describes deficient emotional support and harsh criticism from family members during childhood and into adulthood.

KJ describes significant relationship and financial stresses prior to his first episode of right-sided convulsions. He also experienced consecutive occupational failures leading up to symptom onset. It was at age 30 that his episodes began. They were frequent and disabling, characterized by altered awareness without loss of consciousness and limb-shaking convulsions. During a typical episode, he first feels a sense of anxiety, neck aches, and “light-headedness” for up to 5 min before any movements. He then develops nonstereotyped, hypermotor, convulsive movements of his right arm, right leg, and, sometimes, head, of varying amplitudes. The episodes can last between 3 and 20 min and usually occur in the presence of other people. His most significant episodes occurred while working in retail stores, where he has to engage socially with many people for hours at a time. He feels general weakness for several hours after an episode and is able to recall most details from before and during the episode.

Several typical episodes were captured by video-EEG monitoring over 5 days at our affiliated medical center. There was no electrographic correlation with any episode. He also had several contrast-enhanced brain MRIs over 5 years, remarkable only for mild, symmetric, periventricular leukomalacia (PVL), which is typical for CP (see Supplementary images 1–2). His MRIs were notable for preserved symmetry of cortical volume, without apparent lateralized or segmental tissue loss.

Following his diagnosis of FNSD, he was referred for individual, psychodynamic-oriented psychotherapy as well as couples' psychotherapy. These interventions have led to a significant reduction in episodes, improved self-report on depression scales, and improved personal relationships.

3. Discussion

We will first review a framework for understanding the lateralization of symptoms in FNSD and then discuss how this case might inform emerging FNSD models.

3.1. Detecting FNSD

Functional neurological symptoms are often difficult to detect and differentiate from those more easily explained by neurologic disorders, like dyskinesias, seizures, or myoclonus. Because functional symptoms can be unilateral or bilateral and occur in patients both with and without MRI or EEG abnormalities, these features are not sufficient alone for diagnoses. Functional neurological symptom disorder is also well described in patients with epilepsy, multiple sclerosis, and stroke, among other disorders [10–12], which complicates the diagnosis further. There does seem to be a predilection toward bilateral rather than unilateral symptoms in FNSD [7]. When unilateral, it was long hypothesized by observation [13,14] and then multiple case series [15–18] that symptoms occur more commonly on the left side of the body than on the right side.

3.2. Left-sided symptom predominance: not enough evidence

Historically, the presumed higher frequency of left-sided symptoms was explained by a model of hemispheric dominance or “specialization” [19,20]. Since one hemisphere is “dominant” for major functions, like motor or language, the other hemisphere is “nondominant” and specialized for more “unconscious” or affective processing [16,21,22]. From this model, a link is hypothesized between functional neurological symptoms and dysfunction of this nondominant, affect-focused hemisphere. The link is driven by the strong clinical observation that

significant psychological stress often immediately precedes symptom onset. Since most people are right-handed, with affect processing in the right hemisphere, left-sided symptoms were thought to be more common because of the direct, downstream contralateral effects of non-dominant hemisphere dysfunction. Most series that have reported this left-sided symptom gradient have used patient handedness as a surrogate to predict dominant/nondominant hemispheres.

However, recent studies [7,23] have shown that both left- and right-handed patients who have unilateral FNSD symptoms have a virtually equal distribution of left- and right-sided symptoms. In a meta-analysis [23], researchers looked at 121 studies with 1139 patients, where symptom side was either incidentally recorded or mentioned in the study title. They found no left-sided FNSD predominance or any relationship between handedness and the side of symptoms. The authors cite an outcome variable reporting bias to explain the discrepancy between their results and those of prior case series. Described as a “headline bias”, the authors found data buried within many of the prior series which are contrary to the prevailing conclusion declared in the paper headline.

3.3. Structural brain disease and FNSD: from “functional overlay” to neural dysfunction

While symptom side may not be a useful clinical sign to optimize detection of FNSD, having MRI- or EEG-evidenced neurologic disease may be important. In addition, regardless of symptom side, right-hemispheric neural dysfunction may still be implicated in FNSD.

First, long-standing clinical observation and studies suggest that functional symptoms are more common in patients with structural or electrophysiological brain disease than in those without [24–26]. The term “functional overlay” was used in the past to describe the presence of “pseudoneurologic” symptoms, like PNES, in patients with known, structural brain disease [27,28]. These studies include patients with epilepsy, multiple sclerosis, stroke, and brain tumors, among other conditions.

Importantly, up to 12% of patients with MRI- or EEG-based neurologic disease have concurrent symptoms which their neurologist believe is excessive for, or “unexplained” by, their disease [26]. Specific to patients with hypermotor functional symptoms like PNES, studies have shown that between 5 and 40% of them also have concomitant epilepsy or a history of clinical seizures [29].

It has been shown in a series of 79 patients with both PNES and epilepsy that these patients are much more likely to have right hemisphere pathology on EEG or brain MRI than patients with epilepsy alone [11]. It has also been suggested that disorders of motor neglect, motor intention, and action authorship, for which right hemisphere dysfunction is often implicated, are also related to the mechanism of FNSD [9].

Numerous functional neuroimaging studies, using SPECT, fMRI, and FDG-PET, have implicated both hyper- and hypofunctioning of various brain regions during active FNSD symptoms. These include the left frontal–striatal circuits, the right frontal cortex, the thalamus (i.e., contralateral to symptom side), and the dorsolateral prefrontal cortex (DLPFC), among others [30–32] (for a thorough review, see [9]).

3.4. This case: unilateral FNSD in CP

It is intriguing that our patient's functional convulsions reliably occur on the same side as his chronically paretic limbs. Specifically, his convulsions occur in limbs for which the subserving motor system was definitely injured in early brain development. While there is certainly anatomic overlap between the output motor tracts and control regions damaged by hypoxic injury and those that subserve his current convulsions, it is difficult to describe the sequence of neural events that

link those injured regions to the neural dysfunction that has to be occurring during active, functional symptoms.

Of interest in this case is that our patient was left-handed and demonstrated intact language function throughout life. His family history of left-handedness is not known. One might hypothesize that his early left-hemispheric brain injury necessitated some degree of cortical reorganization. This process may have resulted in compensatory, right hemisphere-mediated language or motor function. This is a well-described phenomenon in children with perinatal brain injury [33]. It is interesting to consider whether the neuroplastic process of cortical reorganization, which leads to aberrant mapping of motor and probably many other functions, is a factor that predisposes these patients to develop FNSD more frequently than patients without brain injury. Larger studies are certainly needed to explore this further.

Also of interest in this case is the association of psychological conflict preceding the onset of FNSD. Why did our patient first experience FNSD at age 30 when he had experienced chronic psychological stress, including social ostracism, depression, and emotional negligence, since childhood? The association of recent, psychological stress prior to FNSD is well described and has even been required in some past iterations of disease criteria [34,35]. This association has been removed as a requisite criterion in the DSM-5 in recognition that relevant psychological factors may not be demonstrable at the time of diagnosis.

Because our patient had little psychological or psychiatric intervention prior to symptom onset, we suggest that he developed a host of maladaptive cognitive and behavioral coping skills. These can greatly increase one's vulnerability to later stressors. It may have been a confluence of stressors in our patient, including his worsening romantic relationship and a series of occupational failures, which was the psychological "straw that broke the camel's back". Future studies might explore the relationship between the time period of one's brain injury, the existence of psychological stressors, the cognitive and emotional processing style, and the development of FNSD.

4. Conclusion

In a broad sense, our case offers additional evidence that FNSD can develop in patients with structural brain injury of varying etiologies, including CP. We know that structural brain injury, by itself, puts patients with different neurologic disorders at increased risk for FNSD. More work is needed to describe the involved neural structures and the physiologic sequence of activation and deactivation that occurs during symptoms. It is important for clinicians to be aware that patient handedness and side of symptoms may not be reliable clinical features to help increase detection of FNSD.

Functional neurological symptom disorder has only rarely been reported in patients with CP. Recent epidemiologic work has demonstrated that children and adolescents with CP have a high prevalence of psychiatric disorders, up to a 3- to 4-fold increase over healthy controls in some series [36,37]. The main neuropsychiatric disorders appear to be attention-deficit disorder (ADD), oppositional defiant disorder (ODD), and anxiety spectrum disorders, including generalized anxiety and compulsiveness [37]. Our study and the previous case reports suggest that attention be paid as well to assess for the presence of comorbid FNSD. Problems with affect regulation in patients with CP may be neglected as there is often a medical and neurologic focus on their treatment, with interventions like speech therapy, spasticity treatment, and cognitive rehabilitation. Treating the psychiatric symptoms and unique psychological stressors during childhood and adolescence in these patients may significantly decrease psychiatric morbidity as they transition to adulthood [37].

Diagnosing FNSD may be especially challenging in patients in which both "explainable" and "unexplainable" symptoms are anatomically colocalized (e.g., additional weakness in an already paretic limb) or behaviorally similar (e.g., nonepileptic convulsions in a patient with

epilepsy). Early, accurate detection of FNSD is critical as it increases the chance that treatment will lead to a sustained decreased frequency or resolution of symptoms.

Supplementary data to this article can be found online at <http://dx.doi.org/10.1016/j.ebcr.2014.01.005>.

Conflict of interest

The authors have no conflict of interest to report.

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