



A case of functional dystonia: clinical cues to differentiate between organic and functional dystonia

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Dear Editor,

Dystonia is an involuntary and sometimes painful muscles contraction that causes abnormal movements and/or posture [1]. Functional dystonia belongs to the wide spectrum of functional movement disorders (FMD) that are abnormal movements not due to an organic condition, varying in intensity and pattern with distraction or suggestibility [2]. Distinguishing between organic and functional dystonia is not always easy: organic dystonia is often task specific and the complexity of the movement itself could resemble a voluntary movement; in addition, the pathophysiology of organic dystonia itself is still poorly understood [2].

Here, we report the case of a 50-year-old woman, born and grown up in Italy. Her past medical and psychiatric history was negative. In 2007, she slipped on ice while she was walking and injured her right arm. After that fall, she suddenly developed persistent pain, fatigue, and functional limitation to her right upper limb. Between 2007 and 2010, she underwent several treatments (analgesic drugs, massages, and physiotherapy) without any benefit. In December 2010, during a session of electrical muscle stimulation, she suddenly developed painful spasms of the right side of her face, mainly involving her upper platysma and her mouth with tonic lip deviation. The spasms quickly affected her ipsilateral upper and lower limb and, in few minutes, also the contralateral side of her body. There was some arching of her back, without loss of consciousness neither sphincter release. After this episode, she came back home and she started presenting spasms of all

her body daily. Spasms were triggered by several perceptual stimuli (e.g., loud noises, bright lights, fatigue, strong emotions, specific tactile stimulation, or distinct movements such as abduction of the right arm).

She was evaluated by several neurologists and she underwent the following investigations: video-electroencephalogram, brain and brain stem MRI with and without contrast dye, brain CT without contrast dye, rachicentesis, electromyography, motor evoked potential for the upper limbs, thorax X-ray, routine hematochemical examinations, wide hormone, auto-antibody, tumor specific-antibody, and tumor markers blood panel. All the examinations were within the normal range. During that period, she was also evaluated by a dentist who suggested her to use a common bruxism tooth brace. Curiously, when she had her tooth brace on, her facial spasms completely disappeared (but suddenly appeared again when she removed it).

Given the negativity of all the investigations, the inconsistency of the presented symptomatology with any neurological or organic disease and the unusual triggers of the dystonic crises, she was diagnosed with functional dystonia (according to Fahn and Williams criteria) [3] by a neurologist with an expertise in FMD. Thus, she was referred to a psychiatrist and a psychologist; she started psychotherapy (as a first-line treatment for FMD) twice a week without any significant improvement of dystonic spasms, which remained the same in terms of frequency and intensity, but with enhancement on social dimension (her social functioning, which had been seriously compromised by dystonia, started to get better).

On the mental status examination, she was alert, well-kept, and calm. The mood tone was euthymic, facial expression was normal, and behavior and attitude were appropriated. Thought process and content were normal; no abnormalities of perception were found.

Here, we presented the case of a 50-year-old lady affected by functional dystonia. This case provides the opportunity to define specific clinical characteristics of functional dystonia, in order to help clinicians in the differential diagnosis between organic and functional dystonia. First, the onset of functional

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dystonia in our patient is preceded by a physical precipitating event (fall on ice). According to the recent study by Parees et al., although historically neglected in favor of pure psychological explanation, physical precipitating events may play an important role in FMD development by providing initial sensory data, which along with psychological factors such as panic, might drive subsequent FMD [4]. Second, the patient developed dystonic crises in response to non-specific triggers such as loud noises or distinct movements such as abduction of the right arm [5]. Third, the patient showed a specific placebo response with the use of the tooth brace, which is typical of FMD [4]. Fourth, the presence of tonic lip deviation is a key distinctive feature of functional dystonia, as pointed out by Colosimo [6]. Another cue to help the differential diagnosis between organic dystonia and its functional counterpart is the presentation pattern that often shows an abrupt onset and initial fixed dystonia for the functional type [2].

Supportive features are the presence of pain (more common than in organic dystonia), the unilateral platysma involvement, the presence of laterocollis with ipsilateral shoulder elevation and contralateral shoulder depression, fixed wrist and finger flexion with relative sparing of the thumb and index fingers, and fixed foot plantar flexion and inversion. The pathophysiologic abnormalities in functional dystonia overlap substantially with those of organic dystonia, with similar impairments in cortical and spinal inhibition and somatosensory processing, but with emerging data suggesting abnormalities in regional blood flow and activation patterns on positron emission tomography and functional magnetic resonance imaging, respectively [7].

In conclusion, differentiation between organic and functional dystonia should be mainly based on the clinical history

and neurological examination, with investigations only playing an ancillary role. Recognizing the etiology of the different types of FMS is crucial for all neurologists in terms of prognosis and treatment approach.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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