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Case Report

Abnormal gait and bradykinesia in the preclinical phase of Huntington's disease – psychogenic movement disorder?

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Soltan W, Sitek E, Wichowicz H, Wieczorek D, Slawek J. Abnormal gait and bradykinesia in the preclinical phase of Huntington's disease – psychogenic movement disorder?

Objective: Psychiatric symptoms may occur in individuals at risk of Huntington's disease (HD) regardless of their genetic status. Psychopathological symptomatology is attributed to both genetic and environmental factors. In case of asymptomatic gene carriers, psychiatric symptoms may precede involuntary movements.
Methods: We report the first case with abnormal gait and bradykinesia in preclinical adult HD. A 33-year-old woman blind to her mother's HD diagnosis and her own genetic status developed motor slowing and gait disturbance. The symptoms withdrew due to counselling and antidepressant medications. Subsequently, she was informed her own and her mother's genetic testing results, but 2-year follow-up did not reveal the onset of choreic movements, cognitive deterioration or depressive symptoms in the patient. Personality assessment (Minnesota Multiphasic Personality Inventory) and neurological examination results are presented, accompanied by 2-year follow-up data. Follow-up examination included Unified Huntington's Disease Rating Scale (motor, behaviour and function), Beck Depression Inventory, Hamilton Depression Rating Scale and neuropsychological assessment (trail-making test, Stroop test, verbal fluency trials, symbol digit modalities test, digit span, serial seven subtraction, Hopkins verbal learning test and nine-hole peg test).
Conclusion: Motor abnormalities in individuals at risk of HD may be of psychogenic origin. It is a matter of debate if this psychogenic reaction presented as hypokinetic syndrome may be a result of choreic movements of her mother (hyperkinetic syndrome) and depression or if this psychogenic reaction represents the preclinical psychiatric abnormalities in an asymptomatic gene carrier preceding the onset of the disease.

Keywords: gait disorder; Huntington's disease; psychogenic movement disorder; psychogenic parkinsonism

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Introduction

Huntington's disease (HD) is a hereditary neurodegenerative disorder, presenting with a wide range of motor, cognitive and psychiatric disturbances (1). A majority of HD gene carriers display a lot of subtle abnormalities, even up to two decades before the onset of chorea (2). Despite the fact that psychiatric symptoms may precede motor abnormalities in HD, there is no difference in the lifetime frequency of clinical psychiatric disorders between gene carriers and non-carriers. The former reported only more depressive symptoms (3). Conversion disorder as a psychiatric manifestation of HD is a very rare phenomenon. Arrojo-Romero et al (4) presented the case in whom hypochondriasis (e.g. unspecified somatic complaints, weakness) preceded by 14 years the onset of gait dysfunction (wide-based gait with small steps) and choreic movements.

We present a case of abnormal gait with severe general motor slowing (slowness or bradykinesia) in a presymptomatic gene carrier unaware of her own and her mother's genetic status.

Case presentation

A 33-year-old Caucasian female, with 11 years of education, reported gait impairment of sudden onset, chronic fatigue and apathy. Medical history revealed that 6 months earlier, she recovered from tuberculosis with drug-induced liver lesion. Her family situation was difficult due to the death of her child a couple of years earlier and marriage problems related to alcoholism in her husband. Subsequently, as the walking problems were not a prominent complaint, first she was referred to a psychiatrist and to a psychological examination. Minnesota Multiphasic Personality Inventory confirmed severe adjustment disorder with depressed mood and anxiety and withdrawal as a predominant coping mechanism.

She was admitted to Psychiatry Department, diagnosed with dissociative disorder and discharged after 1 week at own request. Paroxetine therapy was introduced. One month later, as the walking problems and general slowness became severe, she was admitted to Neurology Department. Examination revealed atypical gait with small steps and stiffening knee joints while walking (but with preserved arms balance), impaired initiation of movement, bradykinesia, rather increased but fluctuating muscle tone in lower limbs (withdrawing following distraction, feigned rigidity) and hypomimia. Magnetic resonance imaging and somatosensory evoked potentials failed to identify any abnormalities.

Psychological examination revealed denial as a main defence mechanism, low insight and susceptibility to conversion reactions as well as intelligence below average (Wechsler Adult Intelligence Scale-Revised; IQ = 77). Treatment with paroxetine was maintained (20 mg/day) and buspirone (15 mg/day) was added. Following several days of treatment at the Neurology Department **a**nd explanation of non-organic aetiology of symptoms, marked improvement of gait during 2-week hospitalisation was observed, and she recovered completely within the next month.

At the time of the onset of symptoms, hospitalisation and recovery, the patient was blind to her mother's HD diagnosis and her own genetic status. Choreic movements in her mother had been already present for 3 years, without family history of chorea. Both of them were hospitalised at the Neurology Department at the same time and consented to genetic testing, which confirmed mutation in both the proband (12/42 CAG repeats) and her mother (16/41 CAG repeats).

Despite informing the patient about her own and her mother's genetic testing results, during the 2-year follow-up, no depressive symptoms were observed. She continued treatment with paroxetine (20 mg/day) and buspirone (15 mg/day), and nitrazepam was added (5 mg/day), for the next 8 months.

As evidenced by two follow-up examinations, according to the REGISTRY protocol, by European Huntington's Disease Network, no specific for HD (oculo)motor or psychiatric symptoms developed. In terms of cognitive function, which was at baseline below average but with no dementia (Mini-Mental State Examination 27; Mattis Dementia Rating Scale 128), no deterioration was noted during the observation period (Table 1). Slowed information processing was evidenced by symbol digit modalities test and Stroop test.

Discussion

The clinical presentation of parkinsonism with predominant gait disturbance in the reported case is atypical for HD in the adults. In accordance with the criteria of Williams et al (5), medical history, neurological examination, psychiatric records and remission following treatment seemed to confirm psychogenic origin of symptoms. It can be questioned whether psychogenic movement disorder can be diagnosed in an asymptomatic gene carrier. However, it seems possible as she was not aware of the genetic disorder of her mother.

Psychiatric disturbances in HD gene carriers may be caused by both genetic and environmental (e.g. disturbed upbringing) factors (6). Depression in HD gene carriers may both precede motor symptoms, constituting the first manifestation of the disease, and appear while considering predictive testing or after

Table 1. Neurological, neuropsychiatric and neuropsychological follow-up examinations results

Age (years/months)	34/1	35/1
UHDRS motor	0	0
UHDRS behaviour	0	0
UHDRS function	24	24
Mood		
Beck Depression Inventory	0	3
Hamilton Depression Rating Scale	0	3
Attention and memory		
Digit span – maximum forward/backward	4/3	3/3
Serial seven subtraction (maximum 14)	11	10
HVLT – learning curve/delayed recall/ recognition	5–6–6/6/10	6–10–11/10/11
Trail-making test A/B	53T/50T	59T/49T
Executive function		
Verbal fluency (phonological: MKP/semantic)	25/15	21/19
Stroop test – colour naming/reading/ interference	47/65/35	49/64/35
Motor speed and coordination		
Symbol digit modalities test	28	30
Nine-hole peg test – right hand/left hand	19/18	22/22

HVLT, Hopkins verbal learning test; UHDRS, Unified Huntington's Disease Rating Scale.

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obtaining positive genetic testing result (3,6). In our patient, only the first explanation may be valid. She developed abnormal gait with predominant slowing after being diagnosed with severe adjustment disorder. It is open to debate whether conversion disorder (psychogenic movement disorder) in the presented case is the psychiatric manifestation of HD, heralding the clinical onset (2,3), or is HD unrelated and can be attributable to other factors, e.g. environmental (6). In the patient, several environmental stressors may have triggered conversion: (a) undiagnosed motor symptoms in her mother, (b) returning to the role of mother, wife and housewife after a long hospitalisation due to tuberculosis, (c) alcoholism in her husband, (d) the death of her child a few years before without mourning period.

Recent data suggest the usefulness of functional imaging in the differential diagnosis of psychogenic movement disorders, factitious disorders and malingering (7). However, these methods are not routinely used in the clinical practice, and their applicability in the asymptomatic HD gene carrier seems questionable because of no standards for preclinical HD population. In our patient, none of these techniques were used. In our case, according to the criteria of Williams et al (5), the recovery without specific treatment confirmed the psychogenic origin of symptoms. However, if the recovery is not observed, functional neuroimaging may be helpful to establish the diagnosis (8,9).

Parkinsonian symptoms are typical for juvenile HD, but in our case, the age was beyond the range typical for Westphal form of HD, and the symptoms disappeared without any specific treatment.

To our knowledge, this is the first presented adult case who presented with Parkinsonian features in the preclinical phase of HD, albeit the second case in whom psychogenic symptoms have preceded HD onset (4). It remains a matter of debate if motor symptoms observed in our patient are related to HD and can be regarded as a psychogenic movement disorder or if those motor symptoms of psychogenic origin are rather environmentally related. However, this case report adds dissociative disorder to the spectrum of psychiatric diagnoses in preclinical HD, as the causal relationship cannot be excluded. Nevertheless, it seems important in the clinical practice to be aware of possible psychogenic origin of symptoms in gene carriers as those symptoms, unless recognised and treated, may persist.

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References

1. Rosenblatt A, Leroi I. Neuropsychiatry of Huntington's disease and other basal ganglia disorders. Psychosomatics 2000;**41**:24–30.
2. Paulsen JS, Langbehn DR, Stout JC et al. Detection of Huntington's disease decades before diagnosis: the Predict-HD study. J Neurol Neurosurg Psychiatry 2008;**79**:874–880.
3. Julien CL, Thompson JC, Wild S et al. Psychiatric disorders in preclinical Huntington's disease. J Neurol Neurosurg Psychiatry 2007;**78**:939–943.
4. Arrojo-Romero M, Silva AF, Palha AP. Hipocondria and Huntington's disease [Hypochondriasis and Huntington's disease]. Actas Esp Psiquiatr 2006;**34**:65–66.
5. Williams DT, Ford B, Fahn S. Phenomenology and psychopathology related to psychogenic movement disorders. Adv Neurol 1995;**65**:233–257.
6. Berrios GE, Wagle AC, Markova IS, Wagle SA, Rosser A, Hodges JR. Psychiatric symptoms in neurologically asymptomatic Huntington's disease gene carriers: a comparison with gene negative at risk subjects. Acta Psychiatr Scand 2002;**105**:224–230.
7. Nowak DA, Fink GR. Psychogenic movement disorders: aetiology, phenomenology, neuroanatomical correlates and therapeutic approaches. Neuroimage 2009;**47**:1015–1025.
8. Ginovart N, Lundin A, Farde L et al. PET study of the pre- and post-synaptic dopaminergic markers for the neurodegenerative process in Huntington's disease. Brain 1997;**120**:503–514.
9. Kägi G, Bhatia KP, Tolosa E. The role of DAT-SPECT in movement disorders. J Neurol Neurosurg Psychiatry 2010;**81**:5–12.

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