Turkish Journal of Psychiatry 2021;32(4):286-290
https://doi.org/10.5080/u25540

A Case of Chronic Functional Parkinsonism Treated Over 10 Years for the Diagnosis of Juvenile Parkinsonism

Necati Serkut BULUT[1], Neşe YORGUNER[2], Gresa ÇARKAXHIU BULUT[3], Emine Neşe TUNCER[4]

SUMMARY

Functional (psychogenic) movement disorders (FMD) constitute a cluster of heterogeneous diagnoses involving motor symptoms that cannot be explained by organic pathology and are often associated with underlying psychological problems. In psychiatry, patients with FMD are often placed within the scope of somatoform disorders and conversion disorders. Functional Parkinsonism (FP) is a rare form of FMD seen in 1.5% of all patients presenting with symptoms of parkinsonism. Although almost all symptoms of parkinsonism can be present in FP, clinical features such as sudden onset and a non-progressive course, inconsistent response to pharmacotherapy, and atypical findings in neurological examination are considered as important clues of psychogenic aetiology. Limited data in the literature on FP indicate that the average age of onset is between 37-53 years of age, whereas the average onset age of Idiopathic Parkinson's Disease (IPD) is around 60; and Juvenile Parkinsonism, a rare condition presenting before the age of 21, is often familial and more closely related to genetic mutations.

Here, we present the case of a female patient, who, after the diagnosis of Juvenile Parkinsonism at the age of 17, had been treated with antiparkinsonian medications for about 14 years in the neurology clinic. Even though the age of onset of this case was far earlier than expected for both IPD and FP and the symptoms became chronic despite close monitoring, it is believed that this case is a striking example for the importance of the recognition of FP and early intervention.

Keywords: Movement disorders, conversion disorder, juvenile parkinsonism, somatoform disorders

INTRODUCTION

Functional movement disorders (FMD) are a cluster of heterogeneous symptoms of motor abnormalities including slowness, tremors, jerks, spasmodic movements and sometimes strange postures that cannot be explained by organic causes and often associated with underlying psychological problems. Although interchangeably described in the literature as functional and psychogenic, the term "functional" may be more appropriate in reflecting better the multifactorial etiology and biopsychosocial model associated with the diagnostic cluster (Czarnecki and Hallett 2012). In the absence of evidence for conscious awareness of the mechanism of symptom appearance, FMD cases are often considered within the scope of somatoform disorders and conversion disorders. The clinical presentation may also be associated with malingering and factitious disorder, in which motor symptoms are purposefully feigned by the patient (Jankovic 2011).

Functional Parkinsonism (FP) is quite rare, accounting for about 10% of all FMD cases (Hallett 2011) and approximately 1.5% of the patients followed up for parkinsonism (Sage and Mark 2015). All symptoms of parkinsonism may be present in FP, although some, including a sudden onset of symptoms, a non-progressive course, lack of response to treatment or inconsistent response over the disease course, disappearance of symptoms by distraction, exaggerated slowness, movements generated with great apparent effort accompanied by grimacing, sighing etc., are commonly regarded as important clues for psychogenic etiology (Hallett 2011). Nevertheless, it should be kept in mind that such atypical features can also be

Received: 10.03.2020, **Accepted:** 14.07.2020, **Available Online Date:** 22.12.2020

[1, 2]MD., Marmara University Pendik Training and Research Hospital, Department of Psychiatry, [3]Asst. Prof., Maltepe University Medical School, Department of Child and Adolescent Psychiatry, [4]Prof., Marmara University Pendik Training and Research Hospital, Department of Neurology, İstanbul, Turkey.

e-mail: serkutbulut@gmail.com

present in Idiopathic Parkinson's Disease (IPD) (LaFaver and Espay 2017).

The largest scale retrospective investigation for FP over 32-years in a movement disorders clinic reported a total of 36 FP patients with an average onset age of 51.5 years and 78% having the female gender (Sage and Mark 2015). The limited literature on FP indicated the age range of FP patients as 37-53 years (Kumar and Kumar 2018). In IPD, on the other hand, the average age of onset is around 60; and the subtype ''early-onset parkinsonism" (EOP), accounting for only 5-10% of the cases, presents under the age of 50 (De Lau and Breteler 2006, Lee and Gilbert 2016). Finally, Juvenile Parkinsonism, a very rare disease presenting under the age of 21, is often inherited in families and is more closely associated with genetic mutations (Schrag and Schott 2006).

Accurate and timely diagnosis of FP is critical for avoiding unnecessary and risky medical interventions and determining appropriate treatment options. Although the patient's psychosocial history with evident stressors, early life traumatic experiences and/or comorbid psychiatric disorders would have a role in suspecting a psychogenic etiology, the basic requirement for the diagnosis is the clinical evaluation by a neurologist with experience in identifying atypical presentations of organic movement disorders (Espay and Lang 2015). Differentiating FP from IPD can in some cases be extremely challenging even for the most experienced clinicians; and, consulting different treatment centers and physicians by the patients may delay the diagnosis for years.

In this article, we present a case of FP, diagnosed as Juvenile Parkinsonism some 14 years previously when the patient was 17 years of age, and followed up at neurology clinics with antiparkinsonian treatment. As a striking example of FP with an early age of onset, atypical presentation and chaotic clinical course of the Parkinsonism symptoms, we aimed to discuss the recognition of the disorder through specific findings and observations during psychiatric evaluations in order to emphasize the importance of early intervention.

CASE

The 31-years old single female patient with an associate bachelor's degree who had worked as a grocery clerk in a supermarket for only 6 months and seeking eligibility for retiring on grounds of disability was referred to the psychiatry committee by her treatment neurologist due to the atypical features of her late clinical course. The patient, youngest of three children born to unrelated parents was living with her mother after losing her father 4 years previously when her Parkinsonian symptoms gradually worsened preventing her from leaving home alone.

The patient's symptoms of movement disorder had first appeared at the age of 16. The first physician, consulted with complaints of tremor in her hands, the knees pulling backwards when walking, slowing down of movements and spasm in her toes, arranged her treatment with carbidopa/levodopa (25/250 mg 4x1). Despite temporary relief with the treatment, she discontinued the medication after the recurrence of her complaints; and was subsequently admitted to the neurology inpatient clinic of a university hospital for further examination and treatment of her symptoms. The results of her hemogram and detailed biochemical and serological tests, were all within normal limits. Screening for Wilson's disease did not show abnormality of urinary copper and ceruloplasmin levels, and Kayser-Fleischer ring was not detected in her ophthalmological examination. Also, pulmonary x-rays, electrocardiography (ECG) and cranial magnetic resonance imaging (MRI) did not indicate pathology. An asymmetrically decreased perfusion in the left thalamus, indicated by brain perfusion single photon emission computed tomography (SPECT) was regarded as clinically insignificant.

Apparent psychopathological observations were not reported and additional treatment was not recommended by the consultant psychiatrist. Genetic tests carried out at an external laboratory did not detect parkin gene mutation. On the basis of the preliminary diagnosis of EOP, the combination of levodopa/carbidopa/entacapone (100/25/200 mg 4x1/day) was started. This resulted in partial improvement in the patient's bradykinesia, rigidity and tremor. Cabergoline (4 mg/day) was also added to the treatment during follow up in the clinic. The patient was discharged with minimal bradykinesia and tremor, and slight bending backward in the right knee. She continued her monthly follow-up in the neurology outpatient clinic for approximately 5 years, during which, rasagiline (1 mg/day) and pramipexole (0.75 mg/day) were added to the treatment, given the fluctuating course of her symptoms. On account of significant peripheral edema, pramipexole was later replaced with ropinirole (4 mg/day). In the sixth year of her treatment, new complaints emerged, including déjà-vu attacks and shortness of breath. During her examination she tended to drag her right foot and her tremor was noted to have increased. It was decided to titrate the ropinirole dose to 24 mg/day, while continuing with levodopa (200 mg/day) and rasagiline (1 mg/day). By the eighth year of the disease, it was noted that the patient could barely stand while walking, experienced tremor attacks and her cigarette consumption had increased significantly. Referred to psychiatry for the second time, she was prescribed duloxetine (30 mg/day) and quetiapine (25 mg/day), which caused her tremor attacks to decrease significantly over time.

However, the patient decided to discontinue her treatment for reasons of alleged side effects of high blood pressure and epistaxis. During the follow up interviews, the patient began to exhibit absence seizures and tremors not consistent with

epileptic phenomenology that disappeared by distraction and speaking. Also, the patient's mother, who accompanied her during the interviews, was observed to make negative suggestions that "the contractions would not disappear and that they would never be able to cope with them". The patient reported having had similar fainting spells in the past when she remained conscious, without concurrent contraction or urinary incontinence and hearing what was spoken although unable to respond back. Her electroencephalography (EEG) was within physiological limits, without any detectable abnormal background activity and/or epileptic activity.

I was learned during follow up consultations that the patient began moaning during sleep and experienced severe tremors in her hands and legs following spells of weeping which ceased only if her mother grabbed her hand and caressed her head. Also, complaints of inability walking uphill or controlling her speed caused inability to leave her home alone. Hence, the patient was again referred to psychiatry with the recommendation for a closer follow-up on these atypical features. During the interviews at the psychiatry outpatient clinic, the 25 year old patient's childish and immature attitudes not appropriate for her age were noted by the clinicians. Her mother, on the other hand, complained that the patient was tampering with belongings of others without permission, that she was impatient, stubborn, and often lied.

Switching to bupropion (300 mg/day) after the poorly tolerated venlafaxine resulted in temporary and partial relief in the patient's depressive symptoms, but the symptoms recurred due to daily life stressors including her mother's health issues, and quarreling with relatives. In the 10th year of the disease, with ongoing neurology follow-up and combined antiparkinsonian therapy, the patient was hospitalized in the psychiatry clinic for further diagnostic evaluation when she requested discharge on the first day of her admission, claiming to be negatively affected by the ambience of the clinic. Although recommended by her neurologist to finalize the antiparkinsonian treatment as the symptoms were mostly psychogenic, the patient kept using the drugs by obtaining prescriptions from different hospitals during the following two years.

In the 13th year of the disease, with the suggestion of her mother, she reapplied to the institution where her past records were available, to initiate an evaluation procedure for disability retirement. She was assessed in a council of physicians specializing in neurological movement disorders. Not having observed any pathology in her repeated cranial MRI, the clinical presentation of the patient was considered to be in line with FP, due to the variability of movement disorder pattern, the disappearance of the symptoms with distraction, and the discordance of her tremor with the typical pattern. Moreover, the absence of dyskinesia, a complex motor phenomenon that develops due to chronic L-DOPA use in Parkinson's Disease, also suggested that the patient did not have a true dopaminergic denervation, which was another finding supporting the diagnosis of FMD.

The patient was once again referred to the psychiatry outpatient clinic for diagnostic evaluation, when a series of interviews with the patient indicated several features deemed clinically noteworthy. In her examination she had reduced spontaneous speech, tended to give short and shallow answers to the questions, and frequently made eye contact with her mother as if to take assurances before speaking. The mother, who insisted on being present, constantly intervened in the conversation, remarking that the information given by the patient might have been inaccurate or incomplete. The patient thought that feeling uneasy when alone and exacerbation of her symptoms by psychological stressors had a minor impact on the overall course of her disease. After 14 years of treatment, the patient was making inconsistent statements on the reasons for the use and the efficacy of antiparkinsonian and psychotropic agents. It was understood that she was still using antiparkinsonian agents prescribed by others against the decision of her neurologist. The patient's occupational and social functionality was significantly impaired and she experienced significant difficulties in maintaining for 6 months a job too simple for her intellectual capacity and level of education. She spent her time at home with her mother for reasons of inability to walk and not liking crowds. Her score on the Minnesota Multiphasic Personality Inventory (MMPI) indicated prominence of dependent personality traits. Diagnosed with chronic conversion disorder, close psychiatric follow-up and extensive psychotherapy were planned.

DISCUSSION

Diagnosing some FP cases can be challenging even for the experienced clinicians. Excluding the organic etiology of parkinsonism is not an adequate diagnostic approach by itself. The physician's familiarity with the disease phenomenology for identifying the clinical features of psychogenic etiology, such as the variable pattern of the motor symptoms and the disappearance of symptoms by distraction during examination as seen in our patient, and careful evaluation of the disease history together with detailed physical examination are of critical importance. Next to the neuropsychological and neurophysiological examinations, relevant laboratory tests and imaging techniques can also support the diagnosis. Hence, it should be noted that diagnosing FMD is not based on elimination but on identification of positive clinical criteria (Jankovic 2011, LaFaver and Espay 2017).

Our patient, with onset of symptoms at 16 years of age, has a very atypical clinical appearance with both IPD (mean age of onset>60) and FP (mean age of onset between 37-53). She was followed up with a preliminary diagnosis of the rare juvenile parkinsonism, and had undergone extensive

diagnostic procedures, including parkin gene mutation test and analyses for Wilson's disease. It should be kept in mind that in the diagnostic algorithm of the cases evaluated for juvenile parkinsonism, *PINK-1* and *DJ-1* gene mutations should also be considered in addition to the parkin gene, when the age of onset and the symptomatic features are consistent with idiopathic parkinsonism (Schrag and Schott 2006, Tang et al. 2006).

The differential diagnosis of functional movement disorder should particularly include *PINK-1* mutations, which may be associated with psychiatric symptoms such as anxiety, mood disorders, psychosis or motor disturbances (Steinlechner et al. 2007). Mutations other than the parkin gene could not be screened in our case, due to technical limitations. Apart from genetic tests, another method to guide the differential diagnosis of Parkinson's Disease is the sophisticated neuroimaging technique DaTScan (de la Fuente-Fernández 2012). Using a radioactive marker that binds to dopamine transporters, DaTScan can demonstrate dopaminergic denervation with very high sensitivity and specificity, although its use in our country is still limited. Since DaTScan could not be performed on our patient, the diagnosis of FP was largely based on the atypical clinical features observed in movement disorder symptoms following the reduction of drug doses. However, due to the aforementioned limitations, Parkinson's Disease or its comorbidity cannot be excluded with certainty in our patient.

Another difficulty in the diagnostic process is the possible comorbidity of organic and psychogenic parkinsonism. The literature indicates that concurrent comorbidity of organic Parkinson's Disease is more likely in FP as compared to other FMD cases. In a 9-case series, 5 patients were given both diagnoses at the end of the follow-up while 4 patients were diagnosed with FP alone (Felicio et al. 2010). Moreover, it is thought that FMDs may appear in some cases as early pre-motor manifestations of Parkinson's Disease (Onofrj et al. 2010).

Another aspect of psychiatric interest is our patient's insistent and inappropriate use of antiparkinsonian medications despite being stopped by the treating neurologist due to their ineffectiveness. Persistence of the disease since the age of 16 with characteristic clinical presentation and aggravation by stressors resulted in the exclusion of simulation and factitious disorder diagnoses, in which symptoms are deliberately produced. The patient's symptoms of parkinsonism, however, were not suggested to be generated voluntarily, nor did they appear to be directly motivated by a conscious gain (e.g. financial gain/legal privilege as in malingering, or to get medical care as in factitious disorder).

The patient's childish attitudes and low sociability, obviously incongruent with her intellectual capacity, had been noted by many physicians during her long-term history of psychiatric treatment for anxiety and depression comorbid with Parkinson's Disease. It is understood that losing her father had caused complete dependence on her mother and experiencing anxiety outside the home had made her housebound. Considering that movement disorder symptoms contributed to a way of life exempt from adulthood responsibilities and the strengthening of the dependence on her mother; the chronic course of disease may accord with the psychodynamic theory of conversion disorders. In other words, the patient's symptoms of movement disorder, triggered by unconscious mechanisms and remaining uncontrollable despite comprehensive treatments, may have led her and her social environment to accept in time the "vulnerable and needy child" identity, thereby fending off the dissociation from the mother and serving the purposes of 'secondary gain' by getting exempted from adult responsibilities (van Egmond 2003). However, conversion disorder be historically associated with histrionic (hysterical) personality, research points to a stronger relationship between conversion and "passive-dependent" or "psycho-infantile" personality traits (Chodoff and Lyons 1958, Ford and Folks 1985). Similarly, to personality disorders, FMD cases may involve more comorbid depression and anxiety disorders, with reported incidences of, respectively, 42.9% and 61.9%, in comparison to Parkinson's disease (Feinstein et al. 2001).

Presenting with conversion disorder in childhood, as in the case of our patient, is not uncommon. Childhood cases, mostly presenting with motor symptoms such as paralysis, tremors, coordination and gait disturbances, are most common between 10-15 years of age, and twice as common among girls than boys. About 10% of the pediatric outpatients admitted to a neurology clinic in England were diagnosed with conversion disorder (Leary 2003). Follow-up studies indicate complete recovery in 85-97% of childhood conversion disorder cases, with the majority being resolved in less than four weeks after accurate diagnosis and appropriate intervention. Presence of multiple symptoms, a history of sexual abuse, depression, anxiety disorders, and pathological personality traits have been shown to be negative prognostic factors (Leary 2003, Pehlivantürk and Unal 2002).

CONCLUSION

In being a rare form of FMD, differentiation of FP from IPD in some cases can be extremely challenging for clinicians. Accurate and timely diagnosis of FP from a multidisciplinary perspective and determination of appropriate psychiatric treatment options in order to avoid the risk of harming the patient with unnecessary examinations and medical interventions are of crucial importance.

REFERENCES

Chodoff P, Lyons H (1958) Hysteria, the hysterical personality and "hysterical" conversion. Am J Psychiatry 114:734-40.

Czarnecki K, Hallett M (2012) Functional (psychogenic) movement disorders. Curr Opin Neurol 25:507-12.

de la Fuente-Fernández R (2012) Role of DaTSCAN and clinical diagnosis in Parkinson disease. Neurology 78:696-701.

de Lau LM, Breteler MM (2006) Epidemiology of Parkinson's disease. Lancet Neurol 5:525-35.

Espay AJ, Lang AE (2015) Phenotype-specific diagnosis of functional (psychogenic) movement disorders. Curr Neurol Neurosci Rep 15:32.

Feinstein A, Stergiopoulos V, Fine J et al (2001) Psychiatric outcome in patients with a psychogenic movement disorder: a prospective study. Cogn Behav Neurol 14:169-76.

Felicio AC, Godeiro-Junior C, Moriyama TS et al (2010) Degenerative parkinsonism in patients with psychogenic parkinsonism: a dopamine transporter imaging study. Clin Neurol Neurosurg 112:282-5.

Ford CV, Folks DG (1985) Conversion disorders: an overview. Psychosomatics 26:371-83

Hallett M (2011) Psychogenic parkinsonism. J Neurol Sci 310:163-5.

Jankovic J (2011) Diagnosis and treatment of psychogenic parkinsonism. J Neurol Neurosurg Psychiatry 82:1300-3.

Kumar R, Kumar R (2018) A case of psychogenic Parkinsonism: late age of onset should not be a barrier to make the diagnosis. Aust N Z J Psychiatry 52:1098

LaFaver K, Espay AJ (2017) Diagnosis and treatment of functional (psychogenic) Parkinsonism. In Semin. Neurol pp. 228-32. Thieme Medical Publishers.

Leary P (2003) Conversion Disorder in Childhood–-Diagnosed Too Late, Investigated Too Much? J R Soc Med 96:436-8.

Lee A, Gilbert RM (2016) Epidemiology of Parkinson disease. Neurol Clin 34:955-65.

Onofrj M, Bonanni L, Manzoli L et al (2010) Cohort study on somatoform disorders in Parkinson disease and dementia with Lewy bodies. Neurology 74:1598-606.

Pehlivantürk B, Unal F (2002) Conversion disorder in children and adolescents: a 4-year follow-up study. J Psychosom Res 52:187-91.

Sage JI, Mark MH (2015) Psychogenic parkinsonism: clinical spectrum and diagnosis. Ann Clin Psychiatry 27:33-8.

Schrag A, Schott JM (2006) Epidemiological, clinical, and genetic characteristics of early-onset parkinsonism. Lancet Neurol 5:355-63.

Steinlechner S, Stahlberg J, Volkel B et al (2007) Co-occurrence of affective and schizophrenia spectrum disorders with PINK1 mutations. J Neurol Neurosurg Psychiatry 78:532-5.

Tang B, Xiong H, Sun P et al (2006) Association of PINK1 and DJ-1 confers digenic inheritance of early-onset Parkinson's disease. Hum Mol Genet 15:1816-25.

van Egmond JJ (2003) The multiple meanings of secondary gain. Am J Psychoanal 63:137-47.