DOI: 10.37990/medr.1007715 Med Records 2022;4(1):117-9

MEDICAL RECORDS-International Medical Journal

Olgu Sunumu/ Case Report

Progressive Supranuclear Palsy Following as Major Depressive Disorder: A Case Report

Majör Depresif Bozukluk Olarak Takip Edilen Progresif Supranükleer Palsi: Bir Olgu Sunumu

Mehmed Burak Erdaş

Artvin State Hospital, Department of Psychiatry, Artvin, Turkey

Abstract

Progressive Supranuclear Palsy (PSP) is a rare neurodegenerative disease that is classified among Parkinson plus syndromes, manifesting itself with behavioral, cognitive and emotional symptoms as well as parkinsonian symptoms such as postural instability, ophthalmoplegia, bradykinesia, rigidity affecting the brain stem, basal ganglia and cerebellum. Although there is sufficient information about the pathological process and clinical presentation of PSP, there is no valid biomarker for diagnosis, clinical findings and neuroimaging are very important in diagnosis, patients often get misdiagnosis at their first application. It has been reported that psychiatric symptoms are common in these individuals due to the involvement of the frontal-subcortical circuits, and therefore patients can get psychiatric diagnoses in the early period. We aimed to present a 72-year-old female patient who was followed up with the diagnosis of major depressive disorder in various centers for about 4 years, who did not benefit from psychiatric treatment, and diagnosed with PSP eventually. We also aimed to emphasize that importance of considering neurodegenerative diseases in differential diagnosis in patients whose depressive symptoms begin at an advanced age, are resistant to treatment and have atypical symptoms such as the presence of accompanying neurological findings, and psychiatric symptoms may be the first symptom of neurodegenerative diseases.

Keywords: Depression, neurodegenerative diseases, Progressive supranuclear palsy

Öz

Progresif Supranükleer Palsi (PSP), Parkinson plus sendromları arasında sınıflandırılan, davranışsal, bilişsel ve duygusal semptomların yanı sıra postural instabilite, oftalmopleji, bradikinezi,rijidite gibi parkinsonyen semptomlarla kendini gösteren beyin sapını, bazal ganglionları ve serebellumu etkileyen nadir bir nörodejeneratif hastalıktır. PSP'nin patolojik süreci ve klinik görünümü hakkında yeterli bilgi olmasına rağmen, tanı için geçerli bir biyobelirteç bulunmamakta, klinik bulgular ve nörogörüntüleme tanıda çok önemli yer edinmekte, hastalar ilk başvurularında sıklıkla yanlış tanı almaktadır. Bu kişilerde frontal subkortikal devrelerin etkilenmesi nedeniyle psikiyatrik belirtilerin yaygın olduğu ve bu nedenle hastaların erken dönemde psikiyatrik tanı alabileceği bildirilmiştir. Burada yaklaşık 4 yıldır çeşitli merkezlerde major depresif bozukluk tanısıyla takipli, psikiyatrik tedaviden yarar görmeyen , değerlendirmeler sonucunda PSP tanısı alan 72 yaşında kadın olgunun sunulması amaçlanmış olup, depresif semptomları ileri yaşta başlayan, tedaviye dirençli ve eşlik eden nörolojik bulguların varlığı gibi atipik belirtileri olan hastalarda ayırıcı tanıda nörodejeneratif hastalıkların düşünülmesinin önemi, psikiyatrik belirtilerin nörodejeneratif hastalıklarda ilk belirti olabileceği vurgulanmak istenmiştir.

Anahtar Kelimeler: Depresyon, nörodejeneratif hastalıklar, progresif supranükleer palsi

INTRODUCTION

Progressive supranuclear palsy (PSP) is a rare neurodegenerative disease affecting the brainstem, basal ganglia, and cerebellum. The age of onset of the disease is often between 60 and 65 years, and its frequency does not differ between genders (1). PSP is a disorder manifested by postural instability, ophthalmoplegia, bradykinesia, rigidity as well as behavioral, cognitive and emotional symptoms. Postural instability and falls are the most common complaints. Patients' gait is slow and unsteady. Bradykinesia and rigidity are symmetrical. Dysarthria and dysphagia can be seen due to pseudobulbar paralysis. Frontal lobe symptoms occur at the early stages (2). It has been reported that psychiatric symptoms are common in these individuals due to the involvement of the frontal-subcortical circuits, and therefore patients can get psychiatric diagnoses in the

Geliş Tarihi / Received: 10.10.2021 **Kabul Tarihi / Accepted:** 17.11.2021
Sorumlu Yazar /Corresponding Author: Mehmed Burak Erdaş, Republic of Turkey Ministry of Health, Artvin State Hospital E-mail: dr.mehmedburakerdas@gmail.com

early period (3). Among the neuropsychiatric symptoms of PSP; apathy, depression, sleep disorders, personality changes, disinhibition, and cognitive disorders can be listed (4). In this study, we aimed to share the clinical features of a patient who had been followed up with depression for a long time and was diagnosed with PSP.

CASE REPORT

72-year-old female, unemployed and primary school graduate patient applied to the psychiatry clinic with complaints of unwillingness, despair, inability to enjoy life, not wanting to leave the house, occasional crying attacks, inability to fall asleep and inability to continue to sleep, which started about four years ago and continued increasingly. The patient applied to various outpatient psychiatry clinics and was diagnosed with depression and Fluoxetine and Trazodone were started. The patient used these medications regularly for 1 year but did not get any response. The patient applied to other clinics due to depression and she was started on Escitalopram, Venlafaxine and Quetiapine due to depression diagnosis and Donepezil due to memory problems, despite not being diagnosed with dementia. The history of patient revealed that she had fallen four times in the last three years and was conscious at the time of the fall and she had balance problems while walking. She had a history of hypertension, and major depressive disorder for 4 years, without a history of family psychiatric or substance abuse. In the psychiatric examination; depressive mood, anhedonia, avolition, difficulty in falling asleep, decreased speech fluency, psychomotor retardation, attention and memory impairment were found. In the neurological examination; hypomimia, hypophonia, bilateral bradykinesia, bilateral rigidity, postural instability, bilateral supranuclear gaze paralysis and slowed saccadic eye movements were detected. Laboratory tests of the patient were found to be normal, the patient got 24/30 points from the Standardized Mini Mental Test. Cranial MRI revealed midbrain-mesencephalon atrophy. Hummingbird appearance was observed in sagittal section (Figure-1).

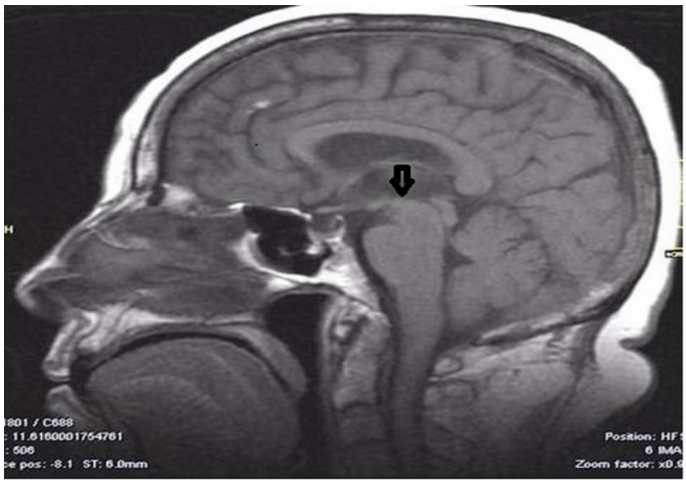

Figure 1. Hummingbird sign; Mid-sagittal T1-weighted sequence MRI brain showing atrophy of the midbrain tegmentum, with a relatively preserved pons, decreased midbrain to pons ratio with a superior aspect concavity, resembling the head and body, respectively, of a hummingbird

After the current clinical picture and imaging tests, the patient was referred to neurology with the prediagnosis of probable PSP, diagnosis was made according to the National Institute of Neurological Disorders and Stroke and Society for Progressive Supranuclear Palsy (NINDS-SPSP) diagnostic criteria and then L-Dopa was started at 500 mg / day. Her psychiatric treatment was arranged as Sertraline 100 mg / day and Zopiclone 7.5 mg / day. The patient who responded poorly to L-Dopa treatment had a partial regression in her depressive mood and insomnia during the follow-up period, and her response status was determined by clinical evaluation and a 25% decrease in the Hamilton Depression Scale score compared to the baseline score. The patient's follow-up continues in neurology and psychiatry outpatient clinics. Written consent was obtained from the patient for publishing case report.

DISCUSSION

Treatment-resistant depression and sleep disorder started about 4 years before the diagnosis of PSP. Although there is a high possibility of depression in patients with PSP in the following years, there are also cases that present with psychiatric symptoms and are later diagnosed with PSP (5). It has been stated that psychiatric symptoms are quite common in these individuals due to the involvement of the frontal-subcortical circuits, and therefore patients can also be diagnosed with psychiatric disorders in the early period (3). Although many neurobiological models related to the frontal lobe and its functions have been proposed, the fronto-subcortical loops start from the cortex and follow a path from the cortex to the striatum, globus pallidum and dorso-medial thalamus. It is thought that there are five cycles following the mentioned path starting from the frontal cortex, two of them are related to eye movements and the other three are closely related to the behavioral dimension. These 3 pathways closely related to the behavioral dimension are the anterior cingulate loop, orbito-frontal loop and the dorsolateral prefrontal loop, and the damage in the pathway is clearly reflected in the clinic (6). Supranuclear gaze paralysis diagnosed in the neurological examination of our patient was indicating fronto-subcortical circuit dysfunction which is responsible from eye movements. The loss of energy and motivation, depressive mood and apathy detected during the psychiatric examination attributed to involvement of the anterior cingulate loop. Difficulty in focusing and maintaining attention, cognitive impairment, and decrease in speech fluency occurred as a result of the involvement of the dorsolateral prefrontal cycle which is associated with executive functions.In Parkinson's disease and Huntington's disease, the involvement of subcortical pathways is asserted to be responsible from the dementia accompanying movement disorders (7). In our case, the findings and neuroimaging results support the diagnosis of progressive supranuclear palsy. The hummingbird sign image is diagnostic for the diagnosis of PSP and has 100% of sensitivity (8). It has

been reported that falls usually occur within three years after the onset of the disease, and MRI findings correlate with the stage of the disease (9). Apathy was reported to be the most common neuropsychiatric symptom in PSP and it is difficult to distinguish it from depression. It has been stated that apathetic individuals generally describe their own moods as happy and pleasant unlike individuals with depression, and depression and apathy may coexist very often. It has been reported that apathy can be treated with antidepressant treatment when depression accompanies apathy (10,11). In the literature, it has been stated that PSP does not respond or respond poorly to L-Dopa treatment, and insomnia is a common symptom in PSP, patients have prolonged sleep latencies and have difficulty in maintaining sleep at the same time, REM sleep is shortened while the rate of sleep-related movement disorders and sleep apnea is increased (12,13). It has been stated that Zolpidem can positively contribute to motor functions and supranuclear gaze paralysis in PSP, and it has positive contributions to sleep disorders in Parkinson's patients. Since Zolpidem is not available in our country, Zopiclone, which is another nonbenzodiazepine GABA agonist, was started (14,15).

CONCLUSION

Considering the patient's clinic and neuroimaging findings, we thought that PSP started years ago with psychiatric presentation since depression symptoms and insomnia did not respond to psychiatric treatment for many years and patient's parkinsonism symptoms didn't respond to L-Dopa treatment. Psychiatrists should be more careful in terms of neurodegenerative diseases in patients with atypical symptoms such as the onset of depressive symptoms at an advanced age, resistance to treatment and the presence of accompanying neurological findings, as in our patient. It should also be kept in mind that neurodegenerative diseases may also present with psychiatric symptoms and that depression and insomnia may be the first symptom of neurodegenerative diseases.

Financial disclosures: *All authors report no financial interests or potential conflicts of interest.*

Conflict of Interest: *The authors declare that they have no competing interest.*

Informed Consent: *Informed consent was taken from the patient.*

REFERENCES

1. Morris HR, Wood NW, Lees AJ. Progressive supranuclear palsy (Steele-Richardson-Olszewski disease). Postgrad Med J. 1999;579-84.
2. Litvan I, Mangone CA, McKee A, et al. Natural history of progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome) and clinical predictors of survival: a clinicopathological study. J Neurol Neurosurg Psychiatry. 1996:615–20.
3. Grafman J, Litvan I, Gomez C, Thomas NC. Frontal lobe dysfunction in progressive supranuclear palsy. Arch Neurol. 1990;553-8.
4. Madhusoodanan S, Wilkes V, Campbell RP, et al. Psychiatric symptoms of progressive supranuclear palsy: A case report and brief review. Neuropsychiatry. 2014;27–32.
5. Chiu YW, Lee SH, Yeh TH. Diversified psychiatric presentation in a case of progressive supranuclear palsy. J Clin Gerontol Geriatrics. 2016;7:164-7.
6. Cummings JL. Frontal-subcortical circuits and human behavior. Arch Neurol. 1993:873-80.
7. Erkol G, Karaali Savrun F. Frontotemporal dementia. Cerrahpaşa J Med. 2002;256-60.
8. Groschel K, Kastrup A, Litvan I, Schulz JB. Penguins and hummingbirds: midbrain atrophy in progressive supranuclear palsy. Neurology. 2006;949-50.
9. Halliday GM, Hardman CD, Cordato NJ, et al. A role for the substantia nigra pars retriculata in the gaze palsy of progressive supranuclear palsy. Brain. 2000:724-32.
10. Levy ML, Cummings JL, Fairbanks LA, et al. Apathy is not depression. J Neuropsychiatry Clin Neurosci. 1998;314–19.
11. Rittman T, Coyle-Gilchrist IT, Rowe JB. Managing cognition in progressive supranuclear palsy. Neurodegenerative Disease Management 2016:499-508.
12. Litvan I, Bhatia KP, Burn DJ, et al. Movement Disorders Society Scientific Issues Committee. Movement Disorders Society Scientific Issues Committee report: SIC Task Force appraisal of clinical diagnostic criteria for Parkinsonian disorders. Mov Disord. 2003;18:467-86.
13. Walsh CM, Ruoff L, Walker K, et al. Sleepless Night and Day, the Plight of Progressive Supranuclear Palsy. Sleep 2017;154.
14. Abe K, Hikita T, Sakoda S. A hypnotic drug for sleep disturbances in patients with Parkinson's disease. No to Shinkei = Brain and Nerve 2005;57:301-5.
15. Cotter C, Armytage T, Crimmins D. The use of zolpidem in the treatment of progressive supranuclear palsy. J Clin Neurosci. 2010;17:385-6.