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Case Report

Atypical Parkinsonism-A Masked Face: Case Report

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ABSTRACT

Progressive Supranuclear Palsy (PSP) is an uncommon neurological condition identical to Parkinson's Disease (PD) in clinical presentation. This makes the illness to be often misdiagnosed. A female who was previously misdiagnosed as PD presented with complaints of off-balance, swallowing difficulty, eye-ball movement restriction along with tremors. The patient was initially treated with Levodopa-Carbidopa, but she became unresponsive to the treatment and the condition gradually worsened. Upon comprehensive clinical discussion and examination, the patient was diagnosed with PSP. Currently, there are no interventions available to manage PSP, whereas physiotherapy and regional exercises are the cornerstone to improve the patient's state. This is a case of misdiagnosis, and the conclusion of this instance may pave the way for more accurate diagnosis and consequent patient care.

Keywords: Progressive Supranuclear Palsy (PSP), Parkinson's Disease (PD), Atypical Parkinsonism, Misdiagnosis.

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INTRODUCTION

Atypical Parkinsonism, also known as Progressive Supranuclear Palsy (PSP), is an uncommon neurological condition marked by aberrant tau aggregation in the form of round neurofibrillary tangles and crocheted astrocytes.[1] The illness is frequently mistaken as Parkinson's Disease (PD) because of this process. On the therapy front, PSP trails a majority of neurological illnesses. PSP's lacklustre reaction to dopamine supplementation or dopaminergic receptor activation may be explained by the degeneration of postsynaptic striatal neurons.[2] PSP's generic manifestation with postural instability or behavioural abnormalities and the lack of an accessible, prompt diagnostic test are further factors. These factors combine to cause a three- to four-year delay in an accurate diagnosis, by which time half of the clinical process has passed and neuroprotective therapy is unlikely to benefit.[3] The focus of the subsequent case study is also on the patient's illness being misdiagnosed and misunderstood.

CASE PRESENTATION

On August 16, 2023, a 67-year-old woman of average build reported to the emergency room concerning swallowing problems that had been plaguing her for a month but had gotten worse during the previous week. As a result, the patient lost her appetite and frequently complained of having difficulty swallowing. Over time, the patient developed severe throat spasms and was unable to consume even watery beverages like milk. In addition to dysphagia, she occasionally had uncontrollable movements of her extremities and other sensory problems such as blurring, paraesthesia, and numbness across her entire body below the neck. Additionally, she reported having trouble sleeping and having walked off balance. The patient originally was diagnosed with PD and was prescribed, Levodopa-Carbidopa 1-1-1, as well as a delayed release tablet HS. At night, she was on Escitalopram and Clonazepam. She was a known case of Hypertension, Diabetes and Bronchial Asthma.

The patient's vital indicators showed blood pressure of 80/60 mmHg, heart rate of 84 beats per min, respiratory rate of 26 cycles per min, and SpO_2 measurement of 84% in room air and 98% on $4LO_2$/min. The patient appeared pale, had a hunched posture, and was drowsy yet cognizant of regional discomfort. Her GCS was found to be $E_4V_2M_4$ (10/15) and her random blood sugar level was 167 mg/dL. Other investigations like ECG, 2D Echo and abdominal scan were normal with senile alterations. Whereas, after 48 hr of incubation, her urine culture and sensitivity test confirmed the development of E. coli (1,00,000 cfu/mL), while her chest X-ray showed cardiomegaly and haziness in the right lower zones.

She was bed-bound and her daughter-in-law was assisting her daily activities. Additionally, she had a history of cough during feeding and complained of sore throat. Initially, she was diagnosed with bronchial asthma flare. Therefore, she was admitted to the ward and was administered Cefoperazone-Sulbactam 1.5 g 1-0-1,

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Table 1: Summary of Progressive Supranuclear Palsy rating scale.

Parameter	Scores range	Test Scores of the Patient
History	0-24	14
Mentation	0-16	11
Bulbar	0-8	6
Ocular	0-16	4
Limb	0-16	6
Gait	0-20	17
Total	0-100	58

Pan 40 mg 1-0-0, Multivitamin, Salbutamol nebulization TID, and Prucalopride 1mg 0-0-1 along with routine medications. On second day of hospitalization, it was discovered that the patient was unresponsive to Levodopa-Carbidopa, thus the frequency was altered from 1-1-1 (7 am-1 pm-7 pm) to 1-1-1-1 (7 am-1 pm-4 pm-7 pm). Even on the third day, the patient remained unresponsive to anti-Parkinson's medications, and the patient was suspected of Progressive Supranuclear Palsy (PSP). The speech-language pathologist conducted a comprehensive assessment since the patient had complained of dysphagia. The patient exhibits limited jaw movement, hypokinetic dysarthria, weak lingual and labial movement and strength, whereas dysphagia is evident in the patient. Oral-motor exercises along with speech and swallowing therapy were recommended for the patient. The patient showed a positive Babinski sign and a 58/100 PSP rating scale (Table 1).[4]

On this basis, the patient was advised to perform daily oral-motor exercises in addition to chest and limb physiotherapy. To assist feeding, she was advised with PEG-tube insertion. Upon consent, the patient underwent PEG insertion. After receiving speech and swallowing treatment, the patient gradually improved lingual mobility and ability to consume thin liquids like milk. Severe throat discomfort and constipation were reported by the patient on the seventh day and was treated with analgesics and laxative suppositories, respectively. Following discharge, the patient was instructed to continue regular medications, frequent physiotherapy, assisted walking, and engage in moderate physical activity.

DISCUSSION

PSP is an unusual neurological condition that gradually impairs balance, eye movements, along with stiffness, dysarthria, and dysphagia. Although it is estimated that PSP affects 5–17 in every 1,00,000 people, post-mortem investigations have discovered PSP histopathology in 2-6% of geriatrics who had no diagnosis of PSP prior to death.[2] The condition usually manifests between the ages of 40 and 77 years, with a mean age of 61.5 years with no identifiable risk variables.[5,6] The pathophysiology of PSP was a revolutionary discovery that allowed clinicians to distinguish between illnesses like Parkinson's and Alzheimer's Disease that share identical neuro-hormonal characteristics. PSP, also known as Steele-Richardson-Olszewski syndrome, is becoming more widespread in modern times and is frequently misunderstood since it can present to a variety of medical specialisations.[7]

In the instance being discussed, the illness was initially misdiagnosed as PD, and when the patient became unresponsive to levodopa, PSP was suspected. When the patient's background was pulled out, it was discovered that the patient previously had appeared with practically identical concerns. Because the pathophysiology of PSP and PD coincides, the earlier therapy was effective for a time. The patient was later diagnosed with PSP after a comprehensive examination and the presentation of characteristic symptoms. The differentiating characteristic of eye-related motions, particularly those impacting downward motion in contrast to PD, is one of the distinctions to bear in consideration, whilst other symptoms align with PD. Whilst, axial stiffness is a crucial differentiating feature of PSP. In the above scenario, patient displayed both axial stiffness and tremors. The patient was 67 years old and had presented with off-balance and dysphagia which was progressive. There was no family history of PSP. Based on the clinical parameters, the patient can be classified as Level 1, indicating a high likelihood of PSP.[8]

The patient had blurry vision, and an ophthalmologic assessment revealed that she had restricted vertical flexibility of gaze, especially to the downward motion, making her eligible for O1 classification. Additionally, the fact that the patient is bed-bound, has experienced many falls in recent years, and needs assistance with daily tasks suggests that the patient falls into P1. The patient also had rigidity, tremors, and an inability to respond to levodopa hence meeting the criteria for A1. In parallel, the patient also displayed classic dysphagia and dysarthria which indicates the inclusion of C1. Levodopa and axial unresponsiveness, along with the patient's akinetic-rigidity, rank her even in the A2 category, but the severity and presentation of other symptoms forth her overall at Level 1. Considering the patient exhibits tremor as well as modest cognition symptoms, the described case may represent a Parkinson-like variation of PSP.

CONCLUSION

PSP is a rare neurological illness with an unclear etiology that calls for particular consideration in treatment since it has a direct impact on quality of life. There are currently no pharmaceutical interventions that prevents progression; nevertheless, interdisciplinary collaborative efforts may speed disease identification and maximize living standards. This is a case of misdiagnosis, and the conclusion of this instance may pave the way for more accurate diagnosis and consequent patient care.

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CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

PATIENT CONSENT

The patient has been informed about the publishing and assured that the information will only be used for scientific and research purposes and identity of patient will not be disclosed.

ABBREVIATIONS

PSP: Progressive Supranuclear Palsy; **PD:** Parkinson's Disease; **GCS:** Glasgow Coma Scale; **ECG:** Echocardiogram; **PEG:** Percutaneous Endoscopic Gastrostomy.

SUMMARY

Atypical Parkinsonism, also known as Progressive Supranuclear Palsy (PSP), is an uncommon neurological condition marked by aberrant tau aggregation in the form of round neurofibrillary tangles and crocheted astrocytes. The illness is frequently mistaken as Parkinson's Disease (PD) because of this process. The focus of the subsequent case study is also on the patient's illness being misdiagnosed and misunderstood. a 67-year-old woman of average build reported to the emergency room concerning swallowing problems that had been plaguing her for a month but had gotten worse during the previous week. As a result, the patient lost her appetite and frequently complained of having difficulty swallowing. Over time, the patient developed severe throat spasms and was unable to consume even watery beverages like milk. In addition to dysphagia, she occasionally had uncontrollable movements of her extremities and other sensory problems such as blurring, paraesthesia, and numbness across her entire body below the neck. The patient originally was diagnosed with PD and was prescribed, Levodopa-Carbidopa 1-1-1, as well as a delayed release tablet HS. At night, she was on Escitalopram and Clonazepam. On second day of hospitalization, it was discovered that the patient was unresponsive to Levodopa-Carbidopa, thus the frequency was altered from 1-1-1 (7 am-1 pm-7 pm) to 1-1-1-1 (7 am-1 pm-4 pm-7 pm). Even on the third day, the patient remained unresponsive to anti-Parkinson's medications, and the patient was suspected of Progressive Supranuclear Palsy. The patient showed a positive Babinski sign and a 58/100 PSP rating scale. On this basis, the patient was advised to perform daily oral-motor exercises in addition to chest and limb physiotherapy. Considering the patient exhibits tremor as well as modest cognition symptoms, the described case may represent a Parkinson-like variation of PSP.

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