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DIAGNOSTIC DILEMMAS OF PROGRESSIVE SUPRANUCLEAR PALSY

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ABSTRACT:

Bckground: In this report authors discuss similarities between psychiatric phenomenon i.e. catatonia and neuropsychiatric syndromes such as progressive supranuclear palsy (PSP) in a 51 years old male, known case of hypertension and diabetes, who presented to the psychiatric services with mutism, stereotypical movements of upper limbs, echolalia, anteriorly flexed neck, swallowing difficulty, decreased oral intake, urinary and fecal incontinence for a period of 1 year worsened for 6 months. Patient was initially treated for catatonia with benzodiazepines and electroconvulsive therapy but did not improve. A neuropsychiatric consultation was sought and a diagnosis of possible PSP was made according to the National Institute of Neurological Disorders and Stroke and Society for PSP (NINDS-SPSP) criteria.

The authors discuss the importance of considering other differential diagnoses such as PSP in patients who present with symptoms that may resemble catatonia and psychiatric syndromes.

INTRODUCTION: Catatonia is a clinical syndrome characterized by motoric immobility or excitement, profound negativism, echolalia (mimicry of speech) or echopraxia (mimicry of movement).[1] It may occur secondary to a general medical condition, as a result of primary psychiatric disorder such as schizophrenia or psychotic depression or during the course of delirium. Benzodiazepines and electroconvulsive therapy are the mainstay treatment of catatonia.[2]

Progressive supranuclear palsy is a rare and progressive neurodegenerative condition with vertical supranuclear gaze palsy plus postural instability and falls within the first year of symptom onset to diagnose “probable” PSP, and the presence of either supranuclear gaze palsy or a combination of slow vertical saccades and postural instability with falls within the first year to diagnose “possible” PSP.3 However patients with autopsy-confirmed PSP have been reported with variant PSP clinical presentations, including initial predominance of ocular motor dysfunction (PSP-OM), postural instability (PSP-PI), Parkinsonism resembling idiopathic Parkinson’s disease (PSP-P), frontal lobe cognitive or behavioral presentations (PSP-F), including behavioral variant frontotemporal dementia (bvFTD), progressive gait freezing (PSPPGF), corticobasal syndrome (PSP-CBS), primary lateral sclerosis (PSPPLS), cerebellar ataxia (PSP-C), and speech/language disorders (PSP-SL), including nonfluent/agrammatic primary progressive aphasia (nfaPPA) and progressive apraxia of speech (AOS).4 Due to the complicated presentations, low sensitivity of the diagnostic criterion and symptom overlap with some psychiatric conditions, the diagnosis is difficult to make at the first clinical visit and the mean duration is 3-4 years after the onset of first symptoms.[5]

CASE REPORT:

Mr. A, 51 years old male presented with memory problems for the last four years. This was followed by anxiety like symptoms, which included breathlessness, preference to sleep in open places, irritability and slurred speech. He also exhibited confusional states such as getting intimately close to other males who were strangers. He later developed difficulty in

swallowing which was gradual. Gait changes were also reported by the family, which started 6 months prior to his presentation and was described as shuffling with small steps.

He had become completely dependent on family for activities of daily living and also developed double incontinence. Features of dysautonomia such as incontinence and dysphagia were not present from the beginning in the course of the disease. This was elicited on history; autonomic testing was not done. Speech became scanty with element of echolalia.

Due to vague symptoms and complex presentation, he was referred to neurology to rule out any organic cause. Apart from brisk reflexes in lower limb, neurological examination was reported to be unremarkable and a working diagnosis of catatonia was made. Differential diagnoses included schizophrenia, dementia, Parkinson's disease and organic/neurological disorder.

On mental state examination he appeared to have an expressionless face with reduced blinking. No rest tremor was noted. Speech was slurred, with reduced output and echolalia was observed. Affect was restricted and flat. The following day, patient became mute and more perplexed and stopped following instructions. He maintained stooped postures and at other times used to pace. The psychiatry team was the primary team and started treatment of catatonia with oral lorazepam up to 12 mg per 24 hours in 6 divided doses. This was continued for 4 days. However, no improvement was observed and patient started developing increasing drowsiness and so lorazepam was reduced to 10 mg per day and Carbamazepine 50 mg twice a day was started which was later titrated to 100 mg twice a day. Other medications during the course of the disease included Sertraline and Quetiapine, which were stopped at the time of his admission. He was on them intermittently for last 6 months.

MRI of head and neck were carried out to look for any other causes of patients' symptoms.

Brain MRI showed mild diffuse cortical atrophy and mild periventricular and subcortical deep white matter ischemic changes. MRI neck showed multilevel disc osteophyte complexes with facet arthropathy with no signal abnormality in the cord. Psychiatry team decided to continue with catatonia as the working diagnosis and started administering ECT after detailed discussion with the family and taking informed consent. When no significant change in symptoms was observed, neuropsychiatric consultation was sought. On neuropsychiatric evaluation, patient was found to have vertical supranuclear gaze palsy, with rigidity and bradykinesia in the extremities, and dystonia/ anteroflexion of the neck. Speech was also dysarthric and spastic. On gait examination, he had small shuffling steps with reduced arm swing bilaterally and a stooped posture. On cognitive examination, patient was not testable as he was not responding to any questions or instructions. Based on the history, examination, and work up a diagnosis of possible PSP was made using the NINDS-SPSP criteria. MRI brain did not show the typical mid brain tegmentum atrophy or the humming bird sign seen in some typical cases of PSP.

DISCUSSION

This case illustrates diagnostic confusion in differentiating neurodegenerative dementias especially those that have motor and/or psychiatric symptoms with other psychiatric conditions such as catatonia. Initial motoric immobility, profound negativism, echolalia, reduced eating, and incontinence presenting in psychiatric ward led to working diagnosis of catatonia. Some initial Improvement from ECT and absence of any other major pathology on MRI added to further confusion. But later ophthalmologic symptoms, neck dystonia, parkinsonism and cognitive impairment favored the diagnosis of PSP. Although typical cases of PSP present with axial rigidity, gait instability, and falls followed by worsening parkinsonism, dysarthria, dysphagia, and eye movement abnormalities[6], there may be other modes of onset such as non specific dizziness, generalized motor slowing, and personality changes. Some other variants of PSP are also recognized, which may present differently than typical PSP, such as progressive supranuclear palsy-pure akinesia with gait freezing (PSP-PAGF), progressive supranuclear palsy-corticobasal syndrome (PSP-CBS), progressive supranuclear palsy-behavioral variant of frontotemporal dementia (PSP-bvFTD) and progressive supranuclear palsy-progressive non-fluent aphasia (PSP-PNFA).[7] Many times diagnosis of PSP is difficult to make and other conditions such as multiple system atrophy, frontal variant of PSP, or FTD may also be considered in such complicated cases. Absence of marked atrophy of mid brain or signal abnormalities in basal ganglia or pons on MRI, as in this case, added to the complexity of diagnosis. Usually MRI findings of PSP include atrophy of the mid brain, pons, and cerebellum with signal increase in inferior olives, atrophy of frontal and/ or temporal lobes, and often humming bird sign which appears due to rostral mid brain atrophy on mid sagittal images.[8] In this case absence of marked frontal atrophy and absence of dysautonomia including bladder and bowel incontinence early in the course of the disease made FTD and MSA less likely. Parkinson's disease typically presents with features of parkinsonism

including appendicular rigidity and rest tremor and may have focal dystonias at times, and typically responds well to levodopa. The neurochemistry of PCP is not well understood. Various theories have implicated alterations in dopamine, serotonin, acetylcholine and norepinephrine pathways. Treatment is symptomatic with L-dopa, dopamine agonists such as bromocriptine and methysergide, benztropine and amitriptyline in various combinations with varying degree of success. Unfortunately, there is no curative treatment available. [4] Pathologically, PSP is characterized by the accumulation of tau protein and neuropil threads, mainly in the pallidum, subthalamic nucleus, red nucleus, substantia nigra, pontine tegmentum, striatum, oculomotor nucleus, medulla, and dentate nucleus.[7] While PSP is a relatively rare syndrome, affecting less than 1% of the geriatric population, this case illustrates the importance of a thorough history, neurological exam, and neuropsychiatric findings to differentiate such cases from other neurological psychiatric conditions such as catatonia.

Not being able to find the correct diagnosis and non responsiveness to various psychiatric treatment modalities including to benzodiazepines and ECT added to family's distress. Although the prognosis of PSP is limited, making correct diagnosis, educating the family, and providing symptomatic treatment may help manage more effectively as in this case. The family was from a remote area in Sindh and went back to the village with follow up scheduled in 3-6 months.

Conclusion:

There may be some overlap and confusion between some psychiatric and neuropsychiatric symptoms, especially less common neurodegenerative conditions with motor and/or psychiatric presentations, which if not appreciated may lead to delayed diagnosis and mismanagement. It is important to keep PSP on the list of differential diagnoses with other organic diseases causing catatonia and depression.

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Humera Saeed; concept, data collection, data analysis, manuscript writing, manuscript review
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Syeda Masharab Jilani; data analysis, manuscript writing, manuscript review
Syeda Maheen Batool; data analysis, manuscript writing, manuscript review