Progressive Supranuclear Palsy: A case report in Southern Nigeria

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Abstract

Progressive supranuclear palsy (PSP) is a progressive neurodegenerative clinical disorder. It is often diagnosed as Parkinson's disease due to the similar clinical presentation of these neurodegenerative disorders. Diagnosing PSP in low-medium-income countries such as Nigeria can be challenging as it is more of a diagnosis of exclusion and this relies largely on neuroimaging. The increasing availability of MRI in Nigeria in the last decade has helped to shore up the level of diagnostic certainty of this condition.

We hereby report a 70-year-old male case of PSP primarily diagnosed with Parkinson's disease.

***Keywords**: Progressive, supranuclear, palsy, Magnetic resonance imaging (MRI)*

Introduction

In 1964, a clinicopathological syndrome defined by vertical gaze ophthalmoplegia, postural instability, dystonic rigidity of the neck and upper trunk, and cognitive dysfunction was reported by Richardson Olszewski and Steele.[1] Progressive supranuclear palsy (PSP) is a tauopathy with tau species being arranged in straight filamentous conformation.[2,3]

In Europe, there has been an increase in the prevalence of PSP to 8.8 - 10.8 per 100,000.[4,5] A similar trend can be observed in Japan, from 5.8 per 100,000 in 1999 to 17 per 100,000 in 2010.[6,7] This increase in prevalence can be attributed to expanded research criteria for the diagnosis of PSP.[8]

Patients with PSP present with postural instability and falls, dysarthria, bradykinesia, and supranuclear palsy which can be a deficit in downward or upward gaze and late in the disease horizontal gaze.[9]

In Nigeria, there have been cases where PSP was diagnosed as Parkinson's disease. The increase in the availability of neuroimaging techniques has had a significant impact on dealing with this problem.[10]

CASE REPORT

A 70-year-old man presented with complaints of difficulty with walking and slurred speech for a year, and difficulty looking to the sides for two months. There is a history of falls, difficulty with swallowing, and aggressive outbursts. The patient was noticed to be withdrawn as he prefers to keep to himself most of the time. He was previously diagnosed as a case of Parkinson's disease not doing well on levodopa/carbidopa for 6 months and was referred to us. He is not a known hypertensive, with no history of forgetfulness or urinary incontinence or tremors, visual hallucination, or dystonic posturing.

He was a conscious, elderly man with impaired vertical and lateral gaze, rigidity in all four limbs, and worst on the trunk. Muscle power assessment (MRC scale) was 4 globally, deep tendon reflexes were 3/5, and glabella tap was positive. Another general examination was essentially normal. However, Magnetic Resonance imaging showed the classic midbrain atrophy (Humming Bird sign) characterized by flattening of the superior aspect of the midbrain. Figure 1 when compared with findings in figure2 (B&C)[24]

The patient was subsequently placed on a tab of sertraline 50mg daily, a tab of risperidone 1mg nocte, and a tab of baclofen 5mg 12 hourly. The patient and his relatives were counseled on the need for physiotherapy. Patient symptoms improved as evidenced by his increased interaction with others, willingness to feed himself, and reduced aggression.

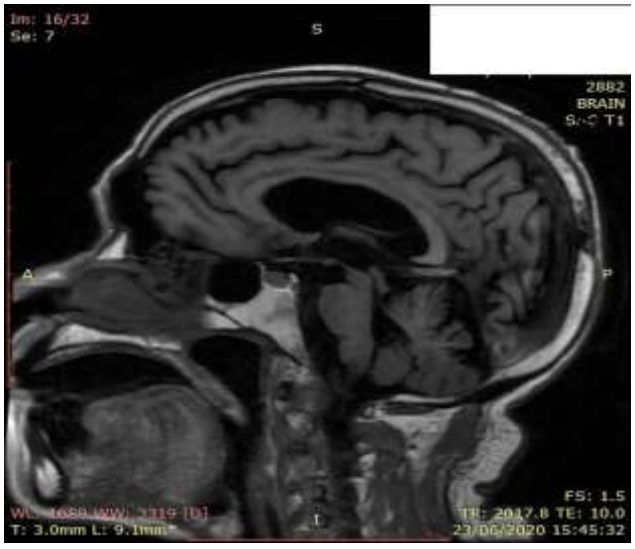

Figure 1: Mid-sagittal T1 weighted brain magnetic resonance image of our patient showing atrophy of the Dorsal Mid-Brain with relative sparing of Pons (the hummingbird's s Sign)

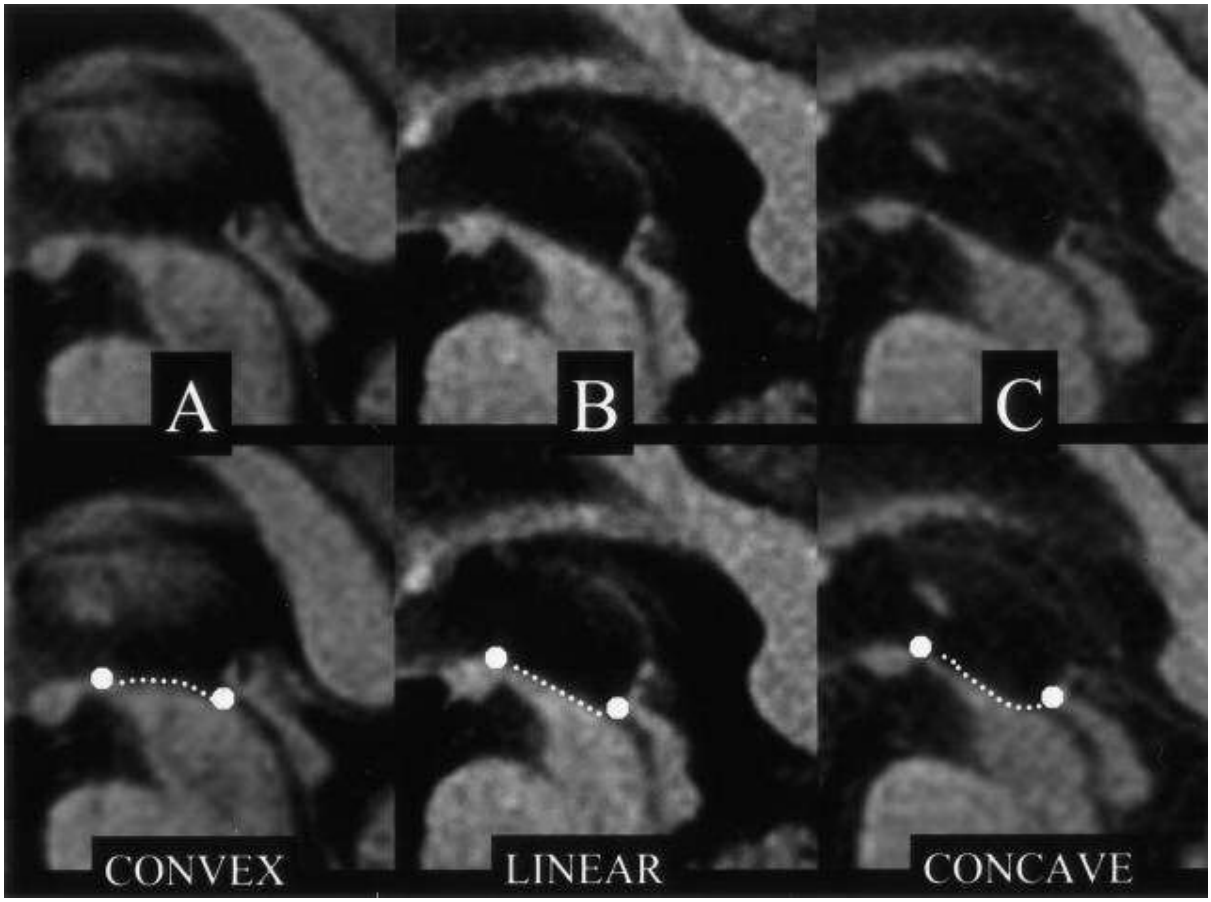

Figure 2 *Top row*, Midsagittal T1-weighted spin-echo sections in PD (*A*) and PSP (*B* and *C*) show the midbrain region. *Bottom row*, the same images with outlined profiles of the upper midbrain, appears convex in *A*, linear (flat) in *B* and concave in *C*.

DISCUSSION

Diagnosing PSP in medium –low-income countries such as Nigeria has received a significant boost due to the increasing availability of neuroimaging techniques. This has greatly improved the diagnostic accuracy of this disorder both locally and internationally. Despite this, the relevance of clinical judgment cannot be overlooked in the diagnosis as classical images may not be present early on in the disease.

The National Institute of Neurological Disorders and Stroke and the Society for Progressive Supranuclear palsy (NINDS-SPS) specify three degrees of diagnostic certainty: possible PSP, probable PSP, and definitive PSP. Definitive PSP requires histopathological evidence to make a diagnosis.[11]

Table1; Criteria for PSP[11]

Criteria for PSP
Inclusion criteria • postural instability with falls in the first year of symptoms • slowing of vertical saccadic eye movements (clinically possible); • vertical supranuclear gaze palsy(clinically probable) Supportive criteria • frontal/sub-cortical cognitive dysfunction • axial rigidity • pseudobulbar dysphagia and dysarthria • blepharospasm/apraxia of eyelid opening

Table2. The MDS-PSP clinical diagnostic Criteria as proposed[11]:

* "Definite PSP" can only be diagnosed by neuropathological examination at present. Currently, no other biomarker, imaging, or genetic finding with close to 100% sensitivity and specificity is available. *"Probable PSP" is diagnosed in the presence of a combination of clinical features that may not be very sensitive for PSP, but are considered to be highly specific, thus being ideally suited for therapeutic and biological studies, where it is important to exclude non-PSP from the subject group. *"Possible PSP" is diagnosed in the presence of clinical features that substantially increase sensitivity but at the possible cost of decreased specificity. This category is therefore suitable for descriptive epidemiologic studies and clinical care, where it is important not to exclude any cases of true PSP. With the addition of biomarkers to increase diagnostic specificity, these individuals might also be reasonably included in a therapeutic study. *Conditions "suggestive of PSP" represent subtle early signs of PSP, but do not meet the threshold for possible or probable PSP, and are suitable for early identification of individuals in whom the diagnosis may be confirmed as the disease evolves,

Progressive supranuclear palsy is currently being recognized as a spectrum of clinical phenotypes with the classic phenotype being " Richardson's Syndrome".[12,13] More recently the International Parkinson's and Movement Disorder Society (MDS) devised a criterion that

considers early forms and another phenotype. These criteria use neuroimaging, physiological, and fluid biomarkers. It recognizes a presymptomatic phase, a suggestive phase, and a symptomatic phase.[13]

The increasing availability of MRI in developing countries has greatly aided in diagnosing PSP. A study involving 21 patients with PSP, 23 patients with Parkinson's disease, 25 patients with Multiple system atrophy, and 31 age-matched normal control by Oba et al. concluded that a mid-brain area of < 70mm^2 was indicative of PSP.[14] The "Pegium" or "hummingbird " sign represents midbrain atrophy without pontine atrophy.[15].This sign on Midsagittal T1-weighted MRI is highly sensitive for making a diagnosis of PSP.[14,15,16] Another radiological evidence of PSP can be recognized by the atrophy of the superior cerebella peduncle as reported by Hiroshi Kataoka et al.[17]

Patients with Parkinson's disease, multiple system atrophy (MSA), and dementia with a lewy body (DLB) have no midbrain atrophy.[18] Selective atrophy of the midbrain tegmentum with relative sparing of the tectum and cerebral peduncles is often referred to as the 'Mickey Mouse' sign (Figures 2 and 3).[18] On the other hand, the 'Morning glory' sign represents the increased lateral concavity of the midbrain tegmentum.[19]

Multiple system atrophy is characterized by autonomic dysfunction, parkinsonian features, and cerebellar ataxia. This disorder can be categorized into MSA-P with predominant Parkinsonism and MSA-C with predominant cerebella features.[20] Atrophy of the transverse pontine fibers results in cruciate hyperintensity on MRI.[21] Dementia with Lewy body typically presents with cognitive impairment, visual hallucination, and parkinsonism.[22] The presence of visual hallucinations at the first visit is highly suggestive of DLB.[23]

Conclusion

Progressive Supranuclear palsy is a rare condition in our environment[10]. There are challenges to the use of MRI in low and Middle-Income countries (LMICs). These obstacles include the cost and ease of getting this investigation done. In the case presented, the patient had to travel to another region and pay a substantial sum to get the investigation done. The classical MRI[15] signs were seen in this patient as well as clinical[8] diagnosis but No pathological Evidence[12]. It is hoped that public health facilities will acquire the machines and make them readily available in the nearest future.

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