Frequent falls and gaze palsy in two patients

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Journal of the Ceylon College of Physicians, 2000, **33**, 48-49

Summary

We report two patients who presented with frequent falls, gaze palsy and other clinical features of Progressive Supranuclear Palsy and wish to highlight the importance of distinguishing this condition from idiopathic Parkinson's disease.

Introduction

In 1964 Steele et al called attention to a distinctive syndrome that they called Progressive Supranuclear Palsy (PSP)[1]. This is a degenerative disorder characterised pathologically by neuronal loss, gliosis and neurofibrillary tangles in the midbrain, pons, basal ganglia and dentate nuclei of the cerebellum[2]. It is an uncommon disorder with an estimated prevalence of 1% among cases diagnosed as Parkinson's disease in a community population and usually presents between the ages of 45 and 75 years[3]. Several sets of criteria have been proposed for the clinical diagnosis of PSP[4]. The core features are a progressive disorder of gait and balance with a supranuclear gaze palsy, bradykinesia, pseudobulbar palsy, memory impairment, rigidity affecting axial structures more often than appendicular structures, and little or no tremor[4]. Certain features such as cerebellar ataxia, alien limb phenomenon (in which the limb moves up and down, without the patient controlling it) and dysautonomia are evidence against the diagnosis[5]. The clinical course is generally progressive with aspiration and inanition leading to a fatal outcome within ten years. Early onset of falls, dysphagia and incontinence are associated with a shorter survival time; age at onset, sex, vertical gaze palsy or axial rigidity have no effect on survival (6).

Case Reports

Case 1

A 63 year old lady complained of frequent falls during the last year. She had difficulty in articulation, swallowing, getting up from a chair and initiating walking; and walked with an extended neck with reduced movements of her arms. She found it difficult in going down stairs as looking down was impaired, was becoming more forgetful and emotional labile. She had a 'quizzical or astonished' facial appearance, an upward gaze palsy, fragmentation of horizontal pursuit movements and axial rigidity. The mini-mental score was 24/30, with mainly recall deficits. There was no tremor, autonomic disturbances or cerebellar signs. All tendon reflexes and the jaw jerk were exaggerated. Routine biochemical and haematological investigations were normal. An MRI scan of her brain showed atrophic changes in the brain stem.

Case 2

A 59 year old man had frequent falls over the last two years, was very forgetful and found it difficult to articulate, swallow and start walking. He had an extended neck posture (Figure 1) with marked axial rigidity,

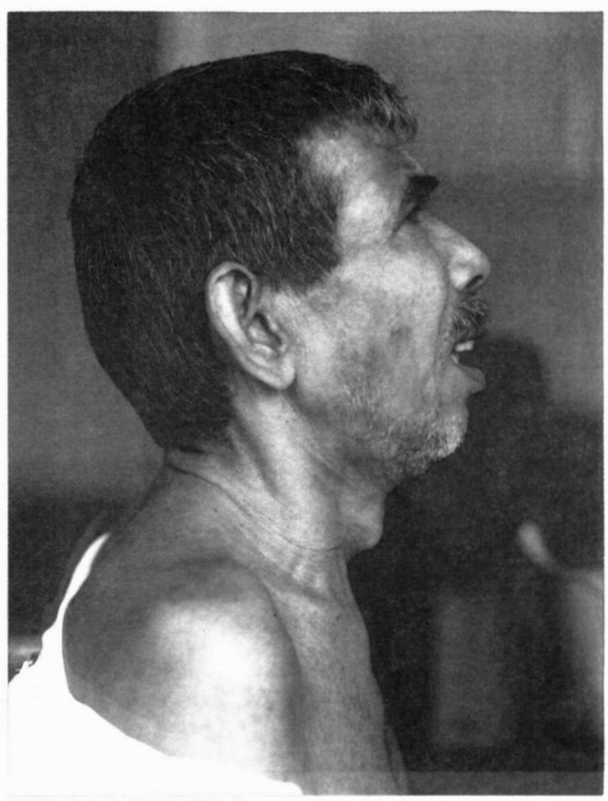

Figure 1. Side view of the patient showing the typical extended neck posture.

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inability to walk more than a few steps, a 'quizzical or astonished' facial appearance (Figure 2) and both an upward and downward gaze palsy. Horizontal eye movements were also greatly impaired. Emotional lability and globally exaggerated tendon reflexes were present. There was no tremor, autonomic dysfunction or cerebellar disturbance. Routine biochemical and haematological investigations were normal. A CT scan of his brain was consistent with brain stem atrophy.

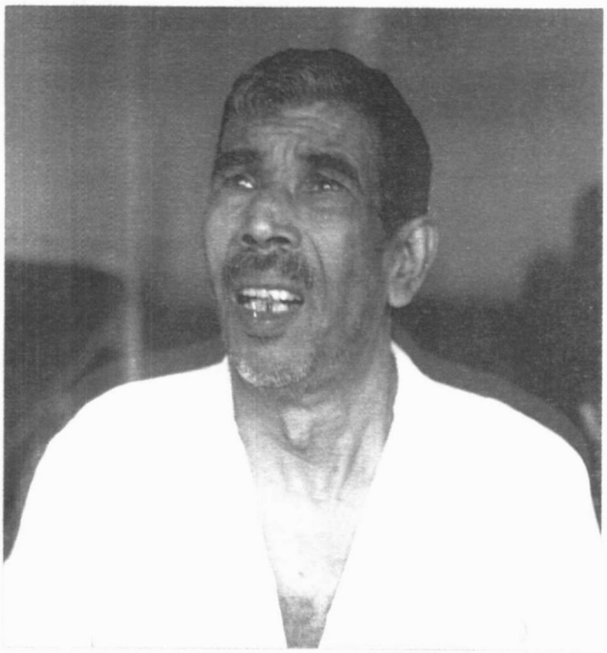

Figure 2. The 'quizzical or astonished' facial appearance which is characteristic of PSP.

Discussion

Both our patients met the classical clinical criteria for diagnosis of Progressive Supranuclear Palsy. This condition has not been reported previously in Sri Lanka, although a recent report on sixteen patients is available from India[7]. Both our patients had the 'full blown' clinical picture of this condition at presentation. However, several other conditions (Parkinson's disease, multi system atrophy, diffuse lewy body disease, corticobasal degeneration) need to be considered when a patient with an akinetic rigid syndrome is seen in the early stages. Certain clinical features help minimise the difficulties experienced when attempting to classify these disorders at this stage (8). Unstable gait, absence of tremor dominant disease and absence of a response to levodopa differentiates PSP from Parkinsons disease. Supranuclear vertical gaze palsy and increased age at symptom onset distinguishes PSP from multi system atrophy. Supranuclear vertical gaze palsy, gait instability and the absence of delusions distinguishes PSP from diffuse lewy body disease. Gait abnormality, severe upward gaze palsy, bilateral bradykinesia and absence of alien limb syndrome separates PSP from corticobasal degeneration.

Although PSP is most often clinically misdiagnosed as Parkinson's disease, the correct recognition of this condition is important as it indicates a poor prognosis and poor response to levodopa[3]. Therefore, this condition should be considered whenever a middle aged or elderly person presenting with repeated falls has extrapyramidal features, axial rigidity, and vertical gaze palsy[8].

References

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