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A RARE CASE OF STEELE-RICHARDSON-OLSZEWSKI SYNDROME.

Medicine

Dr. Parishree Kapadia* 3rd year resident, department of emergency medicine, GMC, Surat. *Corresponding Author

Dr. Grishma Modi 2nd year resident, department of emergency medicine, GMC, Surat.

Dr. P.B. Gupta Professor & Head, department of emergency medicine, GMC, Surat.

KEYWORDS

Introduction

Progressive means continuously increasing in severity and palsy means weakness by causing damage to parts of brain above nerve cells known as nuclei so supranuclear. Progressive supranuclear palsy(PSP) is a rare neurological disorder in which patient mostly comes with repeated history of fall and also affects movements, gait, speech swallowing, vision, mood, behavior and thinking. The progressive supranuclear palsy was first described as a distinct disorder from Parkinson's disease in 1964, when 3 scientists published a paper that distinguished the condition. It was referred to as the steele-Richardson-Olszewski syndrome reflecting the combined names of the scientist who defined the disorder.

Case report

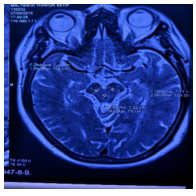
A 68 years old female patient came with complaints of History of recurrent falls progressed over 1 year Difficulty in downward gaze since 2 months Dysarthria since 1 month On examination, patient's vitals were stable.

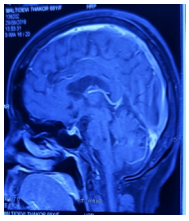
Clinical examination revealed: Loss of vertical and downward gazes Difficulties with convergence (convergence insufficiency), Infrequent blinking, Narrow gait Sway while turning to walk Action tremors on outstretched hands Bilateral brisk knee jerks and equivocal planters Her mini mental state examinations score was 25/30 showing mild cognitive impairment.

Investigations

All the lab parameters were within normal limits. Magnetic resonance imaging (MRI) of the brain showed mild prominence of cortical sulci, basal cisterns, subarachnoid spaces, sylvian fissure and cerebellar folia bilaterally with mild cerebral and cerebellar atrophy with Midsagittal images showed characteristic borderline atrophy of midbrain and superior cerebellar peduncles. This is a typical pattern of MRI in PSP. From clinical findings and MRI, the patient was diagnosed as PSP. She was started a combination of Levodopa (100 mg) with Carbidopa (25mg) 8 hourly and also given balance and gait training. Ophthalmic evaluation was done. She was discharged with advice of follow up after 1 month. On follow up, she came with similar condition.

Discussion





PSP is a neurodegenerative disorder that mainly affects cognition, eye movements and posture. The classical features of PSP is paresis of conjugate gaze. Interruption of the saccadic and pursuit pathways before they reach the eye movement generators results in loss of voluntary eye movements. In Parkinson's disease there is a loss of facial expression while there is an overreaction of the frontalis muscle giving “astonished facial expression” and movements of head in the respective direction to compensate for the weakness of conjugate eye movement in PSP. In Parkinson's disease there is a tendency to fall forward while in PSP, it is more often backward. Present case had history of recurrent falls as early symptoms.

Two main clinical subtypes have been described:

1) Richardson's syndrome in which early appearance of falls, absence of tremor, symmetry of signs, and poor response to levodopa is seen.
2) Progressive supranuclear palsy- parkinsonism, characterized by delayed onset of falls, presence of tremor, asymmetry, and response to levodopa.

The clinical features of Progressive supranuclear palsy more often overlaps with Parkinson's disease so it is usually referred as a form of atypical Parkinson plus syndromes. In Progressive supranuclear palsy, abnormal tau protein is present in nerve cells that are consistent with gait instability and falls, speech and swallowing difficulties early in the disease course, apathy, impaired abstract thought, decreased verbal fluency. MRI features of midbrain atrophy, eye signs, and falling history clinch the diagnosis.

The age of onset of this disease process is usually in the sixth and seventh decades (average age 63 years), and it advances more rapidly than Parkinson's disease, sometimes a family history is also present of autosomal dominant inheritance. Males and females are affected almost equally and there is no racial, geographical, or occupational predilection. The prevalence is age dependent and estimated at 6 to 10% of that of Parkinson's disease, or 6 to 7 cases per 1,00,000. Nearly half of all patients are markedly disabled or wheelchair bound within 4 years of onset. Early onset, the presence of falls, slowness, and inability to move the eyes downward early in the development of the disease predict poor survival time.

No definitive therapies are available, although physical therapy with balancing training and family education may be helpful. In the early stages, there may be some response to dopaminomimetic therapy, but none in later stages. Death, occurring in 2 to 12 years (the median survival is 9.5 years), is often due to the sequelae from falls or dysphagia (may cause aspiration pneumonia).

Conclusions

Progressive supranuclear palsy is one of the diseases collectively known as Parkinson plus syndrome. A case of a 68 years old female with progressive supranuclear palsy is presented. Clinical presentation and MRI brain features are described.

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