PALLIDO-PYRAMIDAL SYNDROME: A RARE ENTITY

Sir,

Pallido-pyramidal syndrome (PPS) is an unrecognized and a rare entity, which was reported for the first time by Davison in 1954.\(^1\) While about 18 patients have been reported in English literature, there are two reports from India.\(^2\) We describe here a 19-year-old male patient born to nonconsanguineous marriage, who presented with insidious onset gradually progressive illness of one and a half year duration. Illness started with complaints of difficulty in walking in the form of dragging of right lower limb, followed by abnormal movements of neck and change in voice, which became slow and monotonous. One year later, he developed postural tremors in both upper limbs along with abnormal posturing of the body. Patient also had peri-oral dyskinesias. He did not have any memory impairment. None of his family members had similar illness; or history of antipsychotic, antiemetic drug intake; or history of head injury or stroke.

Neurological examination revealed an alert, fully oriented patient. Memory and intellectual functions were normal with MMSE score of 27/30. Kayser-Fleischer ring was negative on slit lamp examination. Motor examination revealed asymmetric pyramidal weakness and cogwheel rigidity in all four limbs. His serum ceruloplasmin level, vasculitic profile and serum lactate were normal. MRI brain and EEG were also normal. Single photon emission tomography (SPECT) using Tc99mECD (ethylene cysteine dimmer) revealed hypoperfusion in left fronto-parietal, left basal ganglia and minimal hypoperfusion in left temporal lobe as shown in Figure 1.

In view of the above clinical picture, various possibilities included juvenile parkinsonian disease, Wilson disease, juvenile Huntington disease, Pallido-nigrolyus degeneration, Hallervorden-Spatz and Kufor-Rakeb syndrome. The absence of KF ring and normal serum ceruloplasmin level ruled out the possibility of Wilson disease, while the presence of pyramidal signs was against the diagnosis of both young onset Parkinson's disease and Pallido-nigrolyus degeneration. Hallervorden-Spatz disease was unlikely in the absence of cognitive impairment and a normal MRI brain. Similarly juvenile Huntington disease was not considered due to negative family history and excellent response to levodopa.

![Image](https://example.com/image.png)

*Figure 1: Tc99mECD SPECT study of patient shows hypoperfusion of the left basal ganglia and left fronto-parietal area.*
Our patient’s clinical profile was suggestive of Pallido-pyramidal syndrome, a rare entity. The PPS manifests between 7 and 24 years of age and is suspected to have autosomal recessive mode of inheritance. [3] There is involvement predominantly of extrapyramidal and pyramidal systems and sometimes of cerebellum. Bradykinesia, cogwheel rigidity are seen in almost all patients. Tremors, hypophonia, stooped posture, scissoring gait, hyperreflexia and Babinski sign may be present. The symptoms respond well to combination of anticholinergic and levodopa.[3] So the presumptive diagnosis of Pallido-pyramidal disease was made; it was confirmed by SPECT study, which revealed left-sided basal ganglia and fronto-parietal hypoperfusion, which may be due to degeneration of striatal neurons. The fronto-parietal hypoperfusion due to subcortical lesion has been attributed to cerebral diaschisis. The patient was put on Trihexphenydil in dosage of 2 mg twice daily along with levodopa in a dose of 125 mg four times daily, and he showed marked improvement in motor functions, including tremor, akinesia and cogwheeling, and started walking without support.

Thus although rare, this syndrome must be considered in differential diagnosis of every young patient presenting with extrapyramidal and pyramidal signs, since partial amelioration of symptoms can improve quality of life.

REFERENCES


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