IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 17, Issue 2 Ver. 4 February. (2018), PP 18-19
www.iosrjournals.org

A Prospective Study on Clinical Variants of Progressive Supranuclear Palsy

Dr.N.Shobana[1], Dr.V.Ushapadmini[2]
(Associate professor in Neurology,Coimbatore medical college,The TN DR.M.G.R Medical University,India,)
Associate Professor in Medicine, KAPV GovtMedical College Trichy,The TN DR.M.G.R.Medical University,India)
Corresponding author: Dr.V.Ushapadmini M.D.,

Abstract: *PSP-Progressive supranuclear palsy is a clinical syndrome comprising of supranuclear palsy,postural instability and mild dementia. Since the first description of PSP several distinct clinical syndromes have been described.The aim of our study was to find out the prevalence of these clinical subtypes and their clinical course among patients with PSP(10 Italic)*

Keywords: *PSP, tau protein, PSP-P,PAGF,PSP-CBS,PSP-PNFA*

Date of Submission: 25-01-2018 Date of acceptance: 13-02-2018

I. Introduction

In 1963J.Clifford Richardson described the clinical syndrome of PSP comprising of postural instability,Supranuclear gaze palsy,mild dementia,progressive axial rigidity and bulbar palsy.His collaborators identified pathological findings. Pathologically PSP is defined by the accumulation of tau protein and neuropil threads in the pallidum,subthalamic nucleus,Red nucleus,Substantia nigra,Pontine tegmentum,Striatum,Oculomotor nucleus, Medulla and Dentate nucleus.There are no reliable diagnostic Biomarkers for PSP and accurate diagnosis depends on clinical acumen.

CLINICAL VARIANTS

The variants can be separated by differences. in their severity,regions of pathology,clinical features and are linked by the accumulation of neurofibrillary tanglesand similar natural histories that lead to death within 6-12 years.The classical PSP is referred to as the Richardson syndrome(Steele-Richardson-Olszewski syndrome)The variants are 1)PSP-P Parkinsonism 2)PSP-PAGF-pure akinesia with gait freezing 3)PSP-CBS Corticobasal syndrome 4)PSP with speech apraxia-PNFA Progressine nonfluent aphasia.The early clinical features are subtle and diagnosis delayed for many months. The National Institute for Neurological disorders and stroke(NINDS) PSP criteria are most helpful defining clinical features. They are early falls due to postural instabilityand supranuclear palsy.Richardson syndrome is characterized by lurching gait,Unexplained falls backwards,Cognitive decline first 2yrs. Vertical supranuclear gaze palsy is the definitive diagnostic feature.Apraxia of lid opening,Slowed slurred speech and Difficulty in swallowing are typical early features.Overactivity of the frontalis,procerus cause lid retraction and staring gaze,Median survival is 5-8 yrs.P-P is characterized by normal eye movements,resting tremor,positive levodopa response and focal dementia.Although axial rigidity was the striking early feature limb rigidity was more common and severe than in Richardson syndrome.A Jerky postural tremor was common in patients with PSP-P.PSP-P and Richardson syndrome can be distinguished in the first 2years but there is clinical overlap after 6 years.PSP-PAGF is characterized by progressive onset of gait disturbance,with start hesitation and subsequent freezing of gait.speech or writing without rigidity,tremor,dementia or eye movement abnormality during the first 5years of the disease.There is no benefit with levodopa therapy and no clinical or radiological evidence of lacunar infarcts or diffuse deep white matter ischemia.PSP-CBS is characterized by progressive asymmetric dyspraxia,cortical sensory loss including an Alien limb,Jerky dystonia of the limb with rigidity and bradykinesia that is unresponsive to levodopa.PNFA is characterized by non-fluent spontaneous speech with hesitancy,Agrammatism and Phonemic Errors.There is Apraxia of speech which describes errors in timing,Coordination and initiation of sppech that are secondary to disorders of motor command.

II. Materials And Methods

About 20 aptients with PSP were progressively followed up for 2 years from January 2015 to January 2017 at the Neurology Department, Coimbatore Medical College Hospital.The National Institute of Neurological Disorders and stroke(NINDS) criteria was used to identify patients with PSP.A detailed

neurological examination was done and 10 important components of extrapyramidal system were assessed namely 10Rigidity 2)Bradykinesia 3)Tremor 4)Early Falls 5)Early postural instability 6)Early cognitive decline 7)Early abnormalities of eye movement 8)Response to levodopa 9)Hyposmia 10)Speech.MRI Brain was done to exclude patients with Lacunar Infarcts or Diffuse deep white matter ischemia(Vascular Parkinsonism).

III. Results And Discussion

All the 20 patients diagnosed to have PSP and variants were Males.The average age at presentation was between 55-65 years.Of the 20 patients 14(70%) were diagnosed to have Classic PSP .They showed poor response to levodopa and Rapid progression of the disease.The median duration of symptoms at presentation was between 1-3 years.four patients(20%) presented with PSP-P features.Asymmetrical tremor and Axial rigidity were prominent clinical features.After the moderate response to levodopa in the initial 1-2 yrs they showed progression of symptoms after 2 yrs.1 patient (5%) had PSP-PAGF.Freezing of gait and speech without rigidity or tremors were presenting features.They showed poor response to levodopa and no radiological evidence of white matter ischemia.One patient(5%) had PNFA with prominent apraxia of speech.No patient had PSP-CBS.The clinical heterogeneity could be explained byPathological heterogeneity of tau protein deposition in the brain through postmortem studies.The frontal cortex,Subthalamic nucleus and substantia nigra were more severely affected in Classic PSP.This explains the levodops unresponsivenesss and cognitive dysfunction in Classic PSP.PSP-P patients have less severe tau pathology in the motor cortex,striatum,pontine nuclei and cerebellum.PSP-PNFA had more severe pathology in the temporal cortex and superior frontal gyrus than brainstem and subcortical grey matter regions.(10)

IV. Conclusion

The Classic PSP is still the most prevalent.The clinical variables namely PSP-P,PSP-PAGF,PSP-PNFA and PSP-CBS are increasingly being recognizedNevertheless these clinical subtypes can be included under the generic term PSP owing to clinical overlap and progressive course over time.The regional differences in pathological severity account for the clinical Variability.

Acknowledgements

All patients with PSP and their family members.

References

[1]. HauwJJ,Daniel SF,Dickson D et alPreliminary NINDS neuropathologic criteria for Steele Richardson Olszewski Syndrome-Neurology 1994,44;2015-2019
[2]. Williams DR.Lees AJ,Wheeeeeeerrett JR,SteeleJC,J.Clifford Richardson and 50 years ofsupranuclearpalsy,Neurology008;70;566-73
[3]. Williams DR,HoldenJL,StrandK.Lees AJ.Pure akinesia with gait freezing;A third clinical phenotype of progressive supranuclear palsy Movement disorder 2007;22;2235-41
[4]. Ahmed Z,Josephs KA,Gonzles J,Delle Donne A,Dickson DW;clinical and neuropathological features of supranuclear palsy with severe pallid nigro luysial degeneration and axonal dystrophy,Brain 2008;131;460-72
[5]. Joseph KA,BoeveBF,Duffy JR,SmithGE,KnopmanDS,Parisi JE,etal-Atypical Progressive supranuclear palsy underlying progressive apraxia of speech and nonfluent aphasia,Neurocase 2005;11;283-96
[6]. WilliamsDR,HoltonJ,Strand C etal-pathologica tau burden and distribution distinguishes progressive supranuclear palsy parkinsonism from Riichardson syndrome,Brain2007;130;1566-76
[7]. Jellinger KA,Different tau pathology pattern in two clinical phenotypes of progressive supranuclear palsy,Neuro degenerative dis 2008;5;339-46

Examples follow:
Journal Papers:
[8]. M Ozaki, Y. Adachi, Y. Iwahori, and N. Ishii, Application of fuzzy theory to writer recognition of Chinese characters, *International Journal of Modelling and Simulation, 18(2),* 1998, 112-116.
Books:
[9]. R.E. Moore, *Interval analysis* (Englewood Cliffs, NJ: Prentice-Hall, 1966).
Chapters in Books:
[10]. P.O. Bishop, Neurophysiology of binocular vision, in J.Houseman (Ed.), *Handbook of physiology,* 4 (New York: Springer-Verlag, 1970) 342-366.
Theses:
[11]. D.S. Chan, *Theory and implementation of multidimensional discrete systems for signal processing*, doctoral diss., Massachusetts Institute of Technology, Cambridge, MA, 1978.
Proceedings Papers:
[12]. W.J. Book, Modelling design and control of flexible manipulator arms: A tutorial review, *Proc. 29th IEEE Conf. on Decision and Control*, San Francisco, CA, 1990, 500-506

Dr.V.Ushapadmini M.D., "A Prospective Study on Clinical Variants of Progressive Supranuclear Palsy."." IOSR Journal of Dental and Medical Sciences (IOSR-JDMS), vol. 17,