



Multiple System Atrophy MSA

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It is a Neurodegenerative Disorder

Types

- 1 Shy -Drager Syndrome
- 2 Olivopontocerebellar Atrophy
- 3 Striatonigral Degeneration

History

Described by Milton Shy and Glen Drager in 1960

Incidence

3 to 5 per 100,000 population
USA has 15 000 to 50 000 patients
All races
In Japanese there is a genetic abnormality
SHC3 gene deletion

This genetic abnormality is not seen in the American patients

Age

Average 50 to 59 years
Can start at 30 y

Sex

Males 55%

Course

Lasts for 7 to 10 years
Few patients last 12 years
Very few last 15 years

Pathology

Alpha Synnuclein protein is deposited in the neurons and glia

Types

MSA C Cerebellum

MSA. P. Parkinsonism

Organs involved

Basal Ganglia

Cerebellum

Brain Stem - Autonomic Fn

Breathing. Heart rate. BP

Clinical

Tremors ,Parkinsonism

Ataxia

Postural hypotension

Fainting

Fall

Urinary incontinence

Fecal incontinence

Constipation

Sexual Dysfunction

REM Sleep disturbed

Sweating decreased

Dry mouth

Visual problems

Sleep apnea

Cognitive and emotional disturbances

Anxiety ,depression emotional instability

Laughing and crying

Panic attacks

Suicide

Self harm

Ataxia

Tremors

Wide steps while walking

Eyes nystagmus

Stiffness, rigidity

Slurred speech

Diagnosis

1 clinical

2 CT

3 MRI

4 PET CT

5 Dopamine Transporter scan

6 skin biopsy

7 genetic analysis

Japanese have SHC3 deletion

Treatment
No cure
L Dopa poor response

Symptomatic treatment
Hospice
Palliative care
Walk with a cane or walker
Wheelchair

Cause of death
Infection
UTI
Sepsis
Sudden death
Aspiration Pneumonia
Cachexia

Mental health
Physical health

May need
Tracheostomy
Feeding tube
Indwelling catheter
Colostomy

(Dr. R.G. Wiseman Pinto is a Professor of Pathology, former Head of Department at Goa Medical College, former Dean of Goa University, and the current President of the Asian Society of Cytopathology.)