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MULTIPLE SYSTEM ATROPHY: CASE OF ATYPICALPARKINSON WITH PARKINSON PREDOMINANT PRESENTATION AND PROGRESSIVE CEREBELLAR DEGENERATION WITH EXTRAPYRAMIDAL AND AUTONOMIC INVOLVEMENT

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MULTIPLE SYSTEM ATROPHY: CASE OF ATYPICALPARKINSON WITH PARKINSON PREDOMINANT PRESENTATION AND PROGRESSIVE CEREBELLAR DEGENERATION WITH EXTRAPYRAMIDAL AND AUTONOMIC INVOLVEMENT

Abstract

ABSTRACT

BACKGROUND

Multiple System Atrophy is a rapidly progressive neurodegenerative disease. Symptoms tend to appear in 50s and advance rapidly over the course of 5-10 years with progressive loss of motor and autonomic function.

Some of the common symptoms include frequent falls with or without tremors and b/l akinetic rigidity with involvement of the axial muscles. Postural tremor, severe stridor and cranio cervical dystonia may be seen. Alpha synnuclenopathies are generally characterized by REM behavioral disorders and severe dysautonomia years before the onset of the disease.

Individuals with MSA typically do not have sustained improvement in their symptoms with levodopa.

Currently, there are no treatments to delay the progressive neurodegeneration in MSA and there is no cure. However symptomatic treatment like drugs to improve orthostatic hypotension or anticholinergics for bladder control or compression stockings to overcome fainting can be adopted.[1]

CASE DESCRIPTION

A 60 year old male patient with known history of type 2 DM and HTN presented to JSS hospital with history of difficulty in speaking, walking and multiple episodes of falls, giddiness and involuntary movements of the hand and loss of bladder control progressing over a period of two years which was confirmed to be a case of Multiple System Atrophy (MSA-P) with progressive cerebellar degeneration, extrapyramidal and autonomic involvement.

CONCLUSION

The case report presents a progressive neurodegenerative condition which is a type of Atypical Parkinson with difficult diagnosis in the early stages due to its presenting complaints being very similar to Parkinson itself but progresses more rapidly.

Keywords

MSA, ATYPICAL PARKINSON, PARKINSON PLUS SYNDROME, SHY DRAGER SYNDROME, MSA-P

MULTIPLE SYSTEM ATROPHY: CASE OF ATYPICAL PARKINSON WITH PARKINSON PREDOMINANT PRESENTATION AND PROGRESSIVE CEREBELLER DEGENERATION WITH EXTRAPYRAMIDAL AND AUTONOMIC INVOLVEMENT

AAYUSHI SHARMA

CASE

CHIEF COMPLAINTS

A 60 year old male patient who hails from Mandya and is a farmer by occupation was brought to the hospital by his wife with the chief complaint of

1) Difficulty in speaking since 2 years
2) Difficulty in walking since 1 year
3) Giddiness with multiple falls since 4 months
4) Involuntary movements of the head since 3months
5) Loss of control of urination since 2 months

HISTORY OF PRESENTING ILLNESS

The patient who is a known case of HTN and type 2 DM since 8 years and is complaint to the medications, was apparently in his usual state of health 2 years ago when he developed sudden onset of difficulty in speaking while he was working, in the form of slurring of speech. During this episode patient did not have weakness in any limb.

No h/o difficulty in swallowing or no deviation of angle of mouth present.

Attenders gave history that they could understand the speech for the following 1 year after which the slurring progressed and attenders started having difficulty in understanding. Attenders noticed no tendency to repeat his own or others speech.

Patient also complains of difficulty in walking for the last 1 year. Earlier he was able to walk for 3-4kms per day. Since the last 1 year patient finds it difficult to carry out his daily activities.

no history of weakness in both upper and lower limbs.

Since the last 4 months attenders noticed that the patient is taking multiple small steps while walking and there is reduced arm swinging.

History of multiple episodes of falling with preserved consciousness for last 4 months.

Giddiness present since the past 4 months mostly while walking and it lasts around 1-2 minutes and is associated with prior falls. She noticed that he also had those movements while he was trying to hold an object.

h/o loss of control in passing urine since 2 months. Earlier patient was able to hold his urine and go to the washroom. Then his inability to hold urine when increased urgency,progressed and he was micturating mid way on his way to the washroom.

Attenders also gave history of impaired bowel habits since last 2 months.

PAST HISTORY

No significant history present.

PERSONAL HISTORY

Patient is predominantly vegetarian.

Sleep normal

Appetite normal

GENERAL PHYSICAL EXAMINATION

A male patient, moderately built and nourished is conscious, cooperative and oriented to time,place and person.

Pallor, Icterus, Cynosis, Clubbing, Lymphedema- absent

PR- 85BPM

BP- 140/80 mm hg

SYSTEMIC EXAMINATION

CENTRAL NERVOUS SYSTEM EXAMINATION

CRANIAL NERVES

Cranial nerve examination is normal.

MOTOR SYSTEM

		RIGHT	LEFT
BULK	Arm	31.5cm	29.5 cm
	Forearm	26.5cm	26cm
	Thigh	36cm	35cm
	Calf	30cm	30cm

TONE	UPPER LIMB		
	Flexion	rigidity	cogwheel rigidity
	Extension	rigidity	cogwheel rigidity
	LOWER LIMB		
	Flexion	rigidity	cogwheel rigidity
	Extension	rigidity	cogwheel rigidity

POWER 5/5 in all 4 limbs

REFLEXES	Biceps	2+	2+
	Triceps	2+	2+
	Knee	1+	1+
	Ankle	absent	absent
	Plantar	b/l withdrawl present	

Superficial reflexes are normal.

SENSORY- normal

Hyperesthesia present in both foot while performing plantar

CEREBELLUM

Gait- broad base short stepping gait present with ataxia

Speech- staccato /explosive speech present

Nystagmus- left side fast component present

Resting tremors- absent

Finger nose test-dysmetria present, tremor while reaching finger (left>right)

Rebound- absent

Dysdiadochokinesia present

OTHER SYSTEMS

CVS- S1 S2 present, no murmurs

RS- B/L NVBS present

P/A- soft, no organomegaly

INVESTIGATIONS

Making a diagnosis of MSA can be difficult particularly in the early stages mostly because many of the features are similar to those in Parkinsons. Apart from taking a clinical history and performing neurological examination, assessment of autonomic functions like bladder control, blood pressure fluctuations and heart rate control can be done. MRI of the brain may identify changes which suggest MSA or rule out other causes of the observed symptoms. PET scan is sometimes used to see if the metabolic function of the brain is reduced in specific parts.[1] Individuals with MSA typically do not have a sustained improvement in their symptoms with levodopa, a finding that supports the diagnosis of MSA.[1]

FINAL DIAGNOSIS

Parkinson type of Multiple system atrophy (MSA-P) i.e Atypical Parkinson with progressive cerebellar degeneration with extrapyramidal and autonomic involvement. Peripheral neuropathy present secondary to untreated DM and HTN.

DISCUSSION

Multiple System Atrophy is one of the Parkinson plus syndrome also known as Atypical Parkinson. The aim is to distinguish it from Parkinson as the former is mostly unresponsive to L-dopa and other drugs given in Parkinson.

MSA is an alpha synnucleinopathy, i.e accumulation of intraneuronal cytoplasmic inclusions called as Lewy Body, in the substantia nigra, brainstem and limbic cortex.

There are three types of MSA's

1)the Parkinson type (MSA-P) with primary characteristics similar to Parkinson disease.

2)the cerebellar type (MSA-C) with cerebellar predominant symptoms.

3) the autonomic type(MSA-A) with severe dystonia especially orthostatic hypotension and erectile dysfunction called the Shy Drager Syndrome.

Alpha synnucleinopathies are characterized by REM behavioral disorders as well as ANS problems years before the onset of the disease.

MSA is suspected in cases of absent tremors or b/l symmetrical akinetic rigidity with involvement of axial muscles on presentation. Postural tremors, severe stridor and cranio cervical dystonia may be seen.

Mostly clinical diagnosis is made . MRI- T2W may shows putamen rim or cruciform hyper intensity.

There is no treatment available to cure or slow the progression of the disease but supportive care can be provided.

CONCLUSION

MSA being a rapidly progressive neurodegenerative disorder tends to appear in a person's 50s and advances rapidly over the course of 5-10 years with progressive loss of motor function and eventual confinement to the bed.[1] After making a clinical diagnosis and ruling out Parkinsons, symptomatic treatment and supportive care should be provided to the patient.

ACKNOWLEDGEMENTS

None

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