

Start	End	Topic	Speakers
13:00	13:10	introduction	Ryuji Sakakibara
13:10	13:40	what is MSA, including pathology, clinical	Jalesh Panicker
13:40	14:10	urodynamics	Tatsuya Yamamoto
14:10	14:30	management	Tomoyuki Uchiyama

Description

Background information

Multiple system atrophy (MSA) is a major neurodegenerative disease characterized pathologically by abnormal alpha-synuclein (SNCA) aggregation. In MSA, the SNCA aggregation appears in the oligodendroglia, which are called 'glial cytoplasmic inclusions (GCIs)' as detected by SNCA positron emission tomography (PET) and the measurement of SNCA in the cerebrospinal fluid. The common clinical features of MSA are pelvic autonomic dysfunction (urinary retention with urge incontinence), systemic postural hypotension in some, and parkinsonian/ cerebellar motor disorder.

Although MSA shows a Parkinson's disease (PD)-like motor disorder, rest tremor and laterality are rare, levodopa's effect is not sufficient, and cerebellar ataxia often co-exists. This is because MSA has more widespread central nervous system (CNS) pathology compared to PD. In Asian countries including Japan, the ataxic form of MSA (MSA-C) is more prevalent than the parkinsonian form (MSA-P). This is in contrast to the fact that in European countries, MSA-P is more prevalent than MSA-C. Therefore, clinically MSA should be differentiated among degenerative parkinsonism and cerebellar ataxias.

Sometimes MSA shows PD-like bladder dysfunction, but there is a profound difference between these MSA and PD. Large post-void residual/urinary retention is common in MSA but rare in PD, presumably reflecting MSA's more widespread CNS pathology including the spinal cord. Bladder dysfunction not only impairs an individual's quality of life; it can also cause emergency hospitalization due to retention, and early institutionalization. In this workshop, we summarize the epidemiology, pathophysiology, and management of bladder dysfunction in individuals with MSA.

Key learning points

Pelvic autonomic dysfunction appears as the sole, initial symptom in 18% of the patients. Such patients later develop motor disorder, therefore is referred to as 'double-hit disease'. This reflects complex MSA pathology, including sacral spinal cord for pelvic autonomic dysfunction, and brain pathology for the motor disorder. Continuous care for the patients' pelvic autonomic dysfunction is necessary, by assessing with urodynamics, sphincter EMG, and teaching clean, intermittent self-catheterization (CIC).

Urodynamic features of MSA include so-called DHIC (detrusor hyperactivity/overactivity during bladder filling and impaired contraction/detrusor underactivity during voiding), with detrusor sphincter dyssynergia in some patients. Sphincter electromyogram (EMG) in MSA is pathognomonic, showing neurogenic changes reflecting lesion in the sacral Onuf's nucleus. CIC is a recommended method to manage patients' large post-void residuals (PVR)/ urinary retention. Many patients can continue CIC with care by specialized nurses and doctors.

Take home messages

MSA is sometimes a difficult disease to diagnose initially; however, if we look for the correct diagnosis along with proper tests and care of the patients' bladder, we can maximize the quality of life of MSA patients. Ancient Sophocles tells us: Who seek shall find.

Additional References

The Movement Disorder Society Criteria for the Diagnosis of Multiple System Atrophy.

Wenning GK, Stankovic I, Vignatelli L, Fanciulli A, Calandra-Buonaura G, Seppi K, Palma JA, Meissner WG, Krismer F, Berg D, Cortelli P, Freeman R, Halliday G, Höglinger G, Lang A, Ling H, Litvan I, Low P, Miki Y, Panicker J, Pellecchia MT, Quinn N, Sakakibara R, Stamelou M, Tolosa E, Tsuji S, Warner T, Poewe W, Kaufmann H. *Mov Disord.* 2022 Jun;37(6):1131-1148. doi: 10.1002/mds.29005. Epub 2022 Apr 21. PMID: 35445419

Early presentation of urinary retention in multiple system atrophy: can the disease begin in the sacral spinal cord?

Panicker JN, Simeoni S, Miki Y, Batla A, Iodice V, Holton JL, Sakakibara R, Warner TT. *J Neurol.* 2020 Mar;267(3):659-664. doi: 10.1007/s00415-019-09597-2. Epub 2019 Nov 12. PMID: 31720822

Urological dysfunction in synucleinopathies: epidemiology, pathophysiology and management.

Sakakibara R, Tateno F, Yamamoto T, Uchiyama T, Yamanishi T. *Clin Auton Res.* 2018 Feb;28(1):83-101. doi: 10.1007/s10286-017-0480-0. Epub 2017 Nov 9. PMID: 29124503

Aims of Workshop

Multiple system atrophy (MSA) is a degenerative neurological disease. MSA comprises pelvic autonomic dysfunction (urinary retention with urge incontinence), systemic postural hypotension in some, and parkinsonian/ cerebellar motor disorder. Pelvic autonomic dysfunction appears as the sole, initial symptom in 18% of the patients. Such patients later develop motor disorder, therefore is referred to as 'double-hit disease'. This reflects complex MSA pathology, including sacral spinal cord for pelvic autonomic dysfunction, and brain pathology for the motor disorder. Continuous care for the patients' pelvic autonomic dysfunction is necessary, by assessing with urodynamics, sphincter EMG, and teaching clean, intermittent self-catheterization.

Educational Objectives

Multiple system atrophy (MSA) is a degenerative neurological disease. MSA comprises pelvic autonomic dysfunction (urinary retention with urge incontinence), systemic postural hypotension in some, and parkinsonian/ cerebellar motor disorder. Pelvic autonomic dysfunction appears as the sole, initial symptom in 18% of the patients. Such patients later develop motor disorder, therefore is referred to as 'double-hit disease'. This reflects complex MSA pathology, including sacral spinal cord for pelvic autonomic dysfunction, and brain pathology for the motor disorder. Continuous care for the patients' pelvic autonomic dysfunction is necessary, by assessing with urodynamics, sphincter EMG, and teaching clean, intermittent self-catheterization.

MSA is one of the representative diseases that causes neurogenic pelvic organ dysfunction. Also, MSA is one of difficult neurological diseases that present with bladder dysfunction alone initially. Therefore, understanding MSA and its management is important, and it can be applied to other diseases that neurourologists may encounter. This Neurology Workshop will provide skills to approach patients by both urologically and neurologically.

Learning Objectives

1. Able to know what is multiple system atrophy (MSA)
2. Able to know what are the background pathology and key clinical issues in MSA
3. Able to know how to manage MSA patients

Target Audience

Urology, Urogynaecology and Female & Functional Urology

Advanced/Basic

Intermediate

Suggested Learning before Workshop Attendance

A recent diagnostic criteria of this disease is published:

The Movement Disorder Society Criteria for the Diagnosis of Multiple System Atrophy.

Wenning GK, Stankovic I, Vignatelli L, Fanciulli A, Calandra-Buonaura G, Seppi K, Palma JA, Meissner WG, Krismer F, Berg D, Cortelli P, Freeman R, Halliday G, Höglinger G, Lang A, Ling H, Litvan I, Low P, Miki Y, Panicker J, Pellecchia MT, Quinn N, Sakakibara R, Stamelou M, Tolosa E, Tsuji S, Warner T, Poewe W, Kaufmann H. *Mov Disord.* 2022 Jun;37(6):1131-1148. doi: 10.1002/mds.29005. Epub 2022 Apr 21. PMID: 35445419