



# Urinary retention as first manifestation of probable MSA

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## INTRODUCTION

- Multiple System Atrophy (MSA) is a rare progressive, fatal neurodegenerative disorder with clinical features including parkinsonism, cerebellar ataxia, cardiovascular dysautonomia and urogenital dysfunction (1)
- Urinary incontinence due to an overactive bladder and erectile dysfunction are commonly reported, and may precede the occurrence of motor or cardiovascular autonomic symptoms (2)
- Progressive incomplete bladder emptying and urinary retention usually develop during the course of disease
- We present six cases presenting initially with urinary retention with very little neurological signs, who subsequently developed classical neurological signs of MSA over time

## DISCUSSION

At the time of initial presentation, these 6 patients presented predominantly with urinary retention and bowel and sexual dysfunction were early accompanying features.

Structural urological causes such as bladder outflow obstruction were excluded. Urinary retention in the setting of MSA occurs as a result of neurodegenerative changes affecting regions involved in the neural control of voiding such as the brainstem and sacral spinal cord. There were no signs suggesting brainstem dysfunction at the time of initial presentation. Moreover, the anal sphincter EMG was grossly abnormal, thereby suggesting the lesion to be at the level of the sacral spinal cord. In the sacral spinal cord, MSA-related degeneration can affect the Onuf's nucleus and bladder motor neurons and recently, high resolution MRI sequences of the lumbosacral spinal cord have demonstrated atrophy in the grey matter in patients with MSA (3).

It is therefore likely that this cohort of patients presenting to urology services for urinary retention had a sacral spinal cord-onset of disease. They subsequently developed the classical motor and autonomic signs of MSA. The pathology is known to propagate in the central nervous system with recent evidence suggesting transmissibility of alpha-synuclein in animal models following inoculation of tissue from MSA patients (4), therefore the possibility exists that the pathology might have spread to other regions of the spinal cord and brainstem, causing the classical motor and autonomic symptoms on follow up. Pathological studies would be required to explore this hypothesis further.

## CASE SERIES

Between 2012-2018, six patients (mean age 55 years SD 10,3 years, 2 females) presented to a tertiary-level specialist Uro-Neurology unit with urinary retention who subsequently developed classical signs of MSA.

At initial presentation, 1 was able to void, and 5 were in urinary retention and catheter-dependent (table).

No cause for urinary retention could be identified during urological investigations, and urodynamics studies showed evidence for detrusor underactivity in 3 patients.

Concentric needle EMG of the anal sphincter performed in 5 patients to evaluate the sacral root (S2,3,4) innervation was however abnormal.

5 patients had sexual dysfunction and 4 had bowel dysfunction.

Subtle neurological signs were noted at presentation in 5 patients, however these were insufficient to establish a neurological diagnosis.

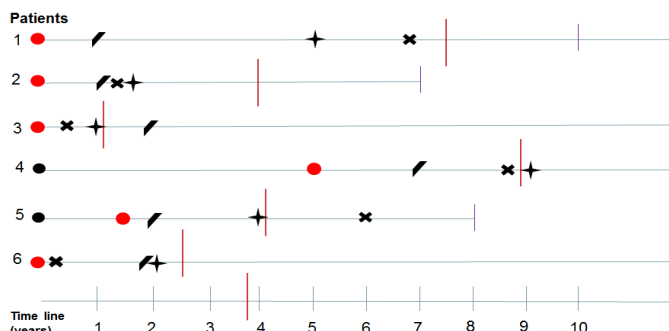
During follow-up, all six patients developed motor and autonomic symptoms/ signs meeting the diagnostic criteria of MSA over a mean period of 4.6 years.

Three patients expired (mean duration of illness 7.67 years). Urinary retention was managed by clean intermittent catheterisation initially, however indwelling catheterisation was required over time as neurological disability progressed.

Patient	(gender/ age at presentation to the Uro-Neurology Unit)	Duration of urinary retention (years)	Duration of catheterisation (years)	Anal sphincter EMG	Duration of sexual dysfunction (years)	Duration of bowel dysfunction (years)	Interval between urinary retention and onset of neurological signs (years)	Interval between urinary retention and diagnosis of MSA (years)
1)	M 37*	5	3	abnormal	5	3	7	8
2)	M 60*	4	3	abnormal	4	/	2	3
3)	M 61	3	3	not performed	10	/	2	2
4)	F 56	2	1	abnormal	/	5	2	3
5)	F 66*	3	3	abnormal	/	2	3	5
6)	M 50	3	3	abnormal	3	3	1	1

\*deceased

Symptoms duration at the time of presentation to the Uro-Neurology Unit.



Interval between urinary retention and Neurology referral 3.6 years (1 year- 7.5 years)!

Timeline of symptoms and disease progression.

## CONCLUSIONS

- In a subset of patients, urinary retention may be the initial presenting feature of MSA
- At the time of presentation to urology services patients may manifest with very little neurological signs.
- The presence of concomitant sexual and bowel dysfunction and abnormal anal sphincter EMG betray a neurological cause for urinary retention.
- Based on the pattern of urological and neurological dysfunction, we postulate the onset of this fatal disease to be in the sacral spinal cord in this cohort of patients with MSA.

## REFERENCES

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