EMERGENCIES IN MULTISYSTEM ATROPHY

VENI ALAGAMUTHU
Department of Neurology,
MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL

Abstract:
Background MSA is a neurodegenerative disorder associated with Parkinsonism, cerebellar ataxia, and autonomic dysfunction. Pituitary apoplexy is an endocrine emergency. Association between these two has not been not described so far. The occurrence of stridor and sleep apnoea in patients with MSA is associated with decreased life expectancy. Cases We present 2 cases of emergencies in multisystem atrophy. 1. Pituitary apoplexy in Multi system atrophy-parkinsonism type (MSA-P) 2. Nocturnal stridor in Multi system atrophy-cerebellar type (MSA-C) Conclusion Pituitary apoplexy in Multi system atrophy-parkinsonism type is presented for its unusual association. Hypotension due to dysautonomia is the cause for pituitary apoplexy. Polysomnography and laryngoscopy are the important diagnostic tools for nocturnal stridor in MSA. Bilateral vocal cord paresis (or) dystonia is the cause for stridor and respiratory compromise. Permanent tracheostomy is the treatment option for this patient.

Keyword: MSA-P Multisystem atrophy parkinsonism type, MSA-C Multisystem atrophy Cerebellar type

Introduction:
Multisystem atrophy is a sporadic and progressive neurodegenerative disorder characterized by neuronal loss in multiple areas of the CNS including nigrostriatal system, cerebellum, inferior olives, intermediolateral column of spinal cord. (1) The disease is usually classified into 2 major motor presentation MSA-P Parkinsonism features predominates in 80% of MSA, whereas cerebellar ataxia (MSA-C) is the presenting feature in about 20%. The other recognised symptoms of multiple system atrophy are dysarthria, dysphonia, dysphagia, stridor, snoring, vocal cord palsy, obstructive sleep apnoea, dystonia, postural instability, absent or atypical levodopa-induced dyskinesia and Raynaud's phenomenon Respiratory emergency secondary to movement disorders are a rare but potentially life threatening phenomenon. (3) Pituitary apoplexy is an endocrine emergency. (2) Association between MSA and pituitary apoplexy has not
been described so far. Other emergencies described in movement disorder are acute spinal rigidity, tics, neuroleptic malignant syndrome, dystonic syndrome, malignant catatonia.

(4) We present 2 cases of MSA, one is MSA-P with pituitary apoplexy, and other is MSA-C with stridor.

**CASE 1:**
Forty two years old male presented with history of sexual dysfunction for 18 months duration, urinary incontinence for 12 months duration, postural giddiness and slowness of activity of daily living for 6 months duration, along with stiffness of trunk and limbs suggestive of multi system atrophy with predominant parkinsonism (MSA-P). With this background problem, the patient also gives history of acute onset of severe headache over bifrontal region for 3 days. No visual disturbances or focal neurological deficit were present. On examination, his MMSE was 30/30, lobar functions were normal except emotional disturbances. Cranial nerve examination showed no field defect and Fundus was normal. Motor system examination revealed Rigidity were present in all limbs, neck and trunk with normal power, Extra pyramidal system: blink – reduced, mask like face was present, glabellar nonaccomodative, bradykinesia was present, no rest tremor, Axial rigidity was noted, Propulsion, retropulsion, lateropulsion were also noted. Festinant gait and Pisa syndrome were noted, Arm swing was reduced, more on RT side, Bedside autonomic function testing revealed dysfunction.

One day after the admission, patient developed altered sensorium. Neuroimaging revealed pituitary adenoma with diffusion restriction, Blooming on GRE was present suggestive of pituitary apoplexy. Hormonal assay was consistent with nonfunctioning pituitary adenoma. Transnasal transphenoidal decompression was done resulting in complete recovery from headache and altered sensorium. Biopsy showed nonsecreting pituitary adenoma. Now patient is better with fludrocortisone, dopamine replacement and anticholinergics. This is the first description of MSA-P subtype coexisting with pituitary adenoma. We hypothesize that dysautonomia could be the cause for pituitary apoplexy.

**CASE-2:**
Forty eight years male presented with unsteadiness while walking, slurring of speech for 6 months, erectile dysfunction for 3 months, postural giddiness for 1 month, increased frequency of micturition for 1 month, urinary retention for 2 days, constipation for 1 week, History of snoring for 1 week. His past medical history was unremarkable. On examination, his respiratory rate was 20 bpm, oxygen saturation was 99%, significant postural fall in blood pressure was present. MMSE was 29/30. Cranial nerve examination was normal. Motor system examination revealed spasticity of all 4 limbs with normal power, brisk deep tendon reflexes, plantar bilaterally extensor. Extra pyramidal system revealed bradykinesia. Cerebellar examination revealed predominant gait ataxia and dysarthria. Bed side autonomic function testing revealed
dysfunction. Patient was subjected to polysomnography, which showed stridor and sleep apnoea. Laryngoscopy examination revealed no abnormality. Next day patient suddenly went for hypoxic ischemic encephalopathy. Patient was intubated and later elective tracheostomy was done. Now the patient is has permanent tracheostomy and T.fludrocortisone and he is better.

Discussion:
Multisystem atrophy presents with motor and autonomic deficits and was first described by Gram and Oppenheimer in 1969. Later during 1990s it was redefined as a progressive neurodegenerative disease of undetermined cause, occurring sporadically. Bilateral vocal cord paresis is well documented in multiple system atrophy and many patients ultimately die of respiratory complication as a result of airway compromise secondary to this. The pathology is related to a selective abductor paresis and tends to be worse during sleep. Patients with symptoms of airway obstruction should receive tracheostomy to bypass the level of obstruction. In sheehan syndrome, hypotension secondary to blood loss is the cause for pituitary apoplexy. In our case, hypotension due to dysautonomia is the cause for pituitary apoplexy. This case is presented for its unusual association and we hypothesize that autonomic dysfunction (hypotension) could be the cause for pituitary apoplexy as cited in Endocrine diseases by Willium F Young.

Conclusion:
Multi system atrophy is an uncommon neurodegenerative disorder which can present itself with emergencies involving autonomic nervous system and respiratory system. Prompt identification and intervention is important to save the life of the patient. Dysautonomia should be managed with caution to avoid life threatening complications. The occurrence of stridor and sleep apnoea in patients with MSA is associated with decreased life expectancy. Polysomnography is the important diagnostic tool for diagnosing stridor in MSA. Our cases were presented to highlight the importance of identifying this uncommon association and timely management of these life threatening complication.

References:
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