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A rare cause of urinary retention - Multiple System Atrophy, a case report.

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Abstract : Multiple system atrophy (MSA) is a rare neurodegenerative disorder with various combinations of pyramidal, extrapyramidal, cerebellar, or autonomic Involvement whose presentation can be any one of these making it difficult to diagnose unless it is considered. More frequently misdiagnosed as idiopathic Parkinsons disease which is subsequently revealed by poor response to levodopa. Here we report 70 year old man admitted with urinary retention which was due to autonomic involvement and also had pyramidal, extra pyramidal, cerebellar features.MR imaging of brain showed hot cross bun sign, so a diagnosis of multiple system atrophy was made.

Keyword :multiple system atrophy , hot cross bun sign , MSA -P, MSA-C.

Introduction: Multiple system atrophy is a progressive and sporadic neurodegenerative disorder. Extrapyramidal involvement (termed *MSA-P*, for *Parkinsonism*) is about three -fold more common than cerebellar involvement (termed *MSA -C* for *cerebellar*). Multiple system atrophy occurs in early fifties and progresses relentlessly with a mean survival of about five (range 1–11) years[1]has estimated prevalence of 4.4 cases per 1,00,000[2]. Here we report a case multiple system atrophy presented with urinary retention, an autonomic nervous system involvement. Case report: A 70 year old man came for not passing urine for one day. Also, gives history of giddiness on standing and swaying while walking for two years. Blood pressure in supine position was 130/90mmhg as against 100/70mmhg. Patient with mask like face had decreased arm swing and turning en-bloc while walking with minimal tremors. Had slowness for reaching objects with normal cranial nerves and higher mental function. Motor system examination showed normal bulk, reduced power in all four limbs, hypertonia with cog wheel rigidity, deep tendon reflexes exaggerated, bilateral plantar extensor.Cerebellar signs like positive finger nose test, heel knee test, pastpointing, dysdiadokinesia, wide based ataxic gait, stance ataxia.Routine blood investigations with ECG, echocardiogram were normal.MRI showed cerebellar atrophy (fig1) with dilated fourth ventricles, peduncles, and cerebellum on T2-weighted images. Classically a cruciform hyperintensity in the pons on T2-weighted MRI, known as the “hot cross bun” sign is present with no infarct in brain (fig2).So patient with cerebellar signs,parkinsonism features,autonomic nervous system involvement(orthostatic hypotension, urinary bladder dysfunction),a diagnosis of Multiple system atrophy (both MSA-P and MSA-C type) was made which was supported by MR imaging. Since it is neurodegenerative disorder with cure not possible, so patient was started on symptomatic treatment with levodopa, fludrocortisone and liberal salt intake.

Discussion: MSA is a rare disease, difficult to diagnose. It was first described by Gram and Oppenheimer in 1969 which has undetermined cause, occurring sporadically.Genetic factors do not seem to play an impor-tant role and no gender predilection exists.At autopsy,MSA is characterized by cell loss and gliosis in the striatum, Subthalamic Nucleus, locus ceruleus, inferior olive, pontine nuclei, Purkinje cells, intermediolateral cell column, and the Onuf nucleus in the sacral spinal cord.Glial cytoplasmic inclusions containing alpha-synuclein are the most characteristic histological feature[3]. A latest consensus conference in 2007 [4] categorised MSA into two simplified form. MSA-P type with predominant parkinsonism features; previously called *striatonigral degeneration.*MSA-C type with predominant cerebellar features ; previously called *sporadic olivopontocerebellar atrophy(OPCA).Shy-Drager syndrome* with predominant autonomic failure (not defined in the present consensus anymore). The designation of MSA-P or MSA-C depends on the dominant feature at the time of evaluation, sometimes both may be present. The most common signs in pathologically confirmed cases are Parkinsonism (87%), autonomic dysfunction (74%), cerebel-lar ataxia (54%), and pyramidal signs (49%).

Also designations of definite, probable, and possible MSA from previous classification is retained. Definite MSA requires demonstration of CNS alpha-synuclein positive glial cytoplasmic inclusions. Probable MSA requires a sporadic, progressive adult-onset disorder including rigorously defined autonomic failure (orthostatic hypotension) with Urogenital dysfunction (Urinary incontinence, incomplete bladder emptying or erectile dysfunction) and cerebellar ataxia or poorly levodopa-responsive Parkinsonism. Possible MSA requires a sporadic, progressive adult-onset disease with at least one feature suggesting autonomic or urogenital dysfunction and Parkinsonism or cerebellar ataxia plus clinical features (Babinski sign with hyperreflexia, stridor) or a neuroimaging abnormality.

MSA-P should be distinguished from idiopathic parkinsonism by the absence of a resting tremor, more severe autonomic dysfunction, and evidence of pyramidal tract involvement (e.g., spasticity, pres-ence of a Babinski sign).Also “red flags “signs[5]used to differentiate MSA-P from idiopathic parkinsonism. The red flags are early instability, rapid progression, abnormal postures (includes Pisa syndrome, disproportionate anterocollis, contractures of hands or feet), bulbar dysfunction (includes severe dys-phonia, dysarthria, dysphagia), respiratory dys-function (stridor, inspiratory signs) [6] and emotional incontinence (inappropriate crying, laughing). A combination of 2 out of these 6 red-flag categories is used as additional criteria for the diagnosis of MSA-P. Cognitive functioning in MSA-P is relatively preserved, but mild executive deficits are observed. MSA-C (formerly sporadic OPCA) is distinguished primar -ily by the presence of dysarthria and ataxia in association with marked cerebellopontine atrophy on brain MRI. Autonomic manifestations are orthostatic hypotension (preganglionic damage of intermediolateral cell columns), impotence, erectile dysfunction, urinary problems, constipation, and hyperhidrosis. Also have emotional liability, with short episodes of crying due to happiness or sadness in response to minor environmental stimulus, such as a song or movie. Involvement of the bladder occurs in most patients. Early complaints are urgency, frequency, and nocturia, later picture is dominated by overflow incontinence, more consistent with a lower motor neuron bladder (Sacral intermediolateral cell column loss). Post-void residuals steadily increases that a large post-void residual in a patient with a Parkinson-like disorder suggests MSA rather than PD [7]. Some MSA patients have sleep apnea which rarely may lead to a critical loss of respiratory drive, the so-called Ondine’s curse. The most common causes of death in patients with MSA are pulmonary embolus, apnea, and intercurrent infection. In this disorder, MR imaging shows olivopontocerebellar atrophy (OPCA), cerebellar atrophy, and the putaminal lesions of striatonigral degeneration. Cruciform hyperintensity within the pons in MR imaging, the so-called hot-cross-bun sign, due to a selective loss of myelinated transverse pontocerebellar fibers may also be a helpful marker but not specific [8, 9]. PET scan with decreased subcortical AChE activity more in MSA-P than in PD.Clinical tests of autonomic dysfunction like abnormal response to Valsalva maneuver, reduced response to isometric exercise (handgrip), diminished response to cold pressor test may be helpful. Denervation pattern on rectal sphincter electromyog-raphy (EMG) is seen. Treatment of MSA is difficult with no specific inter-ventions, and symptomatic therapies provide only partial relief of disability.Patients with MSA-P initially may respond to levodopa, but the duration of benefit typically is short lived unlike idiopathic Parkinsonism where it has good response.There is no effective treatment for the MSA-C type. Orthostatic hypo-tension may improve with nonpharmacological measures such as liberal salt and water intake, compression stockings, and sleeping with the head up, but most patients require phar-macotherapy with fludrocortisone, midodrine, droxidopa. Treatment of orthostatic hypotension often worsens supine hypertension but orthostatic hypotension itself can be exacerbated by levodopa. Urinary catheterization when urine retention is severe. Even in the best hands, MSA has a poor prognosis, with a mean survival of 7 to 9 years but in the last 5 years, several promising developments have occurred. Rifampicin was shown to inhibit aggregation of -synuclein invitro [10] leading to a National Institutes of Health–sponsored trial of rifampicin in MSA begun in 2011. A trial of intravenous globulin has now been completed, and the results are pending. Finally Antiparkinsonian drug, rasagiline, an inhibitor of monoamine oxidase type B usefulness in MSA [11] has been studied.

References:

1.G. K. Wenning, C. Colosimo, F. Geser, and W. Poewe, “Multiple system atrophy,” Lancet Neurology, vol. 3, no. 2, pp. 93–103, 2004.

2.Schrag A., Ben-Shlomo Y., Quinn N.P.: Prevalence of progressive supranuclear palsy and multiple system atrophy: a cross-sectional study. *Lancet* 1999; 354: 1771-1775.

3.Wenning GK, Jellinger KA. The role of alpha-synuclein in the pathogenesis of multiple system atrophy. *ActaNeuropathol*. 2005;109:129-40.

4.Gilman S, Wenning GK, Low PA, Brooks DJ, Mathias CJ, Trojanowski JQ. Second consensus statement on the diagnosis of multiple system atrophy. *Neurology*. Aug 26 2008; 71(9):670-6.

5.Zesiewicz TA, Sullivan KL, Gooch CL. Nat ClinPract Neurol. 2008 Nov; 4(11):596-7. Epub 2008 Aug 12.

6.Mehanna, Jankovic, 2010. Mehanna R., Jankovic J.: Respiratory problems in neurologic movement disorders. Parkinsonism RelatDisord 2010; 16:628-638.

7.Hahn, Ebersbach, 2005. Hahn K., Ebersbach G.: Sonographic assessment of urinary retention in multiple system atrophy and idiopathic Parkinson's disease. MovDisord 2005; 20(11):1499-1502.

8.Massano J, Costa F, Nadais G. Teaching neuroImage: MRI in multiple system atrophy: "hot cross bun" sign and hyperintense rim bordering the putamina. *Neurology*. Oct 7 2008;71(15):e38.

9.Brooks, Seppi, 2009. Brooks D.J., Seppi K.: Proposed neuroimaging criteria for the diagnosis of multiple system atrophy. MovDisord 2009; 24:949-964.for the Neuroimaging Working Group on MSA.

10.Ubhi K, Rockenstein E, Mante M, Patrick C, Adame A, Thukral M, et al. Rifampicin reduces alpha-synuclein in a transgenic mouse model of multiple system atrophy. Neuroreport. 2008;19:1271–1276.

11.Stefanova N, Poewe W, Wenning GK:Rasagiline is neuroprotective in a transgenic model of multiple system atrophy.Exp Neurol. 2008 Apr; 210(2):421-7.

Legends:

Fig1: MRI brain showing diffuse cerebellar atrophy

Fig2: MRI brain showing a cruciform hyperintensity signal in pons called “ hot cross bun “ sign due to atrophy of transverse pontocerebellar fibers.