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Case Report

Role of MRI in diagnosis of multiple system atrophy: A case report

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Abstract

Multiple system atrophy (MSA) is a sporadic, progressive neurodegenerative disorder of unknown etiology, characterized by various combinations of autonomic, cerebellar, pyramidal and extra pyramidal signs. Based on the consensus criteria, patients with MSA are classified as MSA-C and MSA-P. MRI plays an important role in the early diagnosis. The characteristic finding “hot cross bun’’ sign is seen in MSA of cerebellar type. Here we present a case of MSA-C in 52 years old male patient.

Key words

Multiple system atrophy, MSA, Cerebellum, MRI.

Introduction

Multiple system atrophy (MSA) is a sporadic, progressive, idiopathic neurodegenerative disorder of adult onset [1]. It is characterized by several varying combinations of cerebellar, pyramidal, extra pyramidal and autonomic signs [1, 2, 3, 4, 5, 6]. The annual incidence of MSA is 0.6/ 1,00,000. MSA is a distinct clinic-pathologic entity previously known as olivopontocerebellar atrophy, striatonigral degeneration and shy-dragger syndrome are now all named as MSA. Three clinical subtypes are described.

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- MSA-C when cerebellar signs predominates the clinical picture.
- MSA-P when parkinsonian features predominate.
- MSA-A when the patient presents with autonomic signs and symptoms.

Neuroimaging studies, especially the MRI show changes, that are not specific, but helps in differentiating various forms of MSA. Basing on MRI findings two distinct subtypes are described: MSA-C and MSA-P. We herein report a case of MSA-C, who presented with difficulty in walking, slurring of speech, urinary retention and constipation.

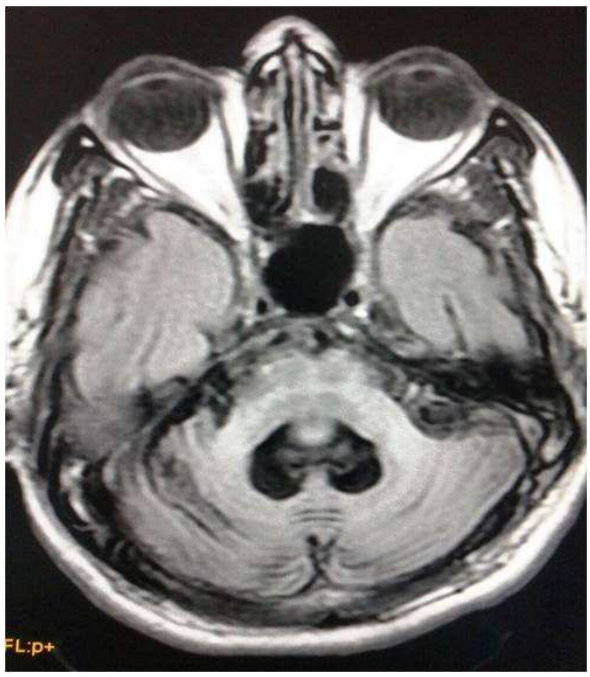
Case report

52 years old male patient presented with complaints of difficulty in walking since 3 years with slowing of walking and intentional tremors in the right hand. He developed ataxic gait 1 year back. He had complaints of urinary retention and constipation along with slurring of speech which started 20 days back. Family history and personal history were insignificant. Blood biochemical investigations were within normal limits. MRI brain revealed flat pons and medulla with atrophic cerebellar hemispheres and vermis. The characteristic finding of cruciform pontine hyper intensity (hot cross bun sign) was observed. **(Photo – 1, Photo – 2, Photo – 3)** Basing on the clinical picture and classical imaging findings the case was diagnosed as multiple system atrophy of cerebellar type.

Discussion

MSA is a rare progressive neurodegenerative disease. It has two main subtypes: MSA-C (Cerebellar) and MSA-P (Parkinsonian). In a study conducted by the European MSA study group (EMSA-SG), statistically significant red flags for the differential diagnosis were noted. The presence of at least two of six red flags (early instability, rapid progression, abnormal posture, bulbar dysfunction, respiratory dysfunction and emotional incontinence) was reported to be 98.3% specific and 82.4% sensitive for the diagnosis of MSA [7]. Rapid progression is one of the very important warning signs for MSA patients.

Photo - 1: Axial FLAIR image showing cruciform hyper intense signal in the atrophic pons (hot cross bun sign).



Cerebello-olivary atrophy, Friedrich's ataxia, Progressive non familial adult onset cerebellar degeneration, hereditary olivopontocerebellar atrophy are considered in the differential diagnosis [1]. Histopathological finding in MSA-C include glial cytoplasmic inclusions and neuron loss that is predominant in cerebellar white matter, pons, inferior olives and middle cerebral peduncle. These inclusions are constituted by alpha-synuclein, ubiquitin and tau protein [6, 8, 9].

MRI plays an important diagnostic tool in the early course of MSA-C. Hot cross burn sign

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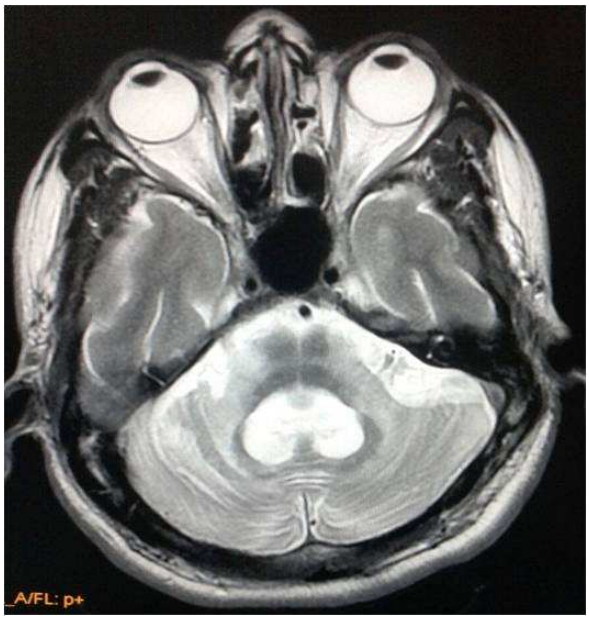
appears early in MSA-C [10]. Though characteristic T2 hyper intense sign in pons and cerebellar peduncle reflects pontocerebellar fiber degeneration is characteristically seen in MSA-C, it can be found in other forms of parkinsonism [11]. This is caused by loss of myelinated transverse pontocerebellar fibers in the pontine raphe with preservation of the pontine tegmentum and corticospinal fibers [1]. There is no specific treatment to MSA until now, except for symptomatic interventions [12].

Photo - 2: Axial T2 WI showing cruciform hyper intense signal in the atrophic pons (hot cross bun sign).



Our case was classified as MSA-C according to criteria in consensus, since the diagnosis of MSA was defined just with pathological analysis. We conclude that the brain MRI changes might increase the accuracy in diagnosis of MSA.

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Photo - 3: T2 sagital image showing atrophy of pons.