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Multiple system atrophy - The cerebellar type with “hot cross bun sign” on MRI

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CASE REPORT

Multiple system atrophy — the cerebellar type with "hot cross bun sign" on MRI

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Abstract

Multiple system atrophy (MSA) is an adult onset, progressive, idiopathic neurodegenerative disease that clinically manifest as MSA-C with cerebellar signs predominant, MSA-P with Parkinsonian features predominant and MSA-A with autonomic signs and symptoms. MRI has an important role in its diagnosis with a characteristic finding described as "hot cross bun" sign seen in MSA of cerebellar type (MSA-C). We have seen two cases of MSA-C referred to Radiology Department, Civil Hospital Karachi in February 2017 and March 2017 who presented with difficulty in walking and speech and foecal and urinary incontinence. The MRI showed atrophy of pons, cerebellum and middle cerebellar peduncles with cruciform hyperintense signals in pons (hot cross bun sign). Multiple system atrophy has no specific treatment so the patients were on supportive therapy. We are reporting these interesting cases along with the clinical and radiological findings as well as literature review.

Keywords: Hot cross bun sign, Multiple system atrophy, MRI, Neurodegenerative disease.

Introduction

Multiple system atrophy (MSA) is an adult onset, progressive, idiopathic neurodegenerative disease that involves, to varying degree, the basal ganglia, olivopontocerebellar complex and autonomic system.[1-3] It has three clinical subtypes: MSA-C when cerebellar signs are predominant, MSA-P when parkinsonian features are predominant and MSA-A when the patient presents with autonomic signs and symptoms.[3] The presence of hot cross bun sign on T2WI is a powerful tool for diagnosis of MSA-C in appropriate clinical setting.

After taking consent from patients, we are reporting these two cases of MSA-C who presented with difficulty in walking, slurring of speech, urinary and faecal incontinence. Such cases have not been reported earlier from Pakistan. It highlights the importance of considering MRI as a good imaging tool to increase accuracy in diagnosing MSA in the relevant clinical scenario.

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Case-1

A 58-year male patient was referred to the radiology department in February 2017 for MRI brain with history of difficulty in walking and speech for two years. He had complaints of urinary and faecal incontinence for the last one and a half year. Fine tremors in hands, postural faintness and ataxia were added to these complaints one year ago. Initially there was difficulty and slowing of walking and for the last six months he could not walk without assistance.

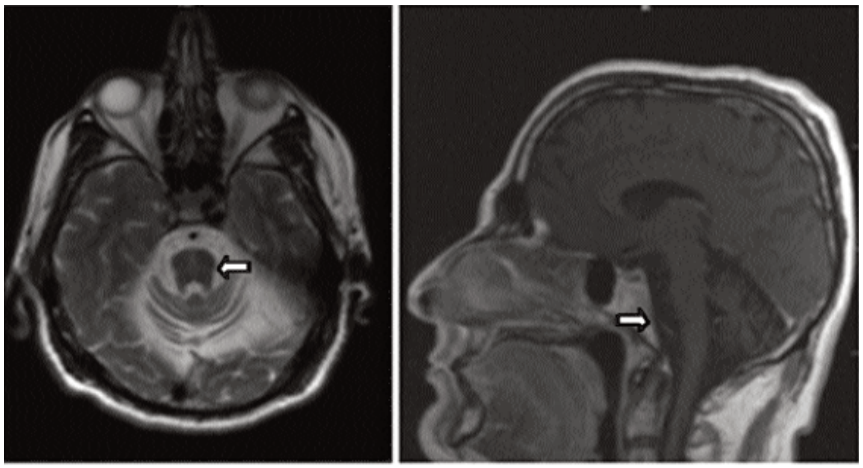

Figure-1 (a-b): Axial T2 (a) and saggital T1 post Gd (c) images of 58-years male patient show infratentorial atrophy (right arrow) and cruciform pontine hyperintensity (hot cross bun sign- left arrow) on axial T2W image.

On examination, he had slurred speech and wide-based ataxic gait; so he could walk with one-person support. Romberg sign was positive with clumsy finger-nose test, dysdiadokinesia and heel shin test. Reflexes were exaggerated in all limbs however muscle tone, bulk and power were normal. Higher motor functions, cranial nerves and sensory system were intact. Pupils were bilaterally normal and equally reacting to light. Patient's

past medical and personal histories were unremarkable. Family history was also unremarkable. Routine laboratory results were within normal limits. His brain MRI showed atrophy of pons, medulla and cerebellum, and high signals in pons and middle cerebellar peduncle with cruciform pontine hyperintensity (hot cross bun sign) on axial T2W image (Figure-1). Supratentorial structures including basal ganglia were unremarkable. On combination of clinical picture and classical MRI findings the patient was diagnosed as multiple system atrophy of cerebellar type (MSA-C).

As there is no specific treatment of MSA, he was managed by the neurology department on supportive and rehabilitative therapy.

Case-2

A 57-year male patient was referred to the radiology department in March 2017 for MRI brain with history of difficulty in walking and speech, and urinary and faecal incontinence for one year. He had fine tremors in hands, postural faintness and ataxia. Initially there was slowed walking but for the last two months, he could not walk without assistance. On examination he had ataxic gait so he could walk with one person support, slurred speech and clumsy finger nose test, and dysdiadokinesia. Reflexes were increased in all limbs however muscle tone, bulk and power were normal. Higher motor function, cranial nerves and sensory system were intact. Pupils were normal bilateral equally reacting to light. The past medical, personal and family history was insignificant. His routine laboratory investigations were unremarkable. His brain MRI showed atrophy of pons, medulla and cerebellum, and high signals in pons and middle cerebellar peduncle with cruciform pontine hyperintensity (hot cross bun sign) on axial T2W image (Figure-2). Cerebrum and basal ganglia were unremarkable. On combination of clinical picture and classical MRI findings the patient was diagnosed as multiple system atrophy of cerebellar type (MSA-C).

He was also managed by neurology department on supportive treatment.

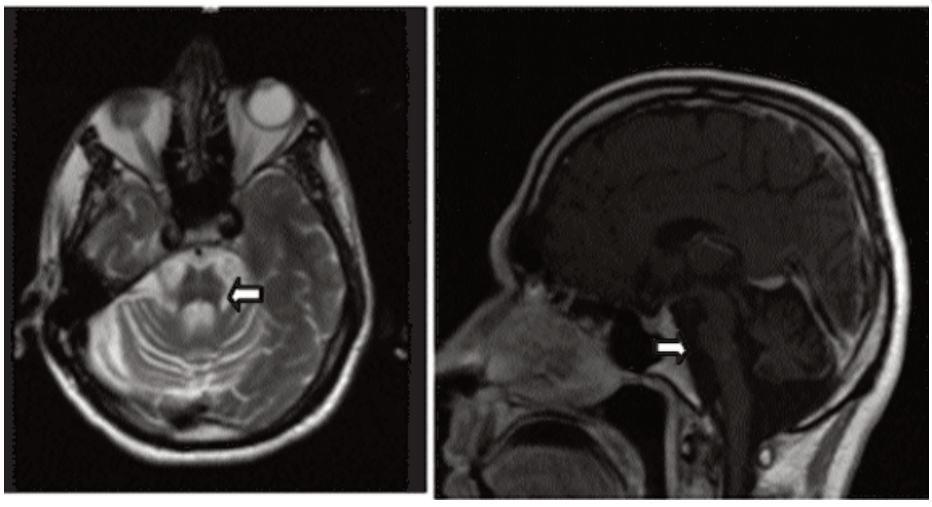

Figure-2 (a-b): Axial T2 (a) and saggital T1 post Gd (c) images of 57-years male patient showing atrophy of pons, medulla and cerebellum (right arrow) with cruciform hyperintensity in pons (hot cross bun sign- left arrow) on axial T2W image.

Discussion

Multiple system atrophy (MSA) is a rare, progressive neurodegenerative disease of unknown etiology presenting in adults.[1-3] Both these patients were adult males towards the elderly age. It is manifested clinically by various combinations of autonomic, cerebellar, pyramidal and parkinsonian signs and symptoms. But autonomic dysfunction is mandatory for the diagnosis of MSA according to the revised consensus criteria,[1] our both patients has urinary and fecal incontinence. There are three clinical subtypes: MSA-C when cerebellar signs are predominant, MSA-P when parkinsonian features are predominant and MSA-A when the patient presents with autonomic signs and symptoms.[3] Formerly these were synonymous as sporadic olivopontocerebellar atrophy (sOPCA), striatonigral degeneration and Shy-Drager syndrome respectively.[3,4] However, these subtypes share common histopathological changes, characterized by neuronal loss, gliosis and the presence of glial cytoplasmic inclusions (GCI) with a-synuclein.[5] In MSA-P the degenerative changes predominantly affect the basal ganglia, particularly the putamen seen as hyperintense rim at the putaminal edge, atrophy and hypointensity of putaminal body on T2WI while in MSA-C changes predominantly affect infratentorial structures like pons and cerebellum, seen as atrophy and hyperintense signals in pons, cerebellum and middle cerebellar peduncles with pontine hyperintensity (hot cross bun sign) on axial image,[4] which was seen in both of our patient's MRI. Hot cross bun sign refers to the cruciform shaped pontine T2 hyperintensity in multiple system atrophy due to selective loss of myelinated transverse pontocerebellar fibers and neurons in the pontine raphe and sparing of the pontine tagmentum and corticospinal tracts.[3,6] Though hot cross bun sign is seen often in MSA, it is not

pathognomonic but only a supportive sign on clinical background.[7]

In a study conducted by the European MSA study group (EMSA-SG), statistically significant red flags for the differential diagnosis of MSA 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.[2,3,8] Rapid progression from red flags criteria is a very important warning sign for MSA patients;[3] both of these patients were also giving the history of rapid progression of signs and symptoms, early instability and abnormal posture. So both cases were classified as MSA-C according to criteria in consensus and red flags along with MRI findings. Since the diagnosis of MSA is confirmed by pathological analysis but the brain MRI findings increase the accuracy of diagnosis of MSA in correlation with the clinical picture. There is no specific treatment to MSA until now, so patients were discharged with symptomatic treatment.

Conclusion

The aim to present this short series of a rare condition was to emphasize the role of MRI in MSA, and to assist clinicians by reviewing literature and summarizing important features of the MSA.

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Conflict of Interest: None to declare.

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