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HOT CROSS BUN" SIGN IN MULTIPLE SYSTEM ATROPHY CEREBELLAR TYPE (MSA-C): A CASE REPORT

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ABSTRACT

Multiple system atrophy (MSA) may be difficult to distinguish clinically from other disorders, particularly in the early stages of the disease. A careful medical history and meticulous neurological examination remain the cornerstone for the accurate diagnosis of MSA. We report a case of 50 years old male patient with cerebellar ataxia, right side hemiplegia, dysphagia, aphasia, and constipation. MRI revealed atrophy with the abnormal signal intensity of pons, middle cerebellar peduncles, and olivary nucleus features indicating atrophy of the olivopontocerebellar pathway. Cruciform T2 hyperintense signal in pons extending along the middle cerebellar peduncles bilaterally giving ``hot cross bun'' sign with T2 hyperintensities in the medulla & bilateral cerebellar hemisphere with mild cerebellar atrophy possibility of Multiple system atrophy (MSA-C type).

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INTRODUCTION

Multiple system atrophy (MSA) is a rare and mainly sporadic, adult-onset, progressive and fatal neurodegenerative disorder clinically characterized by various combinations of autonomic failure, parkinsonism, and ataxia [1]. Its pedigree with both autosomal dominant and autosomal recessive inheritance patterns has been reported in Europe and Asia [2,3,4]. The different predominance of symptoms gives rise to different MSA subtypes. The parkinsonian subtype (MSA-P) is defined when bradykinesia with rigidity, tremor, or postural instability dominates the clinical picture. The cerebellar form (MSA-C) is considered when cerebellar ataxia is the most prominent symptom. The new diagnostic criteria consider now abundant α-synuclein-positive glial cytoplasmic inclusions as neuropathological hallmarks and try to improve the recognition of patients with early or possible MSA by including red flags and neuroimaging aspects.

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The diagnosis of possible MSA-C is considered in the presence of a cerebellar syndrome (gait ataxia with cerebellar dysarthria, limb ataxia, or cerebellar oculomotor dysfunction) and at least one feature suggesting autonomic dysfunction (otherwise unexplained urinary urgency, frequency or incomplete bladder emptying, erectile dysfunction in males, or significant orthostatic blood pressure decline that does not meet the level required in probable MSA) [5]. Recently, we had a case of a man who had chronic lacunar infarcts and an area of encephalomalacia. He had cortical atrophy and white matter atrophy. In addition, he had middle cerebellar atrophy giving hot cross bun sign possibility of multiple system atrophy (MSA- C type).

CASE REPORT

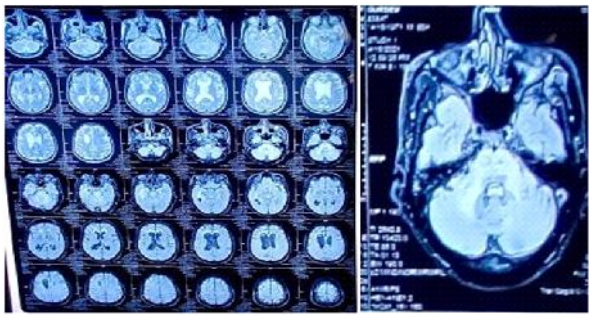
A 50 year old male patient came with a case of ataxia and weakness of the right side. He was unable to speak and had noted weakness in his right arm and right leg one day before (5th April 2021), which was progressive and worsened with time. He had a flaccid tone of muscle with less bulk on the right side, depicting paralysis. Stiffness was also noted in the

right leg. He had constipation, for which an enema was given. The patient urine volume per day was 1-1.5 liters (Foley's catheter was inserted into the attached urobag).

He had difficulty in swallowing, restlessness, otherwise, he responded to commands. His oxygen saturation of 99 was maintained with the help of an oxygen mask. The patient was admitted to the same hospital 8 to 9 months back with similar complaints and was treated accordingly. There was a history of falling to the right side as told by the informant. He had no vomiting, no blurring of vision, no chest pain, or breathlessness. No history of convulsions, dizziness, and leg cramps. The patient showed no history of hypertension, diabetes, or any other chronic ailment. A similar complaint was not noted in any other family member, although there was a history of hypertension in the patient's father and brother. The patient showed a history of chronic alcoholism (almost every day) but was abstained from alcohol for the past 1.5 years. On physical examination, he was conscious and cooperative. He was not able to speak and responded only with gestures. Cranial nerve functions and facial expressions were not normal. Right-sided visual acuity was reduced. Extraocular movements were not normal, unable to deviate his eye's right side as well as the right upper side. Light reflex (constriction of the pupil) and accommodation reflex were normal. Nystagmus was evident and he was unable to close his eyes forcefully. He was not able to clench his teeth with full strength or open his mouth completely. He was unable to blow with his mouth and not able to protrude his tongue out completely.

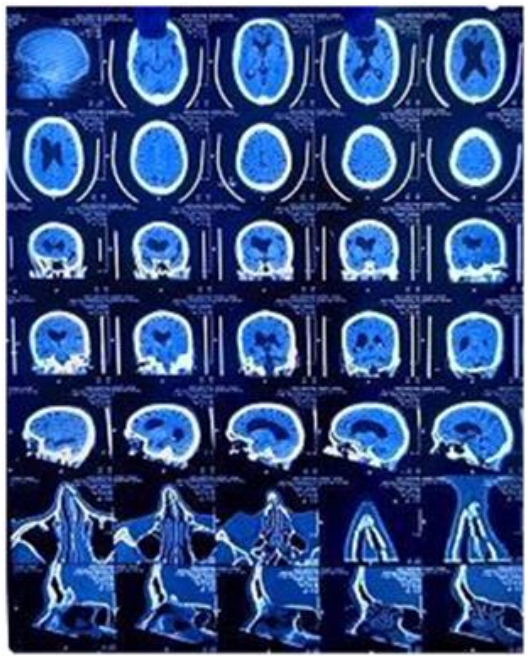
He also had vertigo and tinnitus (cerebellar ataxia) with respect to coordination. He was not able to perform a finger-nose test properly, diadochokinesia, finger to ear test. Tinnitus was present on Rinne's test as the air conduction was more than bone conduction (on both sides). However, on Weber's test, the sound was radiating towards the right. He had motor paresis. He was unable to swallow both liquids and solids properly. He was also not able to shrug his right shoulder and showed resistance while tilting both sides. On clinical examination, motor power of the right upper limb and lower limb was grade 0. Motor power of left upper and lower limb was grade 6 and 3 respectively. He had shown general sensations of pain, touch, pressure, and temperature. There was the presence of tactile localization, two-point sensation, stereognosis, barognosis as well as graphaesthesia. Superficial reflex examination showed, abnormal plantar reflex, dorsiflexion of great toes, and fanning of toes to the left side (Babinski's sign), and nonresponse was noted on the right side. Deep tendon reflexes on all four limbs were evident and were grade 1(biceps jerks, triceps jerk, and ankle jerk) and grade 2 knee jerk.

MRI Brain at 1.5 T was performed. The neuroimaging finding was as follows.e/o cruciform T2 hyperintense signal on axial images in pons extending along the middle cerebellar peduncles bilaterally giving ``hot cross bun'' sign. T2 hyperintensities were also seen extending in the medulla & bilateral cerebellar hemispheres with mild cerebellar atrophy. No restriction or blooming was seen. On post-contrast scans, no significant enhancement was seen. Chronic lacunar infarcts were seen in bilateral frontal lobes. Area of encephalomalacia with gliosis was seen in the right temporal lobe with ex vacuo dilatation of the right lateral ventricle. T2/FLAIR hyperintensities were seen in deep white matter in the periventricular region bilaterally. These lesions did not show restriction on DW/ADC or blooming on SWI c/o chronic white matter ischemic changes. The brain parenchyma otherwise was normal in signal intensity with normal gray white matter differentiation.

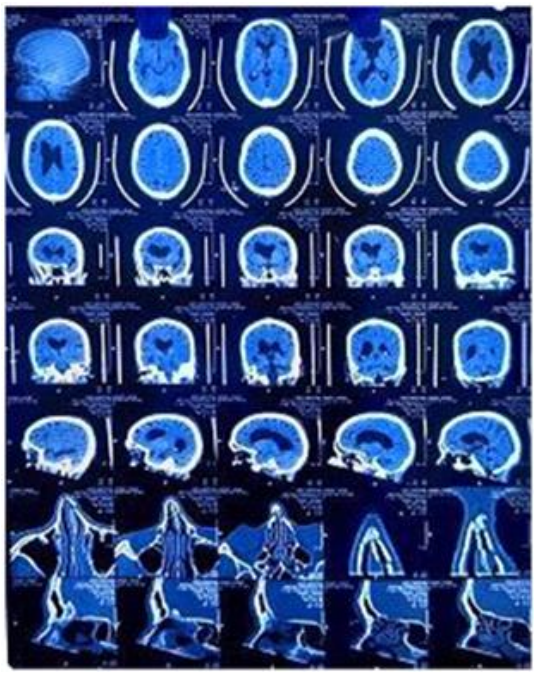


a b

Fig 1. (a & b): Conventional brain magnetic resonance imaging. Axial image: showing the "Hot cross bun" sign



a



b

Fig .2 (a & b): Conventional brain magnetic resonance imaging: showing atrophy in pons medulla and cerebellum

Bilateral thalami & basal ganglia were normal. posterior fossa structures including the brain stem, cerebellum, and fourth ventricle were normal. The ventricular system was prominent. Sulcogyralspaces, Sylvian fissures & basal cisterns were prominent s/o cortical atrophy. No extra-axial collection was seen. No e/o diffusion restriction/blooming was seen on DWI/ADC/SWI images. All these features suggested possible Multiple System Atrophy- C type.

DISCUSSION

The neuropathological hallmark of MSA is glial cytoplasmic inclusions containing α-synuclein, providing a pathological link between Parkinson's disease (PD), dementia with Lewy bodies (DLB), and MSA: all α synuclei[6,7]. The incidence of MSA is 0.6 -0.7 cases per 10,000 person-years, with a range of 0.1 to 3.0 cases per 100,000 person-years. Studies from Russia and Northern Sweden reported incidences of 0.1 and 2.4 per 10,000 persons-years, respectively. Incidences increase with age up to 12/100,000 above 70 years. In the western hemisphere, MSA-P involves about 66-82% and MSA-C is more frequent in the Asian population accounting for about 67% suggesting a role for the different genetic background or environmental factors in the development of MSA subtypes[8]. Patients with MSA have a mean age at onset of 55-60years, and an average survival from the onset of motor symptoms of 8-9 years, although some pathology proven cases survived >15years [1, 9]. In the present case, a 50 year old male patient with cerebellar dysfunction was admitted to the hospital. The patient displayed cerebellar ataxia, right side hemiplegia, dysphagia, aphasia, and constipation. Medical technology such as functional MRI (f MRI) measures activity levels in the brain and can demonstrate areas of impaired brain function. Some atrophic changes are also visible upon the use of MRI.

In the present case study, On MRI, signal changes & atrophy of pons, middle cerebellar peduncles, the cerebellum, inferior cerebellar peduncle & olives have been seen. On Axial T2WI and FLAIR sequences-typically hyperintensity in the cruciform pattern described as "Hot Cross Bun" Sign is seen consisting of the transverse pontine fibers coursing mediolaterally and the pontine raphe coursing anteroposteriorly. Atrophy & hyperintensity involving middle cerebellar characteristic T2 hyperintense sign in the pons and middle cerebellar peduncle ("cross sign") reflects pontocerebellar fibers degeneration and suggestive of MSA it can be found in other forms of Parkinsonism. The "hot cross bun" sign is seen not only in patients with MSA, but also in some patients with other diseases, such as spinocerebellar ataxia, variant Creutzfeldt-Jakob disease, and selective Wallerian degeneration of transverse pontocerebellar fibers due to vasculitis[10,11,12]. Nevertheless, the "hot cross bun" sign is highly specific for MSA and is a trademark of disease progression in general [13,14]. The "hot cross bun" sign is attributable to neuronal loss and astrocytic gliosis in areas of transverse pontine tracts with relative preservation of the pontine tegmentum and corticospinal tracts, which run craniocaudally into the dorsal pons [15,16]. The second consensus statement on the diagnosis of MSA noted that MR imaging could assist diagnosis and that T2-signal change on 1.5-T MR imaging in the brainstem could be helpful, including the "hot cross bun" sign[17]. On T2WI, Horimoto et al. reported that the pontine "cross sign" is useful to distinguish clinical subtypes of MSA, as this sign becomes evident about 5 years earlier in MSA-C than in MSA-P.

CONCLUSION

This is an uncommon case of MSA-C in a 50year old male with predominant cerebellar signs. MRI revealed atrophy with the abnormal signal intensity of pons, middle cerebellar peduncles, and olivary nucleus features indicating atrophy of the olivopontocerebellar pathway. Cruciform T2 hyperintense signal in pons extending along the middle cerebellar peduncles bilaterally giving` `hot cross bun" sign with T2 hyperintensities in the medulla & bilateral cerebellar hemisphere with mild cerebellar atrophy possibility of Multiple system atrophy (MSA-C type). There is no specific treatment for MSA until the present, only symptomatic innervations.

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