International Journal of Contemporary Pediatrics
Mude LN et al. Int J Contemp Pediatr. 2024 Feb;11(2):245-247
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20240105

Case Report

Hot cross bun in pediatric age multi-system atrophy an unusual presentation

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Received: 13 December 2023
Accepted: 11 January 2024

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ABSTRACT

A 10-year-old female was brought to emergency department with complaint fever, cold, and cough for 5 days followed by weakness of upper limb weakness more than Lower limb associated with inability to walk/stand, brought to emergency with altered sensorium. Nervous system: Confused and altered, tone is normal, power: 3/3 in upper limbs 2/2 in lower limbs and reflexes in bilateral knee brisk, bilateral plantar-extensor; On general examination revealed hypomimia, dysarthria and bilateral bradykinesia along with ataxic gait and pyramidal signs. Blood investigation were normal and diagnosis made by neuroimaging s/o**:** Bilaterally symmetrical abnormal signal in both postero-medial thalami, bilateral insular cortices, pons and bilateral middle cerebellar peduncles. It is extending into pons involving transverse pontocerebellar tracts and median pontine raphe nuclei giving 'Hot cross bun sign' (HCBS). During course of treatment child had autonomic disturbances. Child was treated with supportive medication and methyl-presdnisolone followed by oral steroid. Child had responded to treatment given and child has been discharged with no neurological deficit on oral medications. We concluded autonomic dysfunction in any patient presenting with acute onset of weakness with short duration must evaluate for MSA and institute appropriate treatment.

Keywords: Hot cross bun sign, Multisystem atrophy, Paresis, Neurology

INTRODUCTION

Multiple system atrophy (MSA) is a progressive neurodegenerative disorder characterized by various combinations of autonomic dysfunction, Parkinsonism, and cerebellar dysfunction. Autonomic dysfunction has a debilitating course in patients of MSA, compared to other neurodegenerative disorders presenting with parkinsonism or ataxia. We report a case of MSA presenting with weakness followed by paresis.[1]

CASE REPORT

A 10 year old female was brought to emergency with complaint of short duration of fever, cold, and cough for 5 days, followed by weakness of limbs. Upper limb weakness more than lower limb associated with inability to walk/ stand even with support, child was brought to emergency with altered sensorium and irrelevant behaviour without any episode of convulsion. On receiving GCS: E4V2M3 (9/15) altered sensorium her vitals were normal, blood pressure: 140/76 mmHg (95th-99th centile) bilateral pupil reactive. Nervous system: confused and altered, tone is normal in all four limbs power: grade 3/3 in upper limbs grade 2/2 in lower limbs and reflexes in bilateral knee brisk bilateral plantar: extensor; examination revealed hypomimia, dysarthria and bilateral bradykinesia along with ataxic gait and pyramidal signs. Child born out of non-consanguineous marriage with birth order of second with no significant positive history, with normal development milestone till onset of illness. Investigation done in view of acute demyelinating encephalon-myelitis, encephalitis, meningitis, fundus: normal; cerebrospinal fluid analysis turned negative culture report negative viral panel negative.

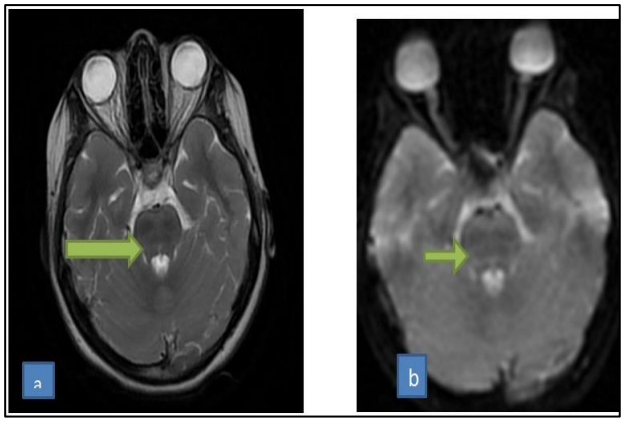

Figure 1 (A and B): Axial T2W image and DWI images of MRI brain of a patient of multisystem atrophy shows HCBS as a cruciform hyperintensity in an atrophied pons.

As a part of making diagnosis MRI BRAIN suggestive of bilaterally symmetrical abnormal signal in both postero-medial thalami, bilateral insular cortices, pons and bilateral middle cerebellar peduncles. It is extending into pons involving transverse pontocerebellar tracts and median pontine raphe nuclei giving 'HCBS'. Possibilities are viral encephalopathy or demyelinating lesions. No significant stenosis in proximal intracranial arteries of circle of Willis.[2]

Diagnosis made by neuroimaging, from the literature, it appears that the hot cross bun appearance on MR images may be seen in more than one disorder. However, in a case where a clinical diagnosis of MSA-C is suspected, the HCBS can provide helpful evidence to support this diagnosis. During course of treatment child had persistent hypertension for which anti hypertensives were given and her renal Doppler normal. Treatment given with oxygen support, IV fluids and antibiotics; methyl presdnisolone followed by oral steroid. Child had been discharged with no neurological deficit after a duration of 18 days on oral medications.

DISCUSSION

MSA is a rare, adult-onset, fatal neurodegenerative disease (ND) characterized by the association of parkinsonism with autonomic failure, cerebellar ataxia and pyramidal signs and with progressive loss of neuronal and oligo-dendroglial cells in various sites in the brain disease onset is usually around 50 years old or later, with the age distribution of onset peaking in the late 50s. MSA belongs to the group of alpha-synucleinopathies, which are morphologically characterized by abnormal accumulation of fibrillary alpha-synuclein in the neurons and oligodendrocytes.[3]

According to the predominant clinical characteristics, it can be further categorised as: MSA-P when parkinsonian clinical features predominate, MSA-C when cerebellar ataxia predominates and MSA-A when autonomic failure predominates.[4]

In MSA-C (historically known as olivopontocerebellar atrophy), we can observe radiological changes, mainly in T2-weighted sequences in brain MRI. Changes are mostly infratentorial, and include the typical hyperintense HCBS in the pons and other anomalies, such as putamen atrophy, putamen hypointensity and hyper intense putaminal rim.[5] MSA is mainly diagnosed based on clinical findings. Cranial magnetic resonance images may show putamen atrophy and infratentorial abnormalities such as the HCBS due to ponto-cerebellar degeneration. Early and severe autonomic dysfunction is a key feature of MSA, in the form of hypertension, urinary dysfunction, erectile dysfunction, constipation, vasomotor and thermoregulatory failure, and nocturnal inspiratory stridor. Hypertension manifests as recurrent syncope, light-headedness, weakness, nausea, tremulousness, headache, or pain in the neck and shoulder region (coat-hanger pain) on standing, but it may be asymptomatic.[6]

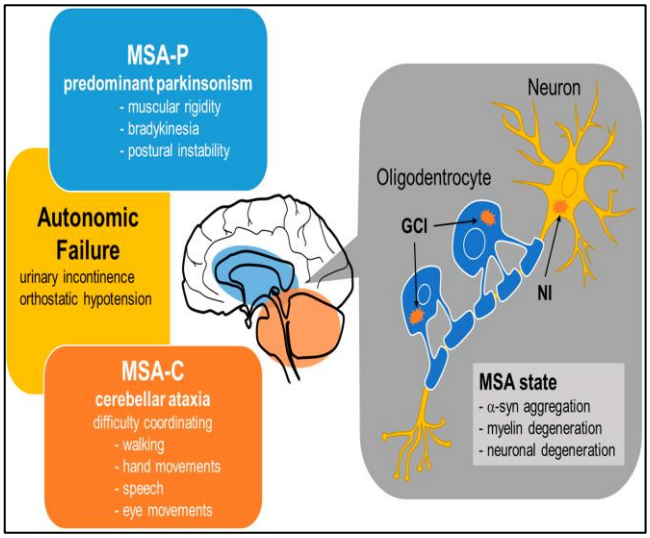

Figure 2: Types of multisystem atrophy with presenting symptoms.

The HCBS is the result of the degeneration of pontine neurons and transverse ponto-cerebellar fibres. This could be considered an expression of degeneration and neuronal loss in the brain stem, and serves as a parameter for measuring disease progression.[7] While the HCBS is typical, it is not pathognomonic to MSA HCBS is a radiologic finding describing a cruciform T2 hyper-intense signal on axial MRI of the pons. It is thought to represent gliosis of pontocerebellar fibers and is most commonly observed in patients with the cerebellar subtype of multiple system atrophy.[8] The pathological hallmark of MSA is glial cytoplasmic inclusion consisting of α-synclein. Autopsy shows widespread neuronal loss and atrophy of the striatonigral, cerebellar, autonomic, and corticospinal pathways in the central nervous system.[9] While the HCBS is typical, it is not

pathognomonic to MSA.[10] It has also been observed in cases of spinocerebellar ataxia (SCA) types 2 and 3, and in parkinsonism with cerebellar and brain stem alterations, presumably secondary to vasculitis.[11]

CONCLUSION

This case report emphasizes the need to recognize autonomic dysfunction in any patient presenting with acute onset of weakness with short duration of history along with autonomical disturbance and also clinicians must evaluate for MSA and institute appropriate treatment.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Mude LN, Shukla OS, Shah RH, Barad RR. Hot cross bun in pediatric age multi-system atrophy an unusual presentation. Int J Contemp Pediatr 2024;11:245-7.