Hot Cross Bun Sign – Multisystem Atrophy (Cerebellar Type)

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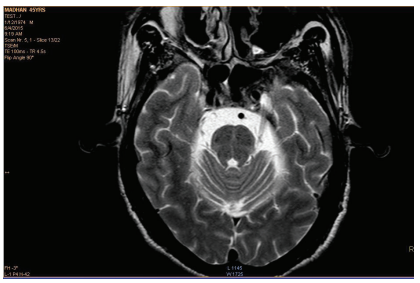

Fig. 1: MRI Brain (T2W Axial) showing typical Hot Cross Bun appearance of Pons

A 43 year old male presented with progressive difficulty in walking, slowness of speech, dizziness with tendency for frequent falls of 2 year duration. The patient was on syndopa and trihexyphenidyl with initial improvement for the first 6 months followed by a rapid deterioration. Later he developed urinary urgency, frequency and incontinence associated with constipation and postural giddiness. On examination, the patient had mask like facies, low volume of speech, bradykinesia, slow saccades, broken pursuits, cog wheel rigidity, symmetrical tremors, festinant gait with a tendency to fall associated with gait ataxia. His blood pressure was 140/90 mm in the lying down position which reduced to 110/74mm after 3 minutes of standing. Heart rate variability showed significant reduction in total power with sympathovagal balance showing sympathetic dominance. Cardiac autonomic function test showed severe autonomic dysfunction. Tilt table test was positive. MRI Brain (T2W Axial) demonstrated typical hot cross bun appearance of pons (Figure 1) which is suggestive of multi system atrophy-Cerebellar type. Patient was managed with dopamine agonists, liberal use of fluids, salts and fludrocortisone. Patient improved symptomatically and is on follow up.

Multi system atrophy is an adult onset, progressive neuro degenerative disease. It can be MSA- P if the features of parkinsonism predominate or MSA-C if the cerebellar symptoms predominate. It most commonly presents in the fifth decade of life. The clinical features can be broadly divided into autonomic dysfunction, parkinsonism, cerebellar dysfunction and corticospinal dysfunction of which extrapyramidal signs are most common.[1] Autonomic dysfunction can be either symptomatic or asymptomatic orthostatic hypotension due to deficient baroreflex regulation of sympathetic circuit. Detrussor hyperreflexia, urethral sphincter weakness and detrusor muscle weakness contributes to urinary symptoms in the form of frequency, urgency, nocturia, incomplete bladder emptying and incontinence.MRI abnormalities are typical of MSA. The Hot cross bun appearance in MSA is due to loss of pontine neurons and myelinated transverse pontocerebellar fibres with preservation of the corticospinal fibres which run craniocaudally[2]. It is seen in MSA- C while the putaminal slit sign is seen in MSA-P.

In conclusion, this case report serves to emphasize the need for a high index of suspicion to diagnose this rare parkinson plus syndrome.

References

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