Case report

Cerebellar type multiple system atrophy (MSA-C)

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Introduction

Multiple system atrophy (MSA) is a neurodegenerative disease characterized by abnormal deposition of the protein α-synuclein in the central and peripheral autonomic nervous system (synucleinopathy). MSA is classified in to Parkinsonian (MSA-P) and Cerebellar (MSA-C) sub types, depending on the predominant motor manifestation. We report a patient with MSA-C with characteristic clinical and radiological features.

Case history

A 61 year old house wife gave a history of progressive unsteadiness of gait for 5 years. She was unable to walk unaided over the last 2 years. There was no history of tremor, postural dizziness or breathing disorders.

Examination revealed normal cognitive functions and intact cranial nerves. Gait was broad based and ataxic needing bilateral assistance to walk. Severely impaired coordination was noticed bilaterally, with a left predominance. She had mild bilateral rest tremor, rigidity and minimal hypokinesis. Her recumbent blood pressure was 112/81 mmHg and dropped to 79/63 after maintaining erect posture for 3 minutes.

Her routine laboratory haematological and biochemical investigations including thyroid functions were normal. Non-contrast CT scan of the brain showed marked cerebellar atrophy (Figure 1). MRI scan of the brain showed pontine and cerebellar atrophy with the characteristic cruciform pontine hyperintensity giving rise to the "hot cross bun" sign (Figure 2). Tilt study confirmed dysautonomia but was negative for vaso vagal syncope. Levodopa treatment did not improve her extra pyramidal manifestations.

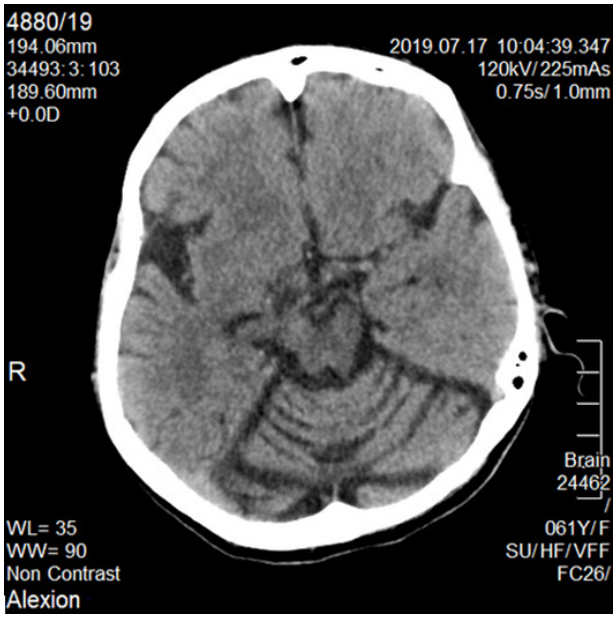

Figure 1. Non-contrast CT scan of the brain showing selective cerebellar atrophy.

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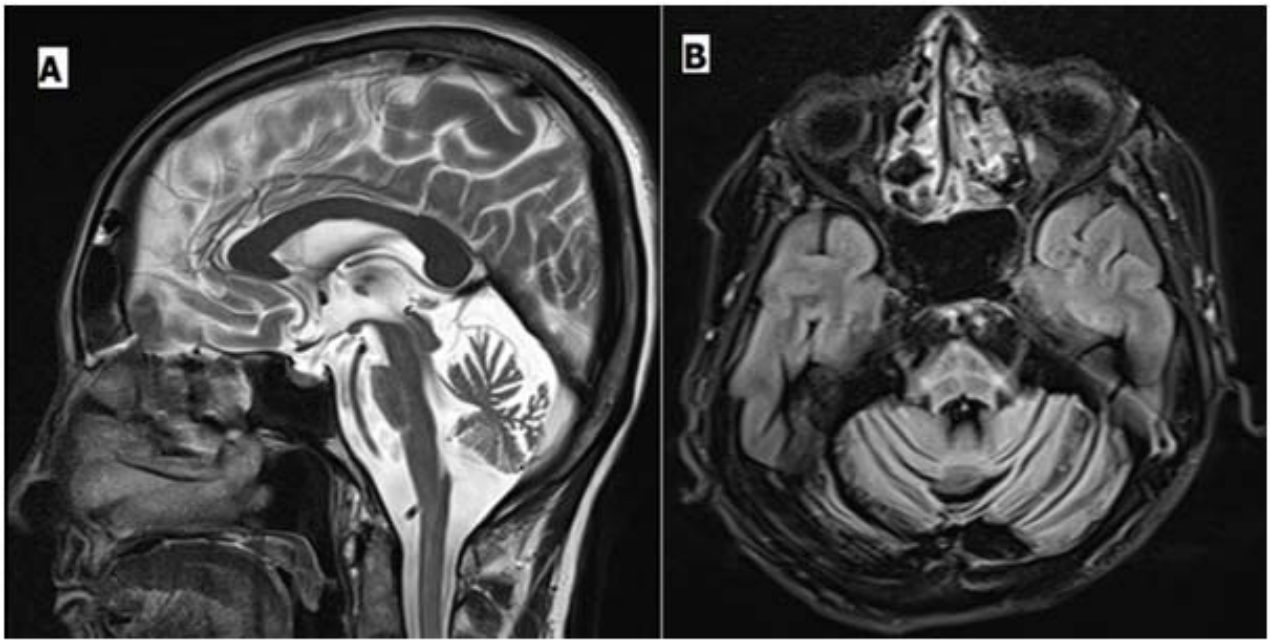

Figure 2. MRI brain T2 sagittal view showing pontine and cerebellar atrophy (A) and T2 axial FLAIR image view of the pons showing the "hot cross bun" sign (B).

Discussion

MSA is the most rapidly progressive synucleinopathy[1]. This patient satisfied the diagnostic criteria for probable MSA (Table 1). It is well recognised that patients with MSA have an initial phase of pure motor manifestation with an isolated Parkinsonism or cerebellar syndrome[3]. This can lead to a delay in the diagnosis. Isolated non-motor phase with pure autonomic failure is rare.

Radiological features are characteristic and aid the clinical diagnosis of MSA-C. These include selective atrophy of cerebellum, middle cerebellar peduncles and pons[4]. Selective loss of myelinated transverse ponto-cerebellar fibres and neurones in the pontine raphe with preservation of the pontine tegmentum and cerebral peduncles produce the "hot cross bun" sign. This sign may also be positive some sub types of Spino-Cerebellar Ataxia[4].

Table 1. Diagnostic criteria for probable multiple system atrophy[1]

Sporadic, progressive disease of adult onset (> 30 years old)
a) Autonomic failure involving urinary incontinence (inability to control the release of urine from the bladder with erectile dysfunction in males) or an orthostatic decrease of blood pressure within 3 min of standing by at least 30 mmHg systolic or 15 mmHg diastolic, ***and***
b) Poorly levodopa-responsive parkinsonism (bradykinesia with rigidity, tremor or postural instability), ***or***
c) A cerebellar syndrome (gait ataxia with cerebellar dysarthria, limb ataxia or cerebellar oculomotor dysfunction)

Conclusion

MSA should be suspected in any patient with otherwise unexplained cerebellar syndrome. Probable diagnosis of MSA could be made by careful evaluation of neurological status and autonomic functions. Characteristic MRI findings manifest early in patients with MSA-C than with alternative diagnoses.

References

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