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CASE REPORT

MULTI SYSTEM ATROPHY: REPORT OF TWO CASES

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ABSTRACT: Multiple system atrophy (MSA) is a sporadic, neurodegenerative disorder, clinically characterized by Parkinsonian, autonomic, cerebellar and pyramidal signs. We describe two patients showing different presentations of the same disease. The patient on case 1 presents features of MSA-C or olivopontocerebellar atrophy with the pontine "Cross sign" on brain MRI. The second case reports a patient presenting MSA-P or striatonigral degeneration and the brain MRI shows lenticular nucleus sign alteration. Multiple system atrophy (MSA) is a sporadic, neurodegenerative disorder, characterized by several combinations of parkinsonian, autonomic, cerebellar and pyramidal signs.[1-5]. The incidence is 0.6 cases/100000/year and the prevalence ranges from 1.86 to 4.9 cases/1000001. The term MSA defines a distinct clinicopathological entity, including olivopontocerebellar atrophy, striatonigral degeneration and Shy-Drager syndrome, which have been described as different diseases for many years and still lead to terminology confusion. The actual consensus tries to define the diagnostic criterions and recommends the term MSA-P when parkinsonian features predominate, and the term MSA-C when cerebellar signs rules the clinical picture. There is some doubt if the term Shy-Drager syndrome should be substitute, as most cases of MSA patients develop autonomic dysfunction during their course.[6] Horimoto et al.[7] believe that some patients present a distinct form of MSA in which dysautonomic symptoms predominate, considering the utility to classify that form as MSA-A. Recently, neuroimaging studies, especially the MRI, have showed some alterations that, although not specific, may help the diagnostic of different forms of MSA. We report two MSA cases with different clinical features and their characteristic MRI alterations.

KEYWORDS: Multi System Atrophy, Hot Cross Bun sign, Synucleopathy.

CASE REPORTS:

Case 1: 55 yrs. old woman complained of gait and speech alteration. On physical and neurological examination he presented slurred speech, gait ataxia and bilateral appendicular cerebellar ataxia. The deep tendon reflexes were +++/4 and there was bilateral Babinski sign. Arterial pressure (AP) was 130X80mmHg and radial pulse frequency 60bpm with the patient laying down; 100X70 and 70bpm when standing up. Brain MRI showed important pontine atrophy with the "Cross sign" (Fig. 1A) as well as cerebellar atrophy (Fig. 1B). There was no neurological disease in her family.

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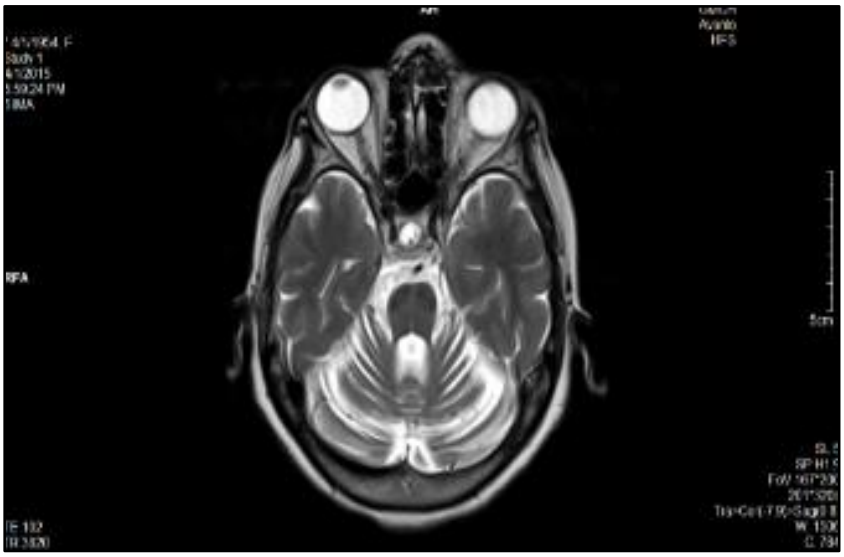

Fig. 1A

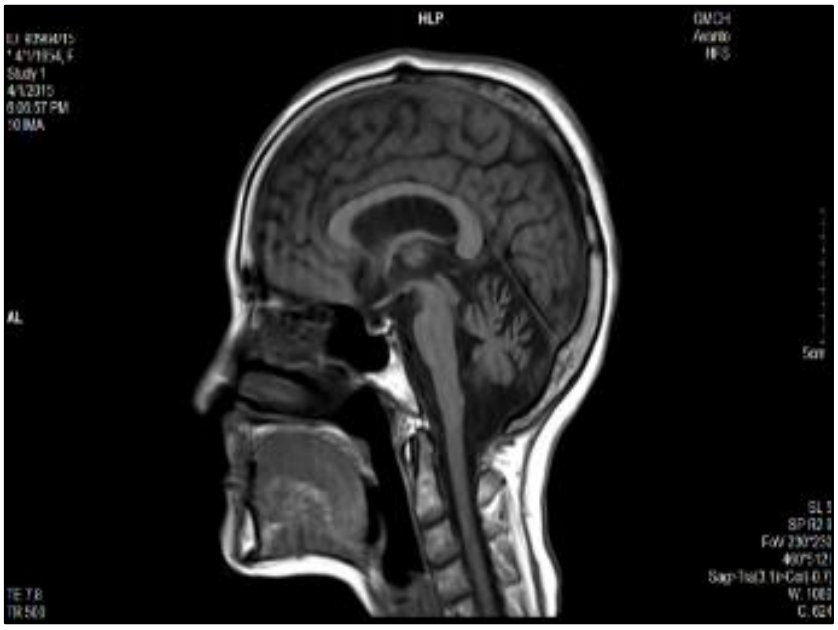

Fig. 1B

Case 2: A 64 year – old man presented with rigidity of arms. Levodopa started as the diagnosis of Parkinson's disease was made but there was no improvement. He reported urinary incontinence. There was progressive deterioration of daily activities and he required aid to walk. On general physical and neurological evaluation, she presented extrapyramidal rigidity, bradykinesia and some periods of akinesia. There was no rest or postural tremor, but the speech was slurred, very difficult to be understood. There was hyperreflexia and Babinski sign on both sides. AP laying down was 149X90 and seated was 90X 50mmHg. Brain MRI shows sign alteration on lenticular nucleus (Fig 2A and 2B). There was no neurological disease in her family.

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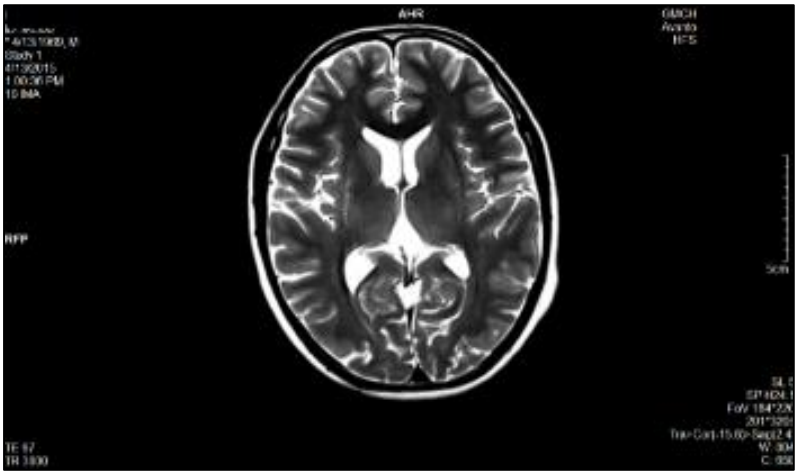

Fig. 2A

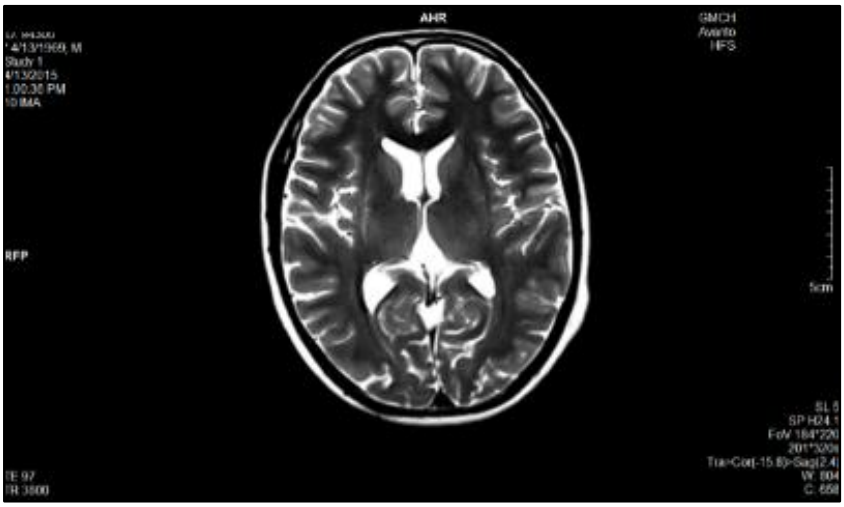

Fig. 2B

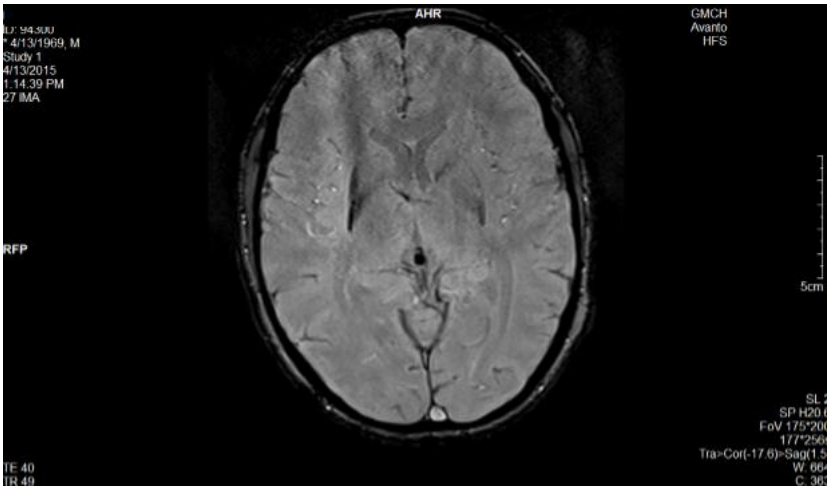

Fig. 2C

We report two patients presenting two different forms of MSA, although in the first case cerebellar syndrome was the main feature and in the second case, parkinsonian symptoms were predominant. Both of them had orthostatic hypotension and the second case also had urinary incontinence (Autonomic system alterations).

Watanabe et al.[8] evaluating the progression of MSA in 230 Japanese patients concluded that patients presenting MSA-P have a more rapid functional deterioration when compared to those with MSA-C. In the other hand, there was no difference in the survival time.

MRI is a useful diagnostic tool in the early course of MSA-C and MSA-P. Horimoto et al.[7] report that pontine "Cross sign" and lenticular nucleus sign alteration appears early in MSA-C and MSA-P respectively. Both of them appears lately in MSA-A. The characteristic T2 hyper intense sign in pons and middle cerebellar peduncle ("Cross sign") reflects pontocerebellar fibers degeneration and despite very suggestive of MSA it can be found in other forms of parkinsonism.[9] Asato et al.[10] have showed that the anteroposterior diameter of the inferior portion of the pons in MSA-C patients is lower when compared to patients in the control group or with progressive supranuclear palsy.

Putaminal abnormalities may be present in MSA-P patients MRI, other findings include hypo intense sign of the putamen with marginal hyper intense sign in T2. Atrophy or hyper intense sign at the pons, middle cerebellar peduncle and cerebellum may be seen. Putaminal atrophy is the most specific finding in MSA-P.[9] Our case number 2 presented putaminal hypo intensity as well as marginal hyper intensity in T2 images.

In conclusion, our cases are classified as likely MSA according to criteria in consensus, since the diagnosis of MSA is defined just with pathological analysis.[6] in the two cases, we try to contribute to the importance of a good MRI interpretation. We concluded that the brain MRI changes might increase the accuracy diagnosis of MSA.

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