

Short Communication

Asymmetry of Symptoms and Signs in Multiple System Atrophy

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Abstract

It has been widely recognized that some of the symptoms and signs of multiple system atrophy (MSA) emerge with laterality, but only a few studies have evaluated this issue comprehensively. Small-scale case reports that have been published thus far suggest that laterality in MSA is a rather rare finding.

In order to examine the prevalence and characteristics of laterality in symptoms and signs in MSA, we retrospectively investigated the records of 155 MSA patients. Among all the patients, 39% had episodes that suggested the presence of laterality in their illness history.

The MSA with predominant parkinsonism (MSA-P) patients presented with more episodes of laterality than the MSA with cerebellar features (MSA-C) patients. Among the episodes, laterality was more frequent in parkinsonism than for the cerebellar symptoms. Interestingly, these symptoms were more prevalent at the left side of the body. Laterality in the cerebellar signs was present in 63% of the MSA-C patients and laterality in parkinsonism was exhibited by 64% of the MSA-P patients. Interestingly, cerebellar signs, parkinsonism, and Horner's sign were all observed predominantly at the left side.

Thus, we concluded that a significant number of MSA patients present with laterality in both symptoms and signs and that more interestingly, these are predominantly observed at the left side.

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Introduction

Multiple system atrophy (MSA) is a sporadic neurodegenerative disease characterized by cerebellar ataxia, parkinsonism, and autonomic dysfunction with a middle age onset^[1]. The disease affects both sexes and is more prevalent in males. It has been widely recognized that some of the symptoms and signs of MSA emerge with asymmetry, and small-scale case reports that have been published thus far suggest that asymmetry in MSA is a rather rare finding^[2,3]. However, in daily practice we notice apparent asymmetry of symptoms and signs in MSA. Thus we tried to evaluate this issue by reviewing past cases.

Methods

In order to examine the prevalence and characteristics of asymmetry in MSA, we retrospectively investigated the records the inpatients' records between 1966 and 2010, we selected 152 patients who satisfied the criteria of probable, and definite MSA in the course of follow-ups according to the Consensus statement on the diagnosis of MSA^[4]. There were 108 MSA with cerebellar features (MSA-C) and 44 MSA with predominant Parkinsonism (MSA-P) patients. The overall male to female ratio was 1.49, and the mean age of onset was 56.2 ± 7.93 years.

Result and Discussion

Among the patients diagnosed with MSA, 39% had episodes such as clumsiness in limb movement, tremors, or a tendency to lean or fall to a certain side during walking that suggested the presence of asymmetry in their illness history. The MSA-P patients presented with more episodes of asymmetry than the MSA-C patients (75% vs. 23.6%,

respectively). Among the episodes, asymmetry was more frequent in Parkinsonism than for the cerebellar symptoms (56% vs. 33%, respectively). Interestingly, these symptoms were more prevalent at the left side of the body (64% for parkinsonism and 83% for cerebellum-related symptoms). Next, we surveyed the neurological signs examined by board certified neurologists. The overall result is shown in table 1.

Asymmetry in the cerebellar signs was present in 63% of the MSA-C patients and asymmetry in parkinsonism was exhibited by 64% of the MSA-P patients. Interestingly, cerebellar signs, parkinsonism, and Horner's sign were all observed predominantly at the left side (cerebellar signs: left, 52%; right, 12%; and bilateral, 36%; parkinsonism: left, 35%; right, 18%; and bilateral, 47%; and Horner's sign: left, 62%, and right, 38%). Pyramidal signs were observed without any asymmetry (left, 13%; right, 11%; and bilateral, 76% if present). Thus, we concluded that this left-side predominance was related to the handedness of the patients; since our two left-handed patients in fact presented with marked right-side-dominant parkinsonism. In 99 cases 123I-IMP-single photon emission computed tomography (SPECT) images were obtained. Trained radiologists evaluated the images. Among the images, 20% were evaluated to be left side dominantly affected, while 12% were evaluated as right side dominant in terms of cerebellar perfusion. Interestingly only 50% of SPECT laterality evaluation matched with laterality in clinical symptoms. This result suggests that in significant numbers of cases, handedness might affect the emergence of the symptoms thus left limbs of right handed patients are more vulnerable than right limbs.

Conclusion

Thus, we concluded that a significant number of MSA patients

diagnosis	MSA-C			MSA-P			all MSA			
number of patients	108			44			152			
male / female	67/41			24/20			91/61			
age at onset (yo ± SD)	55.8±8.14			57.2±7.36			56.2±7.93			
handedness right/left	108/0			42/2			150/2			
initial symptom	autonomic and urinary dysfunction	10			5			15		
	Parkinsonism	0			33			33		
	cerebellar dysfunction	83			2			85		
	others	15			4			19		
neurological signs at diagnosis % (#of patients)	autonomic and urinary dysfunction	94.4 (102)			95.5 (42)			94.7 (144)		
	Parkinsonism	71.3 (77)			100 (44)			79.6 (121)		
	cerebellar dysfunction	100 (108)			54.5 (24)			86.8 (132)		
	corticospinal tract dysfunction	90.7 (98)			93.2 (41)			91.4 (139)		
laterality		left	right	bilateral	left	right	bilateral	left	right	bilateral
	Horner's sign	3	0		2	3		5	3	
	Parkinsonism	21	15	41	21	7	16	42	22	57
	cerebellar sign	56	13	39	9	2	13	65	15	52
	corticospinal dysfunction	15	11	72	3	5	33	18	16	105
MRI cross sign / all MRI	73/74			19/23			92/97			

Table 1: Review of past cases

present with asymmetry in both symptoms and signs and that more interestingly, these are predominantly observed at the left side. We hope that this finding will be useful in diagnosing MSA and understanding its pathomechanism.

Reference

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