Original Article

The High Value of External Anal- and Urethral-Sphincter Electromyography in Differential Diagnosis with MSA-P, PD, and PSP

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Abstract

Objective: It is a challenge to differentiate multiple system atrophy parkinsonism (MSA-P), Parkinson's disease (PD), and progressive supranuclear palsy (PSP). We aimed to explore the value of external anal-sphincter electromyography (EAS-EMG) and urethral-sphincter electromyography (US-EMG) in differential diagnosis with MSA-P, PD, and PSP. **Methods:** A total of 149 subjects, including 27 MSA-P, 100 PD, and 22 PSP, were recruited. The average duration and amplitude of motor unit potentials (MUPs), percentage of polyphasic MUPs, amplitude during strong contraction, and recruitment pattern during maximal voluntary contraction were recorded. The differences in EAS-EMG and US-EMG results between MSA-P, PD, and PSP were analyzed. **Results:** In EAS-EMG examination, the average duration of MUPs of MSA-P was significantly longer than that of PD and PSP; the percentage of polyphasic MUPs and the ratio of simple phase and simple-mix phase of MSA-P and PSP were significantly higher than that of PD; the amplitude during strong contraction of MSA-P was significantly lower than that of PD. In US-EMG examination, the average duration of MUPs in male MSA-P was significantly longer than that in male PD and PSP; the ratio of simple phase and simple-mix phase in male MSA-P was significantly higher than that in male PD; there was no statistical difference in US-EMG indexes between male PD and PSP male. And because only one female PSP was examined, only female MSA-P and PD were compared, the average duration of MUPs in female MSA-P was significantly longer than that in female PD; the ratio of simple phase and simple-mix phase in female MSA-P was significantly higher than that in female PD. **Conclusion:** The average duration of MUPs and the ratio of the simple phase and simple-mix phase of EAS-EMG and US-EMG all can provide the basis for the differential diagnosis between MSA-P and PD. US-EMG can be used as a supplement to differentiate MSA-P from PD when EAS-EMG is limited. The only discriminating indicator between MSA-P and PSP seems to be the average duration of MUPs of EAS-EMG and US-EMG. There is still a lack of diagnostic electromyography indicators between PD and PSP.

Keywords: EAS-EMG, multiple system atrophy, Parkinson's disease, progressive supranuclear palsy, US-EMG

Introduction

Multiple system atrophy Parkinsonism (MSA-P), Parkinson's disease (PD), and progressive supranuclear palsy (PSP) are all neurodegenerative disorders.[1] At present, the diagnosis and differential diagnosis of MSA-P, PD, and PSP mainly rely on presentation, symptoms, and history. Owing to the clinical manifestations of the three overlap,[2] it is difficult to differentiate the three for clinicians. Early accurate diagnosis is very important for patient management and treatment selection. Neurodegenerative diseases such as MSA-P and PSP often require a neuropathological examination to confirm the diagnosis,[3] but biopsy is difficult and risky, and it is difficult to promote the clinical application. Therefore, it is urgent to obtain simple and practical examination methods to provide help for the differential diagnosis among MSA-P, PD, and PSP. Since external anal-sphincter electromyography (EAS-EMG) was first used in the examination of neurological diseases in 1978,[4] its diagnostic value for MSA has been gradually recognized. The study data of Lee *et al.*[5] showed that abnormal EAS-EMG was observed in 90% of MSA patients. Our previous studies also found that both EAS-EMG and urethral-sphincter electromyography (US-EMG) have a high value in the differential diagnosis of MSA.[6] In our study, we observed the characteristics of EAS-EMG and US-EMG

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among the MSA-P, PD, and PSP, and analyze differential diagnostic values for the three.

Methods

Patients

Patients of MSA-P, PD, and PSP were recruited from inpatients and outpatients, and interviewed by two experienced neurologists from the Department of Neurology of the Sixth Medical Center of Chinese PLA General Hospital. This study was approved by the Ethics Committee of the Sixth Medical Center of Chinese PLA General Hospital and carried out in accordance with the Helsinki Declaration. All subjects received written informed consent. Our study was approved by the ethics committee. The approval date was November 6, 2014.

Inclusion criteria

(1) Gender not confined; (2) MSA-P patients were enrolled according to Gilman's MSA diagnostic criteria in 2008[7]; (3) the diagnostic criteria of PD patients are according to the clinical diagnostic criteria of PD Association brain Bank (UKPDBB, international general diagnostic standard)[8] and Parkinson's disease and dyskinesia group of Neurology Branch of Chinese Medical Association in 2009, Chinese guidelines for the treatment of Parkinson's disease [second edition][9]; (4) the inclusion criteria of PSP patients were the clinical diagnostic criteria of progressive supranuclear palsy in China.[10]

Exclusion criteria

(1) It meets the exclusion criteria in Gilman's MSA diagnostic criteria, the UK Parkinson's Disease Association Brain Bank (UKPDBB) PD diagnostic criteria, and the Chinese clinical diagnostic criteria for progressive supranuclear palsy; (2) those who have primary mental disorders, such as consciousness disorder or severe cognitive dysfunction, cannot cooperate with the study; (3) patients with perianal or local infection around the urethra who cannot cooperate with EMG examination; (4) patients with sphincter dysfunction syndrome such as urinary and stool disorders caused by sacral plexus disease; (5) patients with severe diabetic polyneuropathy.

Electrophysiological examination

Instruments and parameters

Keypoint v 3.04 electromyography/evoked potentiometer of Dandi Company. The filter band is 20 H ~10 kHz. The scanning speed of the amplitude of the unit potential of motion is 5 ms/D, the sensitivity is 100 μv/D for light contraction, the sensitivity is 0.5 mv/D for strong contraction, and the scanning speed is 200 ms/D. In the examination room where the environment is quiet, the subjects take a comfortable seat and the room temperature is 20°C ~ 29°C.

The examination method of EAS-EMG

According to the anatomical structure of the anal sphincter, the patient was in the left-lying position. After perineal disinfection, relaxation, bending the knee, and bending the hip, the examiner and the patient cooperate to separate the two buttocks, at the junction of the skin and mucosa of 10mm in the lateral and posterior part of the anus (about 4 o'clock), the concentric needle electrodes were inserted into the external anal sphincter at an acute angle. Record: the average duration of 20 different MUPs, the average amplitude of MUPs, the percentage of polyphasic MUPs, the amplitude during strong contraction, and the recruitment pattern during maximal voluntary contraction.

The examination method of US-EMG

For male patients, a concentric needle electrode of length 50 mm and diameter 0.45 mm was inserted into the middle of the anal-bulbar cavernous muscle connection. For female patients, a concentric needle electrode of length 75 mm and diameter 0.65 mm was inserted vertically into the urethral sphincter at 5 mm away from the external orifice of the urethra. The patients were instructed to hold back urination and urinate, respectively, and the myoelectric activity of the external urethral sphincter was recorded like EAS-EMG.

Statistical analysis

The differences between different groups of variables were analyzed by using SPSS26.0 statistical software. The counting data were compared by Chi-square test or Fisher exact test, expressed in the form of n (%). The Shapiro–Wilk test method was used to test the normality of the measurement data. If the measurement data met the normality, the *t* test was used to compare the measurement data of the two groups, and the analysis of variance was used to compare the measurement data of the three groups, indicating a mean ± standard deviation ($\bar{x} \pm s$). The measurement data did not meet the normality. The Mann–Whitney test method was used to compare the measurement data of the two groups, the Kruskal–Wallis test method was used to compare the measurement data of the three groups, and the Bonferroni correction method was used for pairwise comparison. The median (interquartile range) was expressed in the form of M (P25, P75).

Results

Sample feature

Three groups of patients participated in our research: 27 MSA-P, 100 PD, and 22 PSP. There were 8 male patients and 19 female patients in the MSA-P group, 65 male patients and 35 female patients in the PD group, and 16 male patients and 6 female patients in the PSP group. There were significant differences in gender among the three groups ($P < 0.05$). The average age of patients in the MSA-P group, PD group, and PSP group was 60.63 ± 9.26, 64.70 ± 9.96, 65.64 ± 7.73 years, respectively. There was no significant difference in age among the three groups ($P > 0.05$). The median course of the disease was 2.4 (2.0,4.0) years in the MSA-P group, 3.0 (1.1,5.0) years in the PD group, and 3.0 (1.8,4.0) years in the PSP group. There was no significant difference in the course of disease among the three groups ($P > 0.05$) [Table 1].

Results of EAS-EMG in patients with MSA-P, PD, and PSP

A total of 26 MSA-P patients, 100 PD patients, and 22 PSP patients participated in the EAS-EMG examination, of which one MSA-P patient was excluded due to perianal hemorrhoids and abscesses. There were significant differences in the average duration of MUPs, percentage of polyphasic MUPs, amplitude during strong contraction, and recruitment pattern during maximal voluntary contraction of EAS-EMG among the three groups ($P < 0.05$). There was no significant difference in the average amplitude of MUPs ($P > 0.05$). The average duration of MUPs of EAS-EMG in the MSA-P group was significantly longer than that in the PD and PSP groups [Figure 1]. The percentage of polyphasic MUPs and the ratio of simple phase and simple-mix phase during maximal contractions in the MSA-P and PSP groups were significantly higher than those in the PD group, and the amplitude during strong contractions in MSA-P was significantly lower than that in the PD group [Table 2].

Results of US-EMG in male patients with MSA-P, PD, and PSP

A total of seven male MSA-P patients, 64 male PD patients, and 14 male PSP patients participated in the US-EMG examination, while one male MSA-P patient, one male PD patient, and two male PSP patients did not complete the US-EMG examination due to pain or inability to cooperate. There were statistically significant differences in the average duration of MUPs and the ratio of simple phase and simple-mix phase of US-EMG among male patients ($P < 0.05$). There was no significant difference in the amplitude of MUPs, the percentage of polyphasic MUPs, and the amplitude during strong contraction ($P > 0.05$). The average duration of MUPs of US-EMG in the MSA-P group was significantly longer than that in the PD and PSP groups [Figure 2]. The ratio of simple phase and simple-mix phase in the MSA-P group was significantly higher than that in the PD group. There was no statistical difference in us-EMG indicators in male PD and PSP patients [Table 3].

Table 1: Gender compositions, ages, and courses of disease between MSA-P, PD, and PSP groups

Group	*n*	Gender		Age, year	Course of disease, year
		Male	Female		
MSA-P	27	8 (29.6)	19 (70.4)	60.63±9.26	2.4 (2.0,4.0)
PD	100	65 (65.0)	35 (35.0)	64.70±9.96	3.0 (1.1,5.0)
PSP	22	16 (72.7)	6 (27.3)	65.64±7.73	3.0 (1.8,4.0)
$\chi^2/F/H$		12.870		2.268	0.388
P		❶0.002		❷0.107	❸0.824

❶Kruskal-Wallis test used for analysis because data were not of the standard normal distribution; ❷Pearson χ^2; ❸Analysis of Variance. MSA-P, multiple system atrophy Parkinsonism; PD, Parkinson's disease; PSP, progressive supranuclear palsy

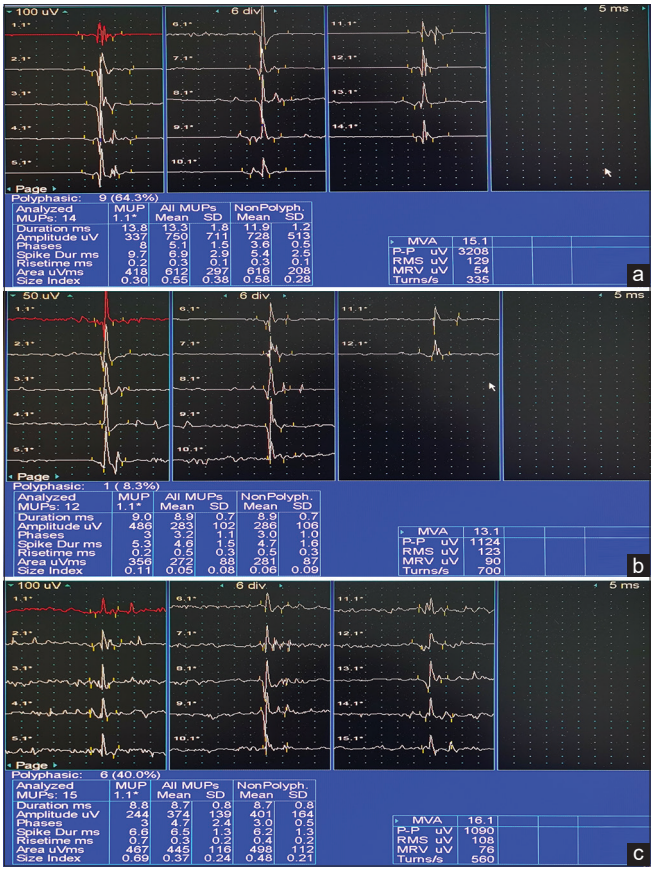

Figure 1: EAS-EMG of patients with MSA-P, PD, and PSP. Notes: (a) EAS-EMG of a patient with MSA-P. (b) EAS-EMG of a patient with PD. (c) EAS-EMG of a patient with PSP. EAS-EMG examination showed that the average duration of MUPs of in the MSA-P group was significantly longer than that in PD and PSP patients. Abbreviations: EAS-EMG, external anal-sphincter electromyography; MSA-P, multiple system atrophy characterized by Parkinson's syndrome; PD, Parkinson's disease; PSP, progressive supranuclear palsy; MUPs, motor unit potentials

Table 2: EAS-EMG results between MSA-P, PD, and PSP groups

	MSA-P *n*=26	PD *n*=100	PSP *n*=22	Statistics	*P*
MUP duration, ms	12 (10.8,12.7)[b]	9.8 (9.4,10.6)[a]	9.9 (9.2,11.2)[a]	H=30.229	❶<0.001
MUP amplitude, μV	463.5 (352.3,625.0)	481.0 (350.0,586.8)	446.5 (384.5,614.8)	H=0.051	❶0.975
Percentage of polyphasic MUPs, %	40.3 (30.8,60.0)[b]	31.6 (22.9,37.0)[a]	38.3 (28.0,43.0)[b]	H=15.727	❶<0.001
Amplitude during strong contraction, mV	1.0 (0.8,1.3)[a]	1.5 (1.0, 2.0)[b]	1.2 (0.7, 1.5)[ab]	H=10.467	❶0.005
Ratio of simple phase and simple-mix phase, %	38.5 [b]	9.0 [a]	36.36 [b]	χ^2=16.983	❶<0.001

❶Kruskal-Wallis test used for analysis because data were not of the standard normal distribution; ❷Fisher exact test. EAS-EMG, external anal-sphincter electromyography; MUPs, motor unit potentials; MSA-P, multiple system atrophy Parkinsonism; PD, Parkinson's disease; PSP, progressive supranuclear palsy. Different lowercase letters indicate statistically significant differences between groups

Table 3: Male US-EMG results between MSA-P, PD, and PSP groups

	MSA-P $n=7$	PD $n=64$	PSP $n=14$	Statistics	P
MUP duration, ms	12.9 (11.1,13.7)[b]	9.8 (9.4,10.7)[a]	10.4 (9.2,11.3)[a]	H=13.311	[❶]0.001
MUP amplitude, μV	421.0 (306.0,580.0)	351.5 (278.8, 461.0)	385.0 (291.8, 591.3)	H=2.637	[❶]0.267
Percentage of polyphasic MUPs, %	36.3 (20.0, 43.2)	20.0 (15.4, 36.2)	20.0 (14.9, 28.2)	H=2.78	[❶]0.249
Amplitude during strong contraction, mV	0.8 (0.7,1.9)	1.1 (0.6,1.8)	1.0 (0.5,2.0)	H=1.094	[❶]0.579
Ratio of simple phase and simple-mix phase, %	57.1[b]	10.9[a]	14.3[a,b]	χ^2=7.956	[❶]0.013

[❶]Kruskal-Wallis test used for analysis because data were not of standard normal distribution; [❶]Fisher exact test. US-EMG, urethral-sphincter electromyography; MUPs, motor unit potentials; MSA-P, multiple system atrophy Parkinsonism; PD, Parkinson's disease; PSP, progressive supranuclear palsy. Different lowercase letters indicate statistically significant differences between groups

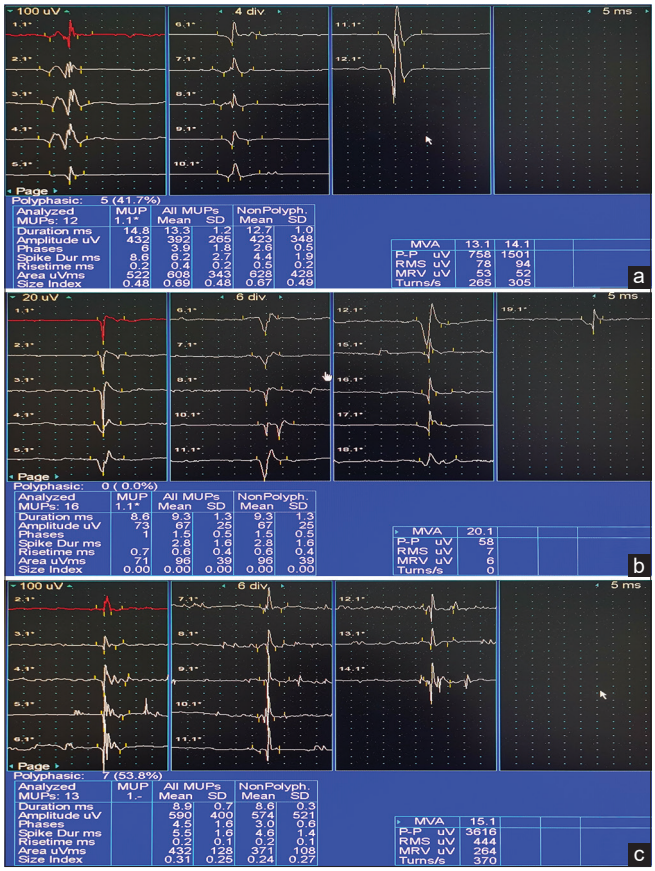

Figure 2: US-EMG of patients with MSA-P, PD, and PSP. Notes: (a) US-EMG of a patient with MSA-P. (b) US-EMG of a patient with PD. (c) US-EMG of a patient with PSP. US-EMG examination showed that the average duration of MUPs of in the MSA-P group was significantly longer than that in PD and PSP patients. Abbreviations: US-EMG, urethral-sphincter electromyography; MSA-P, multiple system atrophy characterized by Parkinson's syndrome; PD, Parkinson's disease; PSP, progressive supranuclear palsy; MUPs, motor unit potentials

Results of US-EMG in female patients with MSA-P and PD

A total of 15 female MSA-P patients, 32 female PD patients, and one female PSP patient participated in the US-EMG examination, while four female MSA-P patients, three female PD patients, and five female PSP patients did not complete US-EMG due to pain or inability to cooperate. The sample size of female PSP patients participating in US-EMG was too small to be included in the statistical analysis. The average duration of MUPs in the MSA-P group was significantly longer than that in the PD group. The ratio of simple phase and simple-mix phase in the MSA-P group was significantly higher than that in PD the group, while other indexes showed no statistical difference [Table 4].

Discussion

With the aging of the world's population, the incidence of senile degenerative diseases such as MSA-P, PD, and PSP is increasing day by day. It is difficult for the three to make an accurate diagnosis in the early stage of the disease based on the medical history, clinical symptoms, physical examination of the nervous system, and imaging manifestations, and they often misdiagnose each other.[11] The process of MSA disease can start from the sacral spinal cord, and then spread to other regions causing motor disorders and cardiovascular autonomic dysfunction,[12] so electrophysiological abnormalities appear earlier. Our previous studies found that EAS-EMG and US-EMG have clinical application value in the diagnosis and differential diagnosis of MSA,[13] and also calculated the specificity and sensitivity of each indicator in the differential diagnosis of MSA.[14]

The abnormal EAS-EMG and US-EMG were caused by damage to the sacral medullary Onuf's nucleus, which was first identified and named by Onufrowicz and whose main function was the sphincter innervating the anus and urethra.[15] Onuf's nucleus was a specific affected site of MSA.[16] When Onuf's nucleus in the anterior sacral horn is damaged, it shows signs of neurogenic damage such as prolonged duration, increased polyphase wave, and spontaneous potential or satellite potential on EAS-EMG and US-EMG.[17] Some researchers[18] found that there was no statistical significance in the differences in EAS-EMG indexes between MSA-P and MSA-C patients, suggesting that the damage of Onuf's nucleus in MSA-P and MSA-C was similar. PD and PSP can also have anal- and urethral-sphincter damage similar to MSA, but the occurrence rate is lower than MSA. The damage degree to the sacral medullary Onuf's nucleus in PD, PSP, and MSA patients increases from mild to severe, and for the same disease, the degree of damage to Onuf's nucleus may also be related to the course of the disease.[19] The more serious the electromyography changes are, the more likely the diagnosis of MSA will be, and the worse the prognosis

Table 4: Female US-EMG results between MSA-P and PD groups

	MSA-P $n=15$	PD $n=32$	Statistics	P
MUP duration, ms	11.5 (10.0, 13.0)	9.8 (9.2, 10.3)	$Z=3.45$	[b]0.001
MUP amplitude, μV	269.0 (226.0, 294.0)	233.0 (156.5, 286.5)	$Z=1.141$	[b]0.254
Percentage of polyphasic MUPs, %	26.5 (14.4, 53.3)	10.0 (7.1, 18.2)	$Z=1.852$	[b]0.064
Amplitude during strong contraction, mV	0.5 (0.5,0.7)	0.6 (0.5,0.8)	$Z=0.447$	[b]0.655
Ratio of simple phase and simple-mix phase, %	60.0	9.4	$\chi^2=13.766$	[b]0.001

[b]Mann-Whitney U test was used for analysis because data were not of the standard normal distribution; [b]Fisher exact test. US-EMG, urethral-sphincter electromyography; MUPs, motor unit potentials; MSA-P, multiple system atrophy Parkinsonism; PD, Parkinson's disease

of patients will be.[20] In our study, there was no statistical difference in the course of disease among the three groups. According to the average duration of MUPs in EAS-EMG and US-EMG, MSA-P had the longest average duration of MUP, followed by PSP and PD, which was consistent with the results of previous studies. Different studies have reached different conclusions as to whether age affects the outcomes of EAS-EMG and US-EMG.[21,22] We found no significant linear relationship between the duration of EAS-EMG and US-EMG and age in the three groups of disease. Winge *et al.*[23] showed that the average duration of anal sphincter MUPs in MSA patients was significantly longer than that in PD patients. Yamamoto *et al.*[24] believed that the average duration of MUPs of EAS-EMG is the most appropriate indicator to distinguish MSA from PD. Similar to previous studies, our study found that the average duration of MUPs of both EAS-EMG and US-EMG was of great value in differentiating MSA-P from PD. Some scholars believed that EAS-EMG could not distinguish between MSA-P and PSP,[25] but our study found that the average duration of MUPs of EAS-EMG and US-EMG for MUP in MSA-P patients was statistically different from that in PSP patients, which has potential significance for differential diagnosis.

Rodi *et al.*[26] suggested that the percentage of polyphasic MUPs >60% can be used to distinguish MSA from PD. Some researchers[27] showed that the percentage of EAS-EMG multiphase waves ≥40% indicates neurogenic damage. In our study, in the three groups of patients with MSA-P, PD, and PSP, the percentage of polyphasic MUPs was 0 in two cases in each group, which may be caused by poor patient coordination, insufficient contraction strength, and poor insertion site, resulting in the single release of motor units. When the percentage of polyphasic MUPs of EAS-EMG was 0 and was removed, the statistical results showed that the percentage of polyphasic MUPs of EAS-EMG of MSA-P and PSP was significantly higher than that of PD, and the difference was statistically significant. In addition, the proportion of neurogenic lesions in the MSA-P, PD, and PSP groups was 50%, 15.3%, and 45%, respectively, based on the percentage of polyphasic MUPs, which was similar to the average duration of MUPs. Similar to the results of previous studies,[14] EAS-EMG showed that the amplitude during strong contractions of MSA-P was significantly lower than that of PD, and the ratio of simple phase and simple-mix phase of MSA-P was significantly higher than that of PD, with statistically significant differences.

Considering the differences in the anatomical structure of male and female urethral sphincters and the differences in US-EMG examination methods between males and females, we compared MSA-P, PD, and PSP groups by sexes in order to reduce the impact of such differences on the results. The results showed that US-EMG could supplement EAS-EMG in the differential diagnosis of MSA-P and PD. For differential diagnosis of MSA-P and PSP in male patients, US-EMG can be used as a supplement to EAS-EMG. However, there is a lack of evidence of US-EMG value between MSA-P and PSP in females. There were still some deficiencies in our study. Due to the lower incidence of MSA-P in the Asian population compared with MSA-C,[28] the sample size of MSA-P in this study was small. In addition, the results of the US-EMG examination in female PSP also needed to be further studied by expanding the sample size.

Conclusion

Both EAS-EMG and US-EMG contribute to the early differential diagnosis of MSA-P from PD and PSP, especially the average duration of MUPs, in the absence of clear clinical and radiographic features. When EAS-EMG is not available, US-EMG can be used as a supplementary method for the differentiation of MSA-P from PD and male PSP. Unfortunately, there is still a lack of valuable electromyography indicators for the differentiation of PD and PSP.

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Conflicts of interest

There are no conflicts of interest.

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