SHEAR WAVE ELASTOGRAPHY IN PATIENTS WITH SPINAL MUSCULAR ATROPHY TYPE 2-3

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INTRODUCTION

Spinal muscular atrophy (SMA) is characterized by proximal muscle weakness and atrophy resulting from progressive degeneration and irreversible loss of the anterior horn cells in the spinal cord[1]. In studies segmental distribution of muscle involvement in SMA, different clinical effects of the upper and lower extremity proximal muscles. In the upper extremities early involvement of the triceps suggests that the disease starts at about the level of C7, and in the lower extremities early and severe involvement of the iliopsoas suggests that the dissease effected firstly L2-4 upper lumbar segments[2,3]. Why the spinal segment is affected particularly is vulnarable and important for the treatment methods. SWE is a non-invasive, ultrasound-based imaging technique which provides quantitative information regarding tissue stiffness has become popular and gained increasing attention for the evaluation of muscle stiffness. Also, due to the possibility of the quantitative evaluation SWE has a promising role in determining the severity of disease and treatment follow-up, monitoring age-related changes of various neuromuscular diseases[4].

OBJECTIVES

This study aimed to investigate selective muscle involvement by shear wave elastography (SWE) in patients with spinal muscular atrophy (SMA) types 2 and 3 and determine the diagnostic contributions of SWE and magnetic resonance imaging (MRI).

MATERIALS & METHODS

Seventeen SMA type 2-3 patients were included in the study. The Medical Research Council (MRC) scale was used for grading muscle power, ranging from 0 to 5. SWE was performed using a high-frequency linear transducer (frequency range, 8–14 MHz) of an ultrasound device (Canon Medical Systems, Aplio 500 Platinum, Japan) within one month of the MRI scan. Spinal MRI performed that requested for possible complications before intrathecal nusinersen treatment on a 1.5-Tesla MR scanner. A 4-point rating scale evaluating the amount of fatty degeneration and atrophy level of muscle bulk.

RESULTS

Seventeen patients were included in the study. Their mean age 10.9 ± 5.5 (3.5-18.4) years and 9 (52.9%) of the patients were girls and 8 (47.1%) were boys. Nine were SMA (52.9%) type 2, 8 (47.1%) type 3.

Muscle Strength: MRC grades were significantly lower in the iliopsoas than gluteus maximus (p<0.001) and in the triceps brachii than biceps brachii (p:0.003).

Comparison of SWE Values Between Muscles: Among the upper extremities, median SWE values were found to be highest (it means also more severe involvement) in triceps brachii, however no significant difference was observed between triceps brachii, biceps brachii and deltoid muscles (p:0.23). In post-hoc analysis, a significant difference was observed in SWE values between triceps-biceps in pairwise group comparisons too (p:0.003). Among the lower extremities muscles, SWE values of the iliopsoas were significantly higher than the gluteus maximus (p<0.001).

Relation of SWE to Disease Duration/Age: Patients with longer disease duration were significantly more likely to have higher SWE values for the triceps brachii (correlation coefficient:0.63, p:0.006). When type 2 and type 3 were examined within themselves, the most affected muscle related to the duration of the disease was again the triceps brachii in both groups (Table 1) (Figure 1).

Relation of MRI Scores to SWE: A positive correlation between SWE values and MRI scores of left and right paraspinal muscles are found (correlation coefficient:0.49, p:0.045) (correlation coefficient:0.67, p:0.003), respectively (Table 2) (Figure 2).

Table 1. Elastography values and disease duration/age correlation analyses

	Deltoid		Triceps		Biceps		Gluteal		Iliopsoas	
	r	p	r	p	r	p	r	p	r	p
SMA type 2-3	0.18	0.47	0.63	**0.006***	-0.13	0.61	0.06	0.81	0.28	0.26
SMA type 2	-0.16	0.65	0.69	**0.02**	0.15	0.67	-0.009	0.97	0.61	0.06
SMA type 3	0.52	0.22	0.91	**0.004**	-0.32	0.48	0.07	0.86	0.25	0.58

SMA, spinal muscular atrophy; r, correlation coefficient; p<0.05

Table 2. Elastography and MRI score correlation analyses

	Elastography (kPa)	MRI score	r	p
Left paraspinal	22.42±9.4 (8-45)	1.67±0.72 (1-3)	0.49	**0.045**
Right paraspinal	20.88±11.6 (8-50)	1.70±0.77 (1-3)	0.67	**0.003**

r, correlation coefficient; p<0.05

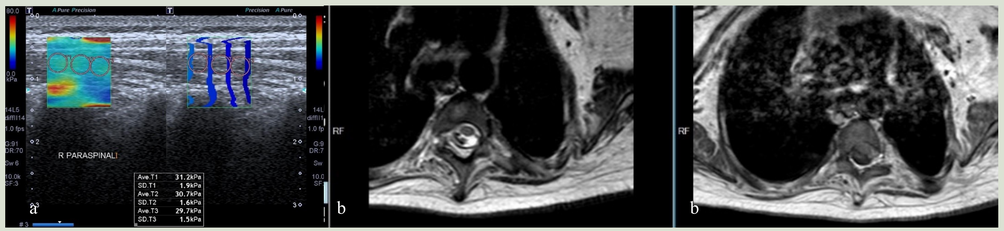

Figure 2. Elastography and MRI images of a patient with SMA type 2

a.SWE image with real-time propagation map reveals the longitudinal orientation of the right paraspinal muscle fibers parallel to the linear transducer. Large distances between the propagation lines and yellow-green color reveal increased muscle stiffness and higher elasticity values as about 30kPa. b. Axial plan T2-W and T1-W images represent significant loss in the muscle volumes with increased signal intensity of thoracal paraspinal muscles.

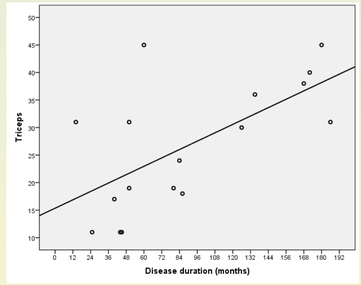

Figure 1. Correlation figure of SWE and Disease Duration/Age in the triceps

CONCLUSION

This is the first study to report the assessment of muscle involvement using SWE in patients with SMA. Our findings confirm the presence of SMA-specific selective muscle involvement pattern shown in previous studies. SWE and MRI findings were similar in showing muscle involvement and SWE can be an alternative diagnostic tool instead of MRI. However, this study has some limitations. First of all, we can include a small sample size for investigating pediatric age group. Second, only the paraspinal muscles could be evaluated, SWE or MRI correlation could not be available for other muscles. And finally, due to the lack of the available normative data for muscle stiffness, the absence of a control group makes it difficult to reach a clear inference.

In conclusion, SWE is a noninvasive practical method that can be used to show muscle involvement in patients with SMA, understand the pathogenesis of segmental involvement, and guide future treatments or monitor the effectiveness of existing new treatment alternatives. For this, larger longitudinal studies are needed and we believe that our study will shed light on these future studies.

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