Prognosis of dysphagia in dermatomyositis

Sirs,

Dysphagia is relatively common complication in dermatomyositis (DM), with 18–58% of patients reported to have this manifestation (1-4). Although risk factors of dysphagia in DM and polymyositis are reported to be age, male gender, anti-TIF1-γ antibody, muscle weakness, and malignancy (5, 6), there is very little published data on the prevalence, treatment outcomes and prognosis of dysphagia in patients with DM.

In this research, which features a cohort of patients with DM, our aims were to (i) reveal the appropriate treatment types and intervention timing for dysphagia recovery, and (ii) identify risk factors for non-recovery from dysphagia.

Serum samples were obtained from adult Japanese patients with DM followed at each medical centre from 2003 to 2016. Detailed medical histories of every patient were retrospectively gathered by unified questionnaire. Eighty-five patients fulfilled the "definite to probable" criterion of Bohan and Peter (7). Autoantibody detection and statistical methods were the same as in our previous study (8). This study was approved by the Ethics Committee of the Nagoya University Graduate School of Medicine and by the individual participating centres.

Of 85 DM patients, 57 (67%) were female. The mean age at DM diagnosis was 61.0±13.9 years. 30 patients were considered to have dysphagia as determined by subjective symptoms judged by their physician's evaluation (10 of 30), examination by otolaryngologists (14 of 30), or examination by speech therapists (5 of 30). The clinical and laboratory characteristics of the 85 DM patients with and without dysphagia are detailed in Supplementary Table S1.

Of the 30 DM patients with dysphagia, we analysed 29 patients' data, excluding one patient with insufficient data (Table I). Sixteen of the 29 patients showed recovery with dysphagia. Survival rates showed strong association with dysphagia recovery (p=0.000003), and high initial dose of prednisolone (PSL) seemed to influence the recovery rate (p=0.045). There was a significant negative correlation between cancer and dysphagia recovery (p=0.025). Other factors, such as age, sex, periods from onset to hospital visit or treatment, intravenous immunoglobulin (IVIG) or other immunosuppressive therapy use did not significantly correlate with dysphagia recovery.

Of the 29 dysphagia-complicated DM patients, 13 patients (33%) died during the follow-up period: 5 from cancer complications (38%), 3 from aspiration pneumonia, and 5 from various other causes. The mean follow-up duration was 15.1±15.5 months. Kaplan-Meier survival curves show the survival probability for patients with or without dysphagia recovery (Supplementary Fig. S1). The mortality rate is significantly higher in patients without recovery from dysphagia than in patients with recovery from dysphagia (p<0.000001).

We had predicted that treatment delay might affect dysphagia recovery, but no such relation was found (Table I). Since the initial dose of PSL was significantly higher in the dysphagia recovery group, early intensive treatment may be effective for recovery. However, in the unrecovered group, 76.9% of patients (10 of 13) had cancer, and this factor may lead clinicians to choose mild immunosuppressive treatments. Medications other than oral PSL, including IVIG, were not significantly related to dysphagia recovery. All but 1 of the 15 surviving patients (93.7%) showed dysphagia recovery. These results suggest that dysphagia in DM might often be reversible without any specific medication.

A major limitation in our study is the lack of standardised dysphagia evaluation methods. Given this limitation, we were unable to compare and discuss the extent of dysphagia recovery between cases. Another limitation was that we did not collect the dates on which the clinician discovered the dysphagia recovery. Future prospective studies on dysphagia recovery and time course are necessary.

Table I. Association between recovery from dysphagia and clinical/laboratory features.

	Improvement of dysphagia		p-value
	(+) n=16 (%)	(-) n=13 (%)	
Age	69.1 ± 7.3	72.5 ± 7.7	0.50
Sex (female)	7 (44)	6 (46)	1
Period (months)			
DM onset to dysphagia*	2.4 ± 2.5	1.6 ± 0.9	0.11
Visit to dysphagia**	0.6 ± 1.1	1.4 ± 2.6	0.77
Treatment to dysphagia***	-0.2 ± 0.9	0.8 ± 2.4	0.39
Visits up to death****	13	5. ± 5.2	0.39
Survival rate	15 (94)	1 (8)	<0.000001
Cancer	5 (31)	10 (76)	0.025
Anti-TIF1-γ	6 (38)	10 (76)	0.06
CK max	3010 ± 2721	2854 ± 3063	0.91
Initial dose of PSL (mg/day)	44.4 ± 19.2	29.2 ± 18.1	0.045
IVIG	3 (18)	0 (0)	0.25
Other medications*****	8 (50)	4 (30)	0.45

*DM onset to dysphagia: period (months) from DM onset to dysphagia onset, **Visit to dysphagia: period (months) from first hospital visit to dysphagia onset, ***Treatment to dysphagia: period (months) from treatment to dysphagia onset, ****Visits up to death: period (months) from first hospital visit up to death. *****Other medications: intravenous steroid pulse therapy, azathioprine, tacrolimus, methotrexate other than oral PSL and IVIG.
CK: creatine kinase; IVIG: Intravenous immunoglobulin; PSL: prednisolone

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M. Ogawa-Momohara, *MD, PhD*
Y. Muro, *MD, PhD*
M. Kono, *MD, PhD*
M. Akiyama, *MD, PhD*

Department of Dermatology, Nagoya University Graduate School of Medicine, Nagoya, Japan.

Please address correspondence to:
Dr Yoshinao Muro,
Department of Dermatology,
Nagoya University Graduate
School of Medicine,
65 Tsurumai-cho, Showa-ku,
Nagoya 466-8550, Japan.
E-mail: ymuro@med.nagoya-u.ac.jp

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