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ORIGINAL ARTICLE

A dysphagia study in patients with sporadic inclusion body myositis (s-IBM)

Ken-ya Murata · Ken Kouda · Fumihiro Tajima · Tomoyoshi Kondo

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Abstract The nature of the swallowing impairment in patients with sporadic inclusion body myositis (s-IBM) has not been well characterized. In this study, we examined ten consecutive s-IBM patients using videofluoroscopy (VF) and computed pharyngoesophageal manometry (CPM). The patients were divided into two groups: patients with complaint and without complaint of dysphagia. VF results indicated pharyngeal muscle propulsion (PP) at the hypopharyngeal and upper esophagus sphincter (UES) in all s-IBM patients. Patients without complaint of dysphagia showed a mild degree of PP, whereas a severe form of PP was observed in patients with complaint of dysphagia. CPM revealed that negative pressure during UES opening was not observed in the s-IBM patients with complaint of dysphagia. Incomplete opening and PP at the UES were observed in all s-IBM patients. These results indicate that the dysphagic processes occur subclinically in s-IBM patients who may not report swallowing impairments.

Keywords Inclusion body myositis · Videofluoroscopy · Pharyngoesophageal manometry · Pharyngeal muscle propulsion · Upper esophagus sphincter

K. Murata (✉) · T. Kondo
Department of Neurology, Wakayama Medical University, 840-1 Kimii-dera, Wakayama 641-8510, Japan
e-mail: kemurata@wakayama-med.ac.jp

K. Kouda · F. Tajima
Department of Rehabilitation Medicine, Wakayama Medical University, 840-1 Kimii-dera, Wakayama 641-8510, Japan

Introduction

Sporadic inclusion body myositis (s-IBM) is an inflammatory myopathy characterized by selectivity of muscle involvement, finger flexor and/or quadriceps femoris involvement, moderate elevation of muscle enzyme concentrations, and a progressive corticosteroid-resistant course. Muscle histopathology shows rimmed-vacuoles, groups of atrophic angular fibers, and endomysial mononuclear cell infiltrations.

Dysphagia has been reported in s-IBM patients. As described by Lotz et al. [1], 10% of the s-IBM patients complained of dysphagia at onset, and 40% of the patients suffered from dysphagia at the time of diagnosis. Patients with progressive dysphagia have a significantly worse functional class rating and poorer quality of life than patients with non-progressive dysphagia [2]. However, the nature of the swallowing impairment in s-IBM and other inflammatory myopathies has not been well characterized. Previous studies suggest that improper contraction of the pharyngeal muscles or cricopharyngeal muscle dysfunction may result in functional obstruction due to dysphagia [3–6].

The purpose of this study was to assess the frequency and nature of dysphagia in s-IBM patients and to identify a possible therapy for dysphagia associated with s-IBM.

Methods

Study subjects were ten consecutive patients (mean age 70.5 ± 7.1 years; 5 males and 5 females) who fulfilled the proposed diagnostic criteria for s-IBM [7] at the Department of Neurology in Wakayama Medical University between January 2000 and July 2011. Muscle biopsy studies were performed on the quadriceps femoris or biceps

brachii in all patients. All specimens were frozen rapidly in isopentane that was chilled in dry ice, and the specimens were stored at −80°C before examination. 5-micron serial sections of each specimen were stained with hematoxylin and eosin (H&E) and modified Gomori Trichrome stain. Muscle biopsy results showed mononuclear cell infiltration around non-necrotic fibers and rimmed-vacuoles in all patients. Swallowing problems were assessed by a personal structured interview, videofluoroscopy (VF) and computed pharyngoesophageal manometry (CPM). All subjects provided written informed consent to the procedures in this study and the ethics committee at the Wakayama Medical University approved all methods used in the study.

Videofluoroscopy (VF)

All ten patients underwent oropharyngeal videofluoroscopic swallowing examination. Patients were placed upright, and the oropharynx was viewed in lateral and anterior-posterior projections. 3 ml of liquid barium and paste barium were administered by teaspoon. Swallowing examinations were repeated in different upright positions. Dysphagia severity was scored using the 8-point Penetration Aspiration Scale (PAS) [8].

Computed pharyngoesophageal manometry (CPM)

CPM was performed in all ten patients. For CPM, a sequential computer manometry system (PC polygraph) (Medtronic, Medtronic Parkway, Minneapolis) with a 4-intraluminal pressure transducer assembly (Mui Scientific, Mississauga, Ontario) was used with the recording sites set at 5 cm apart. The assembly was placed transnasally, and recording sites were chosen at the following four points: oropharynx, hypopharynx, upper esophageal sphincter (UES), and proximal esophagus (Fig. 3a). We evaluated UES pressure and pharyngeal and esophageal peristalsis during barium swallowing.

Results

Clinical findings

Ten patients were examined in this study. The subjects were divided into two groups: patients with complaint of dysphagia (Group A) and patients without complaint of dysphagia (Group B) (Table 1). Group A consisted of three men and two women who complained of dysphagia in the form of regurgitation of liquids and problems with solids. Group B consisted of two men and three women who did not complain of dysphagia. Mean age at examination was 73.8 ± 6.8 years in Group A and 67.2 ± 5.7 years in Group B. The average duration of the disease was 11.0 ± 5.4 years in Group A and 11.0 ± 1.3 years in Group B. There were no statistical differences in mean age, duration of disease and creatine kinase levels between the two groups. Other than one woman (Patient 3) in Group A, all of the patients used a cane or caster walker when walking.

Videofluoroscopy (VF)

Videofluoroscopy results indicated that all patients had a normal oral phase of swallowing and abnormalities in the pharyngeal phase. While barium material did not enter the airway in all patients in Group B, the barium material entered the airway, remained above the vocal folds, and

Table 1 s-IBM patient profiles

Patient	Sex	Complaint of dysphagia	PAS	Age	Duration of disease (years)	Aid for walking	CK (IU/L)	Complications
Group A				73.8 ± 6.8	11.0 ± 5.4		432	
1	M	(+)	2	74	14	Cane	691	HT
2	F	(+)	2	79	12	Walker	482	HT, LS
3	F	(+)	2	64	6	None	353	Sjogren Synd
4	M	(+)	2	83	4	Walker	186	Hepatitis C
5	M	(+)	2	69	19	Cane	447	HT
Group B				67.2 ± 5.7	11.0 ± 1.3		492	
6	M	(−)	1	77	12	Cane	482	HT, DM
7	F	(−)	1	62	9	Cane	503	HT
8	M	(−)	1	62	12	Cane	482	None
9	F	(−)	1	70	12	Cane	643	None
10	F	(−)	1	65	10	Cane	350	None

HT Hypertension, *LS* Lumbar spondylosis, *DM* Diabetes mellitus

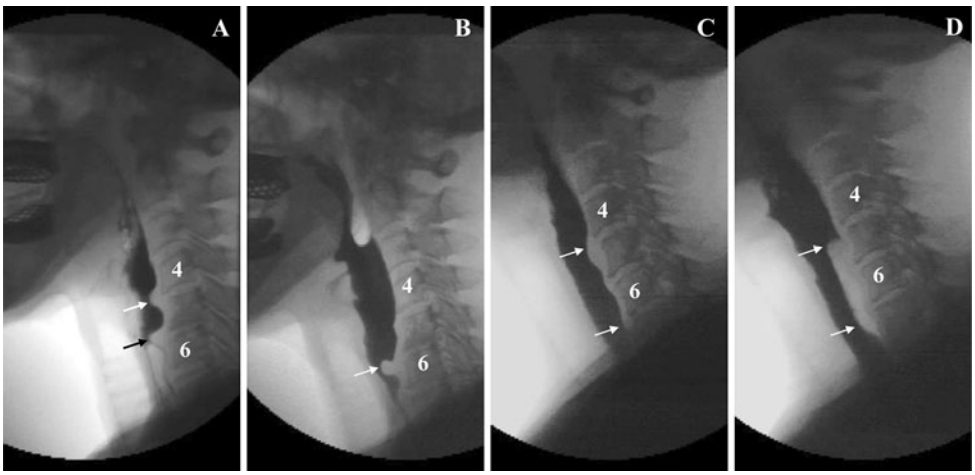

Fig. 1 Videofluoroscopic study in s-IBM patients using paste barium (**a**, **c**) and liquid barium (**b**, **d**). Pharyngeal muscle propulsions (PP) (*arrows*) were observed in Patient 2 (**a**, **b**) and Patient 4 (**c**, **d**). The degree of PP and the narrowing at the upper esophageal sphincter region in Patient 2 is more severe than that in Patient 4. The sites and shapes varied between using paste or liquid barium (**a**, **b** and **c**, **d**)

was ejected from the airway in all patients in Group A. The 8-point PAS showed a score of 2 in all patients in Group A and 1 in all patients in Group B (Table 1). Pharyngeal phase abnormalities included decreased epiglottic deflection and residue in the epiglottic vallecula and piriform recesses. Pharyngeal muscle propulsion (PP) was indicated at the UES in all ten patients without reference to dysphagia (Figs. 1, 2; Table 2). Although, the PP sites in VF ranged from C3 to C7 vertebral levels, PP shapes and sites varied between using liquid and paste barium (Figs. 1a–d, 2a, b). Patients in Group B showed a mild degree of PP, whereas a severe form of PP was observed in all five patients in Group A. An insufficiency of the UES opening was also observed in Group A patients. In addition, prominence of a segment of the hypopharyngeal sphincter muscles was observed in Patient 1 in Group A (Fig. 2a; Table 2).

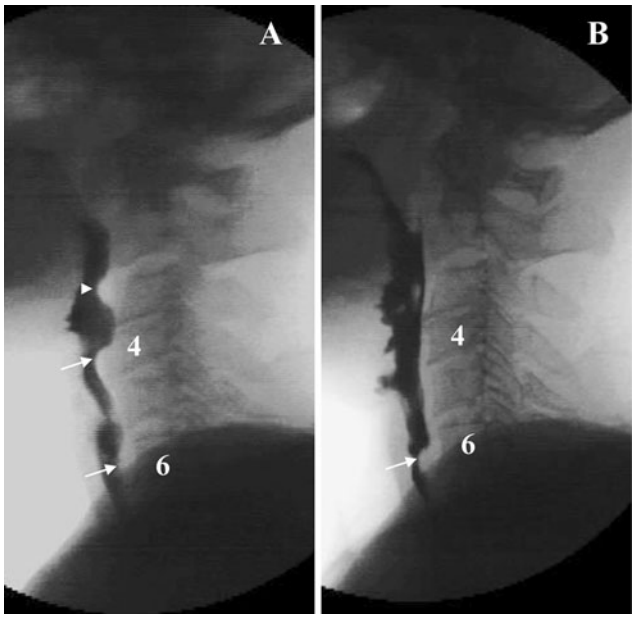

Fig. 2 Videofluoroscopic study in s-IBM patients using paste barium (**a**) and liquid barium (**b**). Pharyngeal muscle propulsions (PP) (*arrows*) and a cephalad prominence (CP) (*arrow head*) were observed in Patient 1 (**a**, **b**). The sites and shapes of PP and CP varied between using paste or liquid barium (**a**, **b**)

Computed pharyngoesophageal manometry (CPM)

In the normal control subjects, the pharyngeal peak pressure at the oropharynx and hypopharynx elevated simultaneously (Fig. 3b-1, 2). The pharyngeal pressure at the oropharynx was higher than that at the hypopharynx. Contrary to the high pharyngeal pressure in the oropharynx and hypopharynx, the pressure at the UES decreased until the UES opened (nadir deglutitive UES pressure) (Fig. 3b-3, arrow). After the barium paste passed through the entrance of the UES, pharyngeal pressure at the UES increased and pushed the paste to the upper esophagus (Fig. 3b-3).

In the s-IBM patients, the pressure at the oropharynx and hypopharynx was very low compared with that of the normal controls (Fig. 3c-1, 2). In addition, the negative pressure during UES opening (nadir deglutitive UES pressure) observed in the normal controls was not observed in the s-IBM patients with dysphagia (Fig. 3c-3). Manometric recordings in all s-IBM patients revealed a lack of oropharyngeal peristaltic activity, a decreased hypopharyngeal peristalsis, and a reduced peak of post-deglutitive UES pressure, while the esophageal resting pressure was normal. In addition, all s-IBM patients in Group A demonstrated no deglutitive UES relaxation in the CPM study.

Table 2 Videofluoroscopic and manometric findings in ten patients with s-IBM

Pt	Group	Videofluoroscopy			
		PP site	Insufficiency of UES opening	Pooling site of barrium	Manometry
1	A	C3, C4-5, C6-7	(+)	EV, piriform	No UES relaxation
2		C4-5, C5-6, C6	(+)	EV, piriform	Decreased oro-hypopharyngeal pressure
3		C5-6, C6-7	(+)	EV	Decreased deglutitive UES pressure
4		C5-6	(+)	EV, piriform	
5		C6-7	(++)	EV, piriform	
6	B	C3-4	(−)	EV	Incomplete UES relaxation
7		C5-7	(−)	None	Decreased oro-hypopharyngeal pressure
8		C5-6	(−)	EV, piriform	Decreased deglutitive UES pressure
9		C4-5, C6-7	(−)	None	Incomplete UES relaxation
10		C5-6	(−)	EV	Incomplete UES relaxation

PP pharyngeal muscle propulsion, *UES* upper esophageal sphincter, E*V* epiglottic vallecula, *piriform* piriform recess

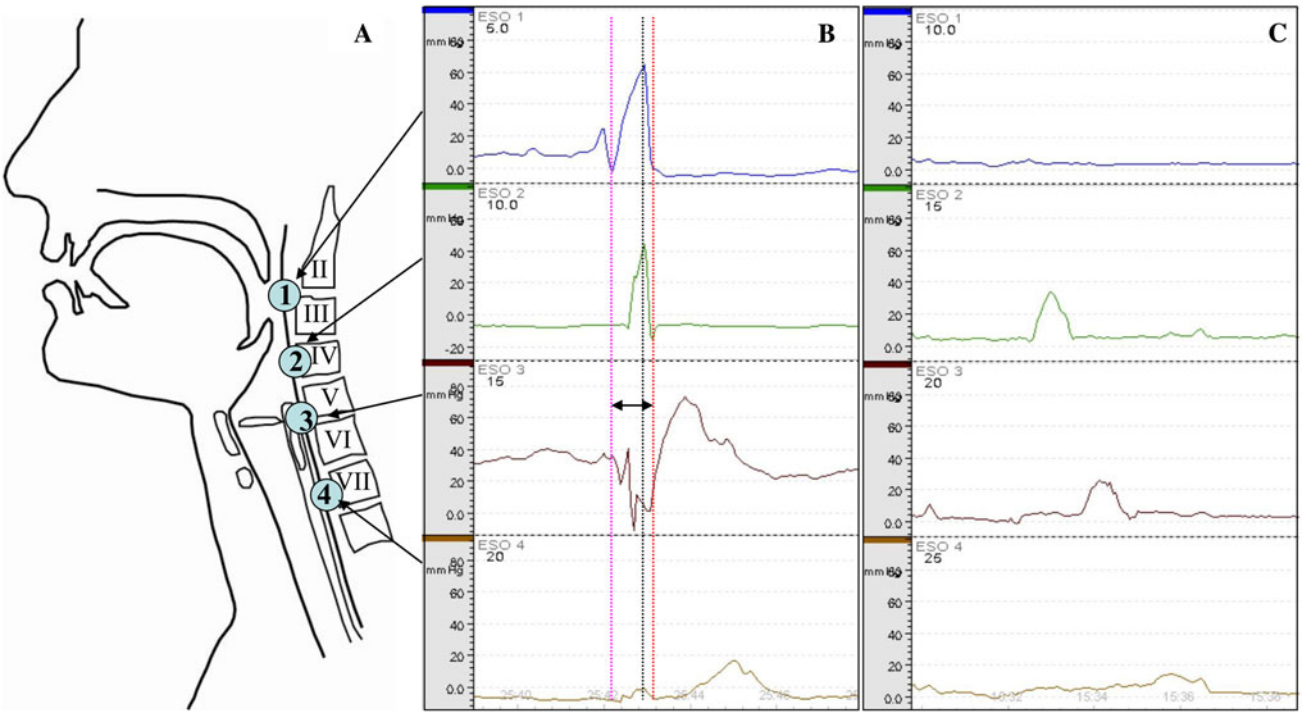

Fig. 3 Manometry **a** Manometry study using simultaneous 4-channel pressure recording during 3 ml barium swallowing. **b** Manometric findings in a healthy control subject: *channel 1* oropharynx, *channel 2* hypopharynx, *channel 3* UES, *channel 4* proximal esophagus. **c** Patient 3: Pharyngoesophageal manometry during swallowing solid barium paste. The pressure at the oropharyngx, hypopharyngx and UES is none or very low compared with that of normal controls. UES negative pressure during UES opening (nadir deglutitive UES pressure), which was observed in normal controls (**b**, *arrow*), was not observed

In contrast, incomplete UES relaxation was observed in Group B patients (Table 2).

Discussion

The consequences of dysphagia include weight loss, the need for modified food consistency and non-oral feeding. Pulmonary infections occur in patients with dysphagia, and aspiration pneumonia is considered a main cause of death in s-IBM patients. We observed PP at the UES and/or hypopharyngeal muscle in all ten s-IBM patients. PP was first reported as a prominent cricopharyngeal impression at the cricopharyngeal muscles [9], and these findings were observed not only in s-IBM patients, but also in mitochondria myopathy patients [10]. PP is also defined as

cricopharyngeal achalasia. We observed PP at the hypopharyngeal muscles in Patient 1, and this PP has been named cephalad prominence [11]. Because PP in VF is observed in front of the vertebral discs, it may be considered the result of disc prolapse. In this study, the PP shapes and sites varied between using liquid and paste barium. Therefore, we conclude that the PP observed here did not represent the result of disc prolapse.

PP was observed at the UES in all ten s-IBM patients, while only five patients (Group A) complained of dysphagia. The local esophageal diameter reduction by PP in Group A was >50% at the UES during swallowing. The degree of PP in Group A was more severe than that of Group B (Fig. 1a–d). In addition, the five patients in Group A showed insufficiency of the UES opening in VF. PP is revealed as barium manages to go through the non-extended pharynx. It represents improper dilation of the pharyngeal muscles during barium passing. A dilation problem of the pharyngeal muscles can occur with the patient being unaware of swallowing difficulties. Although, there was a positive relationship between the severity of PP and the insufficiency of UES opening, PP does not induce cricopharyngeal obstruction. PP represents only the result of dilation problems of pharyngeal musculature.

The lack of negative UES pressure (nadir deglutitive UES pressure) in the s-IBM patients with dysphagia may explain swallowing difficulties. Incomplete pharyngeal musculature opening at the UES during swallowing may have a number of different causes, including impaired relaxation or spasm of the UES, hyperplasia and hypertrophy or fibrosis of the cricopharyngeal muscles, weakness of the suprahyoid muscles, and failure of neural inhibition of tonic sphincter contraction [12]. An examination of cricopharyngeal muscle biopsies of one patient was reported at the time of cricopharyngeal myotomy [4, 13]. Numerous small, round atrophic muscle fibers were observed which varied in size. Because the cricopharyngeal muscles have a sphincteric function, atrophic cricopharyngeal muscles failed to push foods toward the upper esophagus. In our manometry study, post-deglutitive UES pressure was observed, but the peak pressure was greatly reduced. Low pressure at the oropharynx and hypopharynx, and hypo-oropharyngeal peristaltic activities induced problems with the propulsion of the bolus through the sphincter muscles. In addition, there was a marked increase in endomysial connective tissue, some replacement by fat, and proliferative connective tissue. In s-IBM patients in Group A, nadir deglutitive UES pressure was not observed. This finding suggests that endomysial proliferative connective tissue prevented the extension and relaxation of the UES.

The prevalence of s-IBM in Asian populations including Japan has not been examined. Recently, a national survey study revealed that the number of s-IBM patients in Japan is estimated to be around 1,250 and that the prevalence of s-IBM is 9.83 per million [14]. We examined ten patients, representing 0.8% of all s-IBM patients in Japan. Although the number of patients was very low and results of our study are limited in significance, our study revealed a tendency in s-IBM patients with dysphagia.

Recently, cricopharyngeal myotomy was selected to reduce dysphagia in s-IBM patients [15]. The aim of the myotomy is to remove the impaired UES relaxation, which is not overcome by decreased PP. Indeed, myotomy has shown to be useful in improving dysphagia associated with UES hyperactivities in s-IBM, however, other therapies, such as intravenous immunoglobulin [16] or botulinum toxin A [17], and balloon dilation have been employed for s-IBM patients with dysphagia.

The combination test using VF and manometry is needed to assess the preservation of sphincter muscle strength and the efficacy of each therapy. Deglutitive pharyngoesophageal functions should be examined routinely in all s-IBM patients even if they do not complain of dysphagia.

References

1. Lotz BP, Engel AG, Nishino H, Stevens JC, Litchy WJ (1989) Inclusion body myositis. Observations in 40 patients. Brain 112(Pt 3):727–747
2. Houser SM, Calabrese LH, Strome M (1998) Dysphagia in patients with inclusion body myositis. Laryngoscope 108:1001–1005
3. Wintzen AR, Bots GT, de Bakker HM, Hulshof JH, Padberg GW (1988) Dysphagia in inclusion body myositis. J Neurol Neurosurg Psychiatry 51:1542–1545
4. Danon MJ, Friedman M (1989) Inclusion body myositis associated with progressive dysphagia: treatment with cricopharyngeal myotomy. Can J Neurol Sci 16:436–438
5. Dietz F, Logeman JA, Sahgal V, Schmid FR (1980) Cricopharyngeal muscle dysfunction in the differential diagnosis of dysphagia in polymyositis. Arthritis Rheum 23:491–495
6. Kagen LJ, Hochman RB, Strong EW (1985) Cricopharyngeal obstruction in inflammatory myopathy (polymyositis/dermatomyositis). Report of three cases and review of the literature. Arthritis Rheum 28:630–636
7. Griggs RC, Askanas V, DiMauro S, Engel A, Karpati G, Mendell JR, Rowland LP (1995) Inclusion body myositis and myopathies. Ann Neurol 38:705–713
8. Rosenbek JC, Robbins JA, Roecker EB, Coyle JL, Wood JL (1996) A penetration-aspiration scale. Dysphagia 11:93–98
9. Seaman WB (1966) Cineroentgenographic observations of the cricopharyngeus. Am J Roentgenol Radium Ther Nucl Med 96:922–931
10. Kornblum C, Broicher R, Walther E, Seibel P, Reichmann H, Klockgether T, Herberhold C, Schroder R (2001) Cricopharyngeal achalasia is a common cause of dysphagia in patients with mtDNA deletions. Neurology 56:1409–1412
11. Shapiro J, Martin S, DeGirolami U, Goyal R (1996) Inflammatory myopathy causing pharyngeal dysphagia: a new entity. Ann Otol Rhinol Laryngol 105:331–335

12. Cook IJ, Blumbergs P, Cash K, Jamieson GG, Shearman DJ (1992) Structural abnormalities of the cricopharyngeus muscle in patients with pharyngeal (Zenker's) diverticulum. J Gastroenterol Hepatol 7:556–562
13. Verma A, Bradley WG, Adesina AM, Sofferman R, Pendlebury WW (1991) Inclusion body myositis with cricopharyngeus muscle involvement and severe dysphagia. Muscle Nerve 14:470–473
14. Suzuki N, Aoki M, Mori-Yoshimura M, Hayashi YK, Nonaka I, Nishino I (2011) Increase in number of sporadic inclusion body myositis (sIBM) in Japan. J Neurol. doi:10.1007/s00415-011-6185-8
15. St Guily JL, Moine A, Perie S, Bokowy C, Angelard B, Chaussade S (1995) Role of pharyngeal propulsion as an indicator for upper esophageal sphincter myotomy. Laryngoscope 105:723–727
16. Cherin P, Pelletier S, Teixeira A, Laforet P, Simon A, Herson S, Eymard B (2002) Intravenous immunoglobulin for dysphagia of inclusion body myositis. Neurology 58:326
17. Liu LW, Tarnopolsky M, Armstrong D (2004) Injection of botulinum toxin A to the upper esophageal sphincter for oropharyngeal dysphagia in two patients with inclusion body myositis. Can J Gastroenterol 18:397–399