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Chapter 3

Detecting dysphagia in inclusion body myositis

F.M.E. Cox¹

J.J.G.M. Verschuuren¹

B.M. Verbist²

E.H. Niks¹

A.R. Wintzen¹

U.A. Badrising¹

for the Dutch IBM study group*

Departments of Neurology¹ and Radiology², Leiden University Medical Center, Leiden, the Netherlands

* Authors and investigators of the Dutch IBM Study Group include, from the Departments of Neurology:

M.L.C. Maat-Schieman (Leiden University Medical Center); P.A. van Doorn (Erasmus Medical Center Rotterdam); B.G.M. van Engelen (Radboud University Nijmegen Medical Center); C.G. Faber (Maastricht University Medical Center); J.E. Hoogendijk (University Medical Center Utrecht) and M. de Visser (Academic Medical Center Amsterdam).

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Abstract

Dysphagia is an important yet inconsistently recognized symptom of sporadic inclusion body myositis (IBM). It can be disabling and potentially life-threatening. We studied the prevalence and symptom-sign correlation of dysphagia. Fifty-seven sporadic IBM patients were interviewed using a standard questionnaire for dysphagia and 43 of these underwent swallowing video fluoroscopy (VFS). Symptoms of dysphagia were present in 37 of 57 patients (65%). Nevertheless, only 17 of these patients (46%) had previously and spontaneously complained about swallowing to their physician. Both symptoms of impaired propulsion (59%) and aspiration-related symptoms (52%) were frequently mentioned. Swallowing abnormalities on VFS were present in 34 of 43 patients (79%) with impaired propulsion of the bolus in 77% of this group. The reported feeling of impaired propulsion was confirmed by VFS in 92% of these patients.

Dysphagia in sporadic IBM is common but underreported by the vast majority of patients if not specifically asked for. In practice, two questions reliably predict the presence of impaired propulsion on VFS: 'Does food get stuck in your throat' and 'Do you have to swallow repeatedly in order to get rid of food'. These questions are an appropriate means in selecting patients with sporadic IBM for further investigation through VFS and eventual treatment.

Introduction

Dysphagia is one of the main clinical features of sporadic inclusion body myositis (IBM).¹⁻⁵ Together with weakness of quadriceps and finger flexor muscles, dysphagia constitutes a clue for the diagnosis. It may even be the presenting symptom.^{4, 6-8} Dysphagia causes both social embarrassment and life threatening complications. It may lead to avoidance of shared meals and social isolation due to audible deglutition. Besides, it can lead to unintended weight loss. Due to a higher incidence of aspiration pneumonia, life expectancy is assumed to be shorter.⁹ Therapeutical interventions described to be effective for dysphagia in sporadic IBM patients include cricopharyngeal myotomy, pharyngoesophageal dilatation and swallowing exercises such as the Mendelsohn maneuver.⁹

We investigated the prevalence and symptom-sign correlation of swallowing dysfunction in a large group of sporadic IBM patients. Through this approach we aimed at finding specific questions which could reliably predict dysphagia in the sporadic IBM patient in order to influence the selection of patients who need further investigation through video fluoroscopy (VFS) or who need therapeutical interventions.

Patients and Methods

Patients

The patients in this study were selected from a national cohort of 86 sporadic IBM patients. The method of recruitment applied here has been described previously.¹⁰ Of these 86 patients, five patients could not be located, six died prior to assessment and 13 refrained from participation. Five patients were excluded because of prior cricopharyngeal myotomy. This left 57 patients to study fulfilling the ENMC criteria for definite (n=51) or probable (n=6) IBM.¹⁰ To evaluate recruitment bias we compared age (at onset), sex and disease duration of these 57 patients with those of the whole cohort of 86 patients, based on their medical records. All patients gave informed consent. The study was approved by the Ethics committee of the Leiden University Medical Center. All patients answered a questionnaire and were examined by video fluoroscopy (VFS), unless prevented by logistic difficulties.

Questionnaire

Patients were personally interviewed by one single investigator using a previously published standard questionnaire regarding dysphagia (Table 1).¹¹

Table 1. Results of questionnaire

<i>Question</i>	<i>No.</i>	<i>%</i>	<i>Median</i>
Impaired propulsion-related questions			
Does food get stuck in your throat?			
Yes	31	54	
No	26	46	
If yes, how often per month?*			
1-5	4	13	
6-10	0	0	30
11-30	2	6	
>30	25	81	
Do you have to swallow repeatedly in order to get rid of the food?			
Yes	31	54	
No	26	46	
If yes, which consistency of food?			
Fluid	0	0	
Semi-fluid	3	10	
Solid	22	71	
Both fluid and solid	6	19	
If yes, how often during one bite/sip?			
1-5	28	90	
6-10	3	10	3
>10	0	0	
How often per month?*			
1-5	3	10	
6-10	0	0	30
11-30	1	3	
>30	27	87	
Do you use normal or grinded food?			
Normal	56	98	
Grinded	1	2	
Aspiration-related questions			
Do you choke when using solid or fluid food?			
Yes	33	58	
No	24	42	
If yes, how often per month?*			

1-5	11	33	
6-10	4	12	30
11-30	8	24	
>30	10	30	
Do you have to cough often during/after a swallow when eating?			
Yes	19	33	
No	38	67	
If yes, how often per month?*			
1-5	1	5	
6-10	5	26	20
11-30	4	21	
>30	9	47	
Do you have to cough during/after a swallow when drinking?			
Yes	8	14	
No	49	86	
If yes, how often per month?*			
1-5	3	37	
6-10	1	12	11
11-30	2	25	
>30	2	25	
Do you cough while lying down?			
Yes	12	21	
No	45	79	
If yes, how often per month?*			
1-5	3	25	
6-10	1	8	30
11-30	1	8	
>30	7	58	
General questions			
Have you ever reported trouble with swallowing to a doctor spontaneously?			
Yes	17	30	
No	40	70	
Do you sometimes suffer from heartburn?			
Yes	13	23	
No	44	77	

Do you cough daily? (not related to swallowing/lying down)			
Yes	18	32	
No	39	68	
Do you give up phlegm daily?			
Yes	28	49	
No	29	51	
Does fluid come out of your nose during drinking?			
Yes	5	9	
No	52	91	
If yes, how often per month?			
1-3	1	20	4
4-6	4	80	

* Multiple occurrences on one day have been counted as one occasion

Dysphagia due to impaired propulsion (IP) was defined by the presence of at least one of the following symptoms: the feeling of food getting stuck in the throat or repetitive swallowing for one bolus. Aspiration-related (AR) dysphagia was defined by the presence of at least one of the following symptoms: choking for more than five times a month or coughing related to eating, drinking or lying down.

Video fluoroscopy

Video fluoroscopy (VFS) was used to record the movements and anatomy of the pharynx and the cervical oesophagus in lateral view. Patients were seated upright with their heads in neutral position. The head was stabilized occipitally and frontally. Each patient successively swallowed four volumes of opaque fluid (Jopamiro), viz. 3, 6 and 9 ml, followed by a 'dry swallow' (0 ml). The VFS was scored by one single observer who was aware of the diagnosis, but uninformed with regard to the patient's clinical features. Signs were subdivided in two categories: impaired propulsion (IP) or aspiration-related (AR).

IP included repetitive swallowing, residue in the valleculae or piriform sinus and cricopharyngeal sphincter dysfunction. Sphincter dysfunction was defined by a posterior indentation of the sphincter, as a result of contraction of the cricopharyngeal muscle at the moment the pharynx was still dilated and filled with contrast.¹²

Aspiration was defined by fluid entering the larynx. Because inadequate epiglottal downward tilting (IEDT) and residues in the valleculae and/or piriform sinus are generally regarded as risk factors for aspiration,^{13, 14} these three signs were also considered

as AR signs. A normal swallow was concluded in the absence of IP or AR signs. Furthermore, the presence or absence of a Zenker's diverticulum was recorded.

Results

Demographical and clinical characteristics

The mean age of the 57 patients was 67 ± 8 years for men ($n = 41$) and 71 ± 10 years for women ($n = 16$), with a mean age at onset of muscle weakness at 57 ± 9 years for men and 59 ± 10 years for women. Most patients ($n = 39$, 68%) presented with weakness of the quadriceps muscles. Dysphagia was the presenting symptom in four patients (7%), at a mean age of 64 years. Mean duration of symptoms of muscle weakness was 10 ± 7 years for men and 12 ± 5 years for women. The investigated group did not differ significantly from the original population group of 86 patients with regard to age (at onset) and disease duration. The male sex however, was slightly over-represented in the investigated group compared with the group of 86 patients (male to female ratio: 2:1).

Questionnaire ($n = 57$)

Thirty-seven patients (65%) had symptoms of dysphagia. Women reported dysphagia more often compared to men (88 vs. 56%). Of these patients, 26 reported both IP and AR symptoms (46%), seven reported symptoms of IP only (12%) and four had exclusively AR symptoms (7%). The vast majority of these patients had symptoms on a daily basis. Solid food was the most likely to get stuck in the throat ($n = 22$, 71%), yet only one patient (2%) used grinded food (Table 1).

Twenty patients with dysphagia on the questionnaire (54%) had not spontaneously disclosed swallowing complaints to a physician before.

Video fluoroscopy ($n = 43$)

In 34 patients (79%) VFS was abnormal. Abnormal findings were equally frequent in men and women.

Thirty-three patients (77%) had signs of IP (repetitive swallowing ($n = 24$, 56%), residues in the piriform sinus ($n = 19$, 44%) and valleculae ($n = 16$, 37%), cricopharyngeal sphincter dysfunction ($n = 16$, 37%), Figure 1).

Only one patient showed aspiration during VFS; this patient also had repetitive swallowing, vallecular and piriform sinus residues and IEDT. Twenty-three (53%) other

patients had aspiration-related signs of whom 18 (43%) had IEDT. Normal swallowing function was observed in nine patients.

A Zenker's diverticulum was found in eight patients (19%). The ostium of the diverticulum was invariably located just above the upper oesophageal sphincter.



Figure 1. VFS showing indentation of the cricopharyngeal sphincter

Comparison between questionnaire and video fluoroscopy

In 25 patients who reported symptoms of IP, a VFS was performed. Of these, 23 had corresponding IP signs on VFS (positive predictive value: 0.92). The highest positive predictive values were calculated for the questions regarding food getting stuck in the throat (91%) and whether or not repeated swallows (92%) were needed. Nineteen patients (83%) showed repetitive swallowing, 15 (65%) had a piriform sinus residue, 13 (57%) had a vallecular residue and 10 (44%) demonstrated sphincter dysfunction. Remarkably, two (8%) patients had symptoms of IP, but a normal VFS. Ten out of 33 patients with IP signs on VFS had no symptoms of IP on questionnaire. The sensitivity of the questionnaire concerning IP was 0.70, the specificity 0.80 and the negative predictive value 0.44.

Twenty-three patients who underwent VFS reported AR symptoms on the questionnaire. Confirmation of aspiration on VFS was obtained in only one patient. Fourteen (65%) other patients showed one or more different AR signs on VFS (positive predictive value: 0.65). Nine out of 24 patients with AR signs on VFS had no symptoms of aspiration on questionnaire. The sensitivity concerning AR signs was 0.63, the specificity 0.58 and the negative predictive value 0.55.

Abnormalities were more frequently detected by VFS than based on the questionnaire scores, 79% vs. 65%.

Discussion

Dysphagia is a frequent, embarrassing and potentially dangerous symptom in patients with sporadic IBM. Using a questionnaire, we aimed at making an elaborate view on the swallowing status of the patient. For analysis, we took those questions into account which we considered to be indicative of IP or aspiration and categorized them. The 2 categories were made to enable correlation between symptoms and signs.

We encountered dysphagia in 65% of our sporadic IBM patients after excluding patients who had undergone a cricopharyngeal myotomy. The prevalence of dysphagia in sporadic IBM differs considerably between reported studies, ranging from 40-80%.¹⁻⁵ The wide range in prevalence figures most likely originates from the absence of a universal definition for dysphagia.

In the present study, 37% of our patients had abnormal function of the cricopharyngeal sphincter. Other investigators described sphincter dysfunction in 47% to 69% of myositis-patients.^{8, 15} Inflammatory involvement of the cricopharyngeal muscle is most likely the basis for dysfunction, as has been suggested by authors who reported the presence of inflammation or rimmed vacuoles in cricopharyngeal muscle biopsies.¹⁵⁻¹⁸ Due to inflammation, the compliance of the sphincter might be reduced, impeding the opening and thus the trans-sphincteric bolus flow.¹⁹ Ageing may be another cause of a diminished sphincter opening, but ageing does not seem to prolong the pharyngeal bolus transit time, neither does it disturb trans-sphincteric bolus flow coordination.²⁰

Observed aspiration was rare in our patients ($n = 1$). However, aspiration-related signs were present in half the group. Two publications revealed higher prevalences of aspiration on VFS (24 and 61%) in inflammatory myopathies. These studies also included polymyositis and dermatomyositis patients.^{8, 15} These studies, including the present study, used different fluid volumes for swallows. A different fluid consistency and volume, and a different position of the head during VFS may have influenced these

prevalence figures. Differences in mechanisms of dysphagia within the inflammatory myopathies cannot be ruled out, either.

The sensitivity and specificity of questions regarding impaired propulsion were disappointing. However, for the detection of dysphagia through questions from the questionnaire the positive predictive value is the most important. The positive predictive value appeared to be high enough to be of use when selecting patients to be investigated by VFS or for possible further treatment.

Despite the frequent occurrence of dysphagia in sporadic IBM only 54% of patients with dysphagia on the questionnaire had expressed swallowing difficulties to their physician spontaneously. This indicates that a proactive approach to detect dysphagia using a simple set of questions could facilitate early treatment and lead to prevention of complications.

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