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Laryngology

Upper dysphagia in patients affected by systemic sclerosis: prevalence and features

La disfagia orale e faringea in pazienti affetti da sclerosi sistemica: prevalenza e caratteristiche

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SUMMARY

Herein, we describe the prevalence and features of dysphagia in patients affected by systemic sclerosis (SS). We analysed the data of 19 patients obtained by administering the M.D. Anderson Dysphagia Inventory (MDADI) scale that measures dysphagia symptoms and by physical assessment consisting of judging specific lip, mandible and tongue performances (scale 0-3) and diadochokinesis, respiratory and phonatory functions (scale "poor", "fair", "good", "normal") according to Robertson's method. Subjects also underwent flexible endoscopic examination of swallowing. MDADI showed that 74% of answers were included in "*mild*" class of disability, 21% as "*moderate*" and 5% as "*severe*". The performance of lips, mandible and tongue that most frequently scored 1 were the opening (52.6% for the lips and 47.4% for the mandible) and the pop of the tongue (52.7%). The percentage of compromised respiratory, phonatory and diadochokinesis tests ("poor" or "fair") was 81%, 70.1% and 74%, respectively. Flexible endoscopic examination of swallowing revealed pharyngolaryngeal sensory deficit and signs of oropharyngeal dysphagia in more than half of cases (58% and 53%, respectively). This study highlights the presence of dysphagia in SS patients and demonstrates the importance of a multidimensional approach that includes subjective and objective evaluation to characterise specific features of swallowing alterations that have a high-impact on upper dysphagia.

KEY WORDS: dysphagia, systemic sclerosis, upper dysphagia, flexible endoscopic examination of swallowing

RIASSUNTO

Scopo dello studio è stato quello di valutare, per mezzo di un campionamento trasversale, la prevalenza e le caratteristiche dei disturbi di deglutizione in pazienti affetti da sclerosi sistemica (SS). Abbiamo analizzato i dati ottenuti da 19 pazienti sottoposti a tests soggettivi, clinici e strumentali. Per la valutazione soggettiva è stato utilizzato il questionario di autovalutazione "M.D. Anderson Dysphagia Inventory". Per l'esame clinico ai pazienti veniva chiesto di eseguire specifici movimenti e prassie delle labbra, della lingua e della mandibola (score da 0 a 3), performances vocali, respiratorie e di diadococinesi in accordo con il sistema Robertson's ("insufficiente", "quasi sufficiente", "sufficiente" e "normale"). Infine ciascun paziente veniva sottoposto ad esame fibroendosopico della deglutizione con test della sensibilità. Risultati dell'MDADI: il 74% dei pazienti mostrava un'alterazione "lieve" della deglutizione, il 21% ed il 5% rispettivamente un grado "moderato" e "severo" di disfagia. Le performances più compromesse erano l'apertura della mandibola e delle labbra (52,6% and 47,4%) e lo schiocco della lingua (52.7%). La voce, la respirazione e la diadococinesi erano alterate in più del 70% dei casi. La FEES ha dimostrato un'alterazione della fase faringea e la presenza di deficit della sensibilità faringolaringea in più della metà dei pazienti (58% e 53%). Lo studio mette in evidenza l'elevata prevalenza della disfagia alta nei pazienti affetti da sclerosi sistemica e dimostra l'importanza di una valutazione multidimensionale che coinvolga entrambi il logopedista ed il foniatra in grado di eseguire un esame clinico specifico e strumentale mirato, indispensabili per contribuire a riconoscere la sede della disfagia e con essa caratteristiche non altrimenti rilevabili.

PAROLE CHIAVE: *disfagia, sclerosi sistemica, disfagia orale e faringea, valutazione fibroendoscopica della deglutizione*

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Introduction

Systemic sclerosis (SS) is a rare autoimmune disorder characterised by alterations in humoral and cellular immunity, leading to fibroproliferative alterations in microvasculature which in turn causes abnormal collagen deposition in the skin and internal organs [1,2]. The gastrointestinal tract is one of the most commonly affected organ systems, involved in approximately 90% of SS patients. The specific changes contribute to autonomic dysfunctions and dysmotility [4] that cause a variety of morbid symptoms including dysphagia [2-4]. The pathogenesis of dysmotility is related to progression of myopathy, neuropathy and fibrosis leading to abnormalities in compliance and contractility of the GI tract wall [4]. In the literature, dysphagia is reported as a rare presenting complaint of scleroderma in which symptoms occur as the oesophagus becomes more severely affected. When the oesophagus is compromised, the disease process is usually diffuse with involvement of multiple levels of the gastrointestinal tract [5]. Nevertheless, as demonstrated in a previous study, dysphagia (oropharyngeal) is actually not so rare in immunomediated diseases, particularly in cases affected by SS [6]. The recent literature offers no specific studies that are capable of ruling out the presence of primary alterations of swallowing in SS. In addition, almost all published studies describe dysphagia in SS as lower or non-specific dysphagia. Up to now, except for a case reported in 1981 [7], only Rajapakse et al. [8] presented a case series affected by dysfunction of the pharyngoesophageal region. Therefore, based on the available literature, oropharyngeal dysphagia (OD) is infrequent, poorly recognised and poorly documented.

It is known that aspiration, pneumonia, malnutrition, increased mortality, prolonged hospitalisation, advanced disability and declining quality of life may be the consequences of OD. Since early dysfunction is still very responsive to appropriate management, it is clear that early diagnosis and treatment are fundamental issues in preventing such life-threatening complications.

In the light of all the aforementioned considerations, the primary objective of this study was to investigate the prevalence of swallowing dysfunction in SS patients using self-assessment questionnaires in addition to physical evaluation that included clinical and instrumental approaches. The secondary aims were to describe the features of dysphagia focusing on the site and characteristics of symptoms and to provide a detailed description of the structural and functional abnormalities.

Materials and methods

From January to June 2018, at the clinics and rheumatology department of St. Carlo's Hospital at Potenza, we recruited patients affected by SS disease. The inclusion criterion was clinically and laboratory-defined SS disease. The exclusion criteria were thyroid, laryngeal, oesophageal (all except GERD), gastric, respiratory diseases or previous surgery, inability to cooperate, and past or present swallowing rehabilitation therapy. All patients routinely underwent ENT evaluation including flexible fiberoptic rhinolaryngoscopy to evaluate the anatomical integrity of pharynx and larynx. Comorbidities were recorded and patients were asked to answer specific dysphagia symptoms listed in the dysphagic adults assessment questionnaire developed by Travalca Cupillo-Castellini [12] (Tab. I). All persons gave their informed consent prior to inclusion in the study.

In order to assess the impact of dysphagia on the quality of life, we selected the "MDADI" M. D. Anderson Dysphagia Inventory translated into Italian [9,10]. It is a long-standing validated screening self-reported measure of a patient's perceived handicap or impairment from their swallowing. We opted for MDADI because of its simplicity, limited number of questions and direct scoring to assess the handicapping effects of OPD. In order to calculate the prevalence of symptoms (items), we divided the answers into two classes: "No *symptomatic"* that included the "never" and "almost never" answers and "*Symptomatic*" that included the "almost always" and "always". Moreover, we calculated the total score between 0 and 60 and, to obtain a grading scale of dysphagia, we divided the distribution of the scores into four classes of disability: 0-2 (*absent*), 3-14 (*mild*), 15-29 (*moderate*) and 30-60 (*severe*). One additional item is present, whose score is not computed in the total of the MDADI score but which accounts for the general (G) distress reaction to symptoms ("*Does your swallowing problem interfere with your quality of life?").* The interviews were carried out by two trained speech pathologists (DC, MM). With respect to bedside swallowing evaluation (BSE) [11], clinical signs closely related to dysphagia and aspiration were considered: presence of "wet voice", post-swallow residue in the mouth and post-swallow cough. Moreover, using a scale from 0 (not able) to 3 (good) the performance of the lips, mandible and tongue was tested according to the protocol of Travalca Cupillo-Castellini [12], and, diadochokinesis, respiratory and phonatory functions according to Robertson's method [13] (scoring "poor", "fair", "good" and "normal"). Finally, we performed flexible endoscopic examination of swallowing with a sensory test according to Rees [14] and Langmore [15]. The sensory test was performed by lightly touching the aryepiglottic fold or the tip of the epiglottis with the tip of endoscope and ask the patient if he/she feels it. We considered normal subjects who answered affirmatively or who coughed.

Table I. Prevalence of swallowing symptoms and comorbidities in decreasing order.

Symptoms	No. cases	%	Comorbidities	No. cases	%
Food or liquid come back up into the throat	13/19	68	Sicca syndrome	9/19	47
Frequent throat clearing	13/19	68	Osteoarthrosis	8/19	42
The amount of saliva is decreased	12/19	63	Arterial hypertension	8/19	42
Food or liquid come back up into the mouth	12/19	63	Gastroesophageal reflux disease	7/19	37
Globus pharyngeal	11/19	58	Sjogren syndrome	4/19	21
Feeling of food remaining in the upper throat	11/19	58	Hiatal hernia	4/19	21
You clear your throat when you swallow food	10/19	53	Hypovitaminosis D	3/19	16
Difficulty to start swallowing	9/19	47	Thyroid nodules	3/19	16
Cough when you swallow food	8/19	42	Hashimoto's thyroiditis	3/19	16
Feeling of food remaining in the mouth	8/19	42	Hypothyroidism	3/19	16
Loss of saliva during the night	6/19	32	Pulmonary arterial hypertension	3/19	16
Decrease in body weight	6/19	32	Osteoporosis	3/19	16
History of pneumonia by bacterial infection	6/19	32	Dyslipidaemia	2/19	11
Feeling of food in the lower throat	5/19	26	Venous chronic insufficiency	2/19	11
The amount of saliva is increased	4/19	21	Esophagitis	1/19	5
Increase in body weight	4/19	21	Hypercholesterolaemia	1/19	5
Leakage of food or fluid from the mouth	3/19	16	Diabetes type 2	1/19	5
Food or liquid come back up into the nose	3/19	16			

Written informed consent was obtained from all participants included in the study. Statistical analysis was performed using commercially available software (Excel – Microsoft Corporation, Redmond, Washington, USA). Continuously distributed outcomes were summarised as means and categorical outcomes with frequencies and percentages. The numerical data and categoric variables were compared by applying a Student's t test and chi-square test, respectively. The level of significance was set at $p < 0.05$.

Results

From a series of 28 patients, 9/28 met exclusion criteria and 19/28 were considered. Seventeen cases were female and two were males with a mean age of 58.9 years (min. 30 max 78; SD = 13.5). Three of 19 (16%) casess were affected by diffuse cutaneous SS and 16/19 (84%) by limited cutaneous SS.

Comorbidities and dysphagia-specific symptoms

The principal comorbidities and respective prevalence are listed in Table I. Sicca syndrome was the most prevalent occurring in 9/19 (47%) of cases, followed by osteoarthritis 8/19 (42%), arterial hypertension (8/19; 42%), gastro-oesophageal reflux (7/19; 37%) and fibromyalgia (5/19; 26%). The prevalence of specific dysphagia symptoms is shown in Table I. The symptoms referred by more than half of cases were "*Frequent throat clearing*" (13/19; 68%), "*Food or liquid come back up into the throat*" (13/19; 68%), "*Food or liquid come back up into the mouth*" (12/19; 63%), "*The amount of saliva is decreased*" (12/19; 63%), "*Feeling of food remaining in the upper throat*" (11/19; 58%), "*You clear your throat when you swallow food*" (10/19; 53%).

M.D. Anderson Dysphagia Inventory (MDADI)

The total mean score was 11.42 ("mild" dysphagia). In particular, 74% of answers were included in "*mild*" class of disability, 21% as "*moderate*" and 5% as "*severe*". The partial scores for each group of questions were 7.68, 2.42 and 1.31 for the Physical, Emotional and Functional sections, respectively. The score of the Physical (P) section was the highest and significantly greater compared with the other sections ($p < 0.05$). Finally, the mean score of Emotional (E) sub-items was significantly higher than the Functional (F) one ($p < 0.05$). The mean percentage of "Symptomatic" answers was 17.58%, 10.53% and 6.32% for P, E and F group of sub-items, respectively. Nevertheless, these frequencies were significantly less ($p < 0.05$) compared with those of answers with a score between 0-1 (82.42%, 89.47%, 93.68% and for the physical, emotional and functional group of sub-items respectively). Table II showed the frequency of all items in decreasing order. Regarding the item for general (G) distress reaction to dysphagia, 26.3% of answers were included as "Symptomatic".

Bedside swallowing evaluation

The tongue appeared atrophic in 17/19 (89%) of patients, while they were normal in the remaining two cases. On-

ly 2/19 (10%) patients had normal teeth, while the other 17 patients (89%), partially or totally edentulous, used dental prostheses. Three of 19 (16%) patients showed a "wet voice", while post-swallow residue in the mouth was observed in 12/19 (63%) of cases and in only 1/19 (5.2%) case was post-swallow cough present. As shown in Table III the performance of the lips most frequently scoring as 1 was the opening (52.6%). The remaining dynamic tests of the lips were performed almost normally (score 2) in the more than half of cases (54.7%). The opening of mandible and the pop of the tongue were most frequently compromised since a score of 1 was present in 47.4% of patients and in 52.7%, respectively. The percentage of compromised respiratory tests ("poor" or "fair") was 81% and significantly higher compared with "good" or "normal" performances (18.9%) ($p < 0.05$). The test most frequently judged "poor" was "*take a deep breath, then make the /s/ sound again, but start at a whisper then get louder*". The frequencies of the remaining tests are showed in Table IV; 70.1% of phonatory performances were abnormal ("poor" and "fair") and 29.8% were "good" or "normal". The difference was statistically significant ($p < 0.05$). The first two recurrent tests with "poor" score were, respectively, "*Take a deep breath and during expiration say /a/ how long is possible*" and "*Take a deep breath and say a sustained /a/ as soft and as loud possible*". Finally, 76.3% of diadochokinesis tests were impaired ("poor" and "fair"). In particular, 74% of performances were "fair" (Tab. IV).

Flexible endoscopic examination of swallowing

The findings of flexible endoscopic examination are shown in Table V. The oral phase lasted a mean of 21.8 seconds (SD 5.5).

Discussion

Among musculoskeletal diseases, dysphagia is best known as a complication of scleroderma. Nevertheless, the literature refers almost exclusively to the dysfunction caused by the oesophageal abnormalities [16]. In reality, the SS has numerous deleterious effects that compromise more than one stage of the swallowing process. Salivary dysfunction can be seen in up to half of patients with SS as demonstrated by Baron et al. [17]. Consistent with the literature, we observed the co-occurrence of Sicca syndrome in 47% of cases and 63% complained of a sensation of "*dry mouth*". Microstomia (decrease of the mouth opening) and microcheilia (decrease of the lip opening) are common manifes-

Table II. Mean prevalence of MDADI items with score > 1 in decreasing order.

		Prevalence (%)
Physical		
P7	It takes me longer to eat because of my swallowing problem	42
P4	I feel that I am swallowing a huge amount of food	26.3
P5	I limit my food intake because of my swallowing difficulty	26.3
P6	Swallowing takes great effort	21
P8	I cough when I try to drink liquids	21
P2	Swallowing is more difficult at the end of the day	15.8
P3	People ask me, "Why can't you eat that?"	15.8
P1	I cannot maintain my weight because of my swallowing problems	10.5
Functional		
F5	My swallowing difficulty has caused me to lose income	15.8
F2	I feel free to go out to eat with my friends, neighbors, and relatives	10.5
F3	My swallowing problems limit my social and personal life	5.3
F1	People have difficulty cooking for me	0
F4	I feel excluded because of my eating habits	0
Emotional		
E4	I am upset by my swallowing problem	26.3
E7	I do not feel self-conscious when I eat	15.8
E6	I have low self-esteem because of my swallowing problems	10.5
E2	I am embarrassed by my eating habits	5.3
E3	Other people are irritated by my eating problem	5.3
E5	I do not go out because of my swallowing problem	0

tations of SS that are reported to be present in 50-80% of cases [18-20]. Specifically, we observed microstomia in 47% of subjects. Erosions and resorption of mandible and temporomandibular joint involvement are common findings among SS patients and may explain the previously mentioned changes [21]. Overall, reduced oral opening and xerostomia interfere with speech, mastication and oral hygiene predisposing to oral and dental disease. In this regard, it is

Table III. Prevalence of score 0, 1, 2 and 3 for performance of the lip, mandible and tongue.

	0	1	2	3
Lips				
Opening	-	52.7%	36.8%	10.5
Extension	5.3%	26.3%	57.9%	10.5
Protrusion	-	31.6%	57.9%	10.5
Ability to hold a depressor between the lips	-	10.5%	68.5%	21
Exert force against resistance	5.3%	26.3%	52.7%	15.7
Mandible				
Opening	-	47.4%	42.1%	10.5%
Lateralisation	5.3%	36.8%	36.8%	21.1%
Protrusion	5.3%	36.8%	47.4%	10.5%
Tongue				
Protrusion	-	15.8%	57.9%	26.3%
Lateralisation	-	15.8%	57.9%	26.3%
Tongue tip elevation out of the mouth	31.5%	26.3%	36.9%	5.3%
Tongue tip elevation into the mouth	10.5%	26.3%	52.7%	10.5%
Circular movements around the lips	-	15.8%	63.2%	21%
Pop of the tongue	21%	52.7%	10.5%	15.8%
Vertical resistance	36.8%	36.8%	26.3%	-
Lateral resistance	5.2%	26.3%	47.4%	21.1%
Central resistance	-	15.8%	42.1%	42.1%

Table IV. Distribution of respiratory, phonatory and diadochokinesis performance.

	Poor	Fair	Good	Normal
Respiratory				
Take a deep breath, then make the /s/ sound for as long as you can	42.1%	37%	0	21.1%
Take a deep breath, then make the /s/ sound again, but start at a whisper then get louder	73.7%	21.1%	0	5.2%
Take a deep breath, then make the /s/ sound again, but start at a whisper then get softer	52.7%	42.1%	0	5.2%
After a deep breath say repeatedly /s/	47.3%	37%	10.5%	5.2%
Phonatory				
Take a deep breath, then make the /a/	26.3%	21.1%	36.8%	15.8%
Take a deep breath, then make the /a/ sound for as long as you can.	42.1%	36.8%	15.8%	5.3%
Take a deep breath, then make the /a/ sound for as aloud as you can.	26.3%	36.8%	26.3%	10.5%
Begin at your conversational level of speech, say /a/ and sing up a scale	36.8%	47.4%	10.5%	5.3%
Begin at your conversational level of speech, say /a/ and sing down a scale	42.1%	36.8%	10.5%	10.5%
Take a deep breath, then make the /a/ repeating the sound (a-a-a)	10.5%	57.9%	15.8%	15.8%
Diadochokinesis				
Open and close the mouth as many times as you can in 5 seconds	10.5%	79%	10.5%	0
Protrude and retract lips as many times as you can in 5 seconds	31.6%	63.1%	5.3%	0
Protrude and retract tongue as many times as you can in 5 seconds	82.4%	10.5%	5.3%	0
Raise and lower the tongue as many times as you can in 5 seconds	94.7%	5.3%	0	0
Move tongue from side to side as many times as you can in 5 seconds	89.4%	5.3%	0	5.3%

Table V. Percentage endoscopic of fiberoptic evaluation of swallowing findings.

Findings	N. cases	%
1. Laryngopharyngeal sensory deficit	11	58
2. Repeated dry swallows	10	53
3. Residue post-swallow	10	53
4. Mucous secretions in the pharynx and larynx	5	26
5. Facilitating manoeuvres	5	26
6. Dryness appearance of oropharyngeal mucosa	5	26
7. Deficit of oral bolus propulsion	4	21
8. Glottic incompetence	3	16
9. Delayed swallow initiation	2	11
10. Abnormal pharyngeal squeeze	2	11
11. Aspiration or penetration pre-, intra-, post-swallow	0	0

interesting to note that almost all our cases were edentulous and showed atrophic tongues.

Our results were not differentiated based on the stage of the disease. Nevertheless, the percentage of subjective swallow abnormalities resulted from the anamnestic list of symptoms was higher: dysphagia was present in 40-50% of cases and was mostly related to oral and oropharyngeal dysphagia. Among all symptoms, globus pharyngeal was reported in 58% of the sample versus 5-45% estimated in the general population, respectively, for persistent and intermittent globus pharyngeal [22,23]. We hypothesise that the prevalence increases in SS patients because of disease-related xerostomia, pharmacotherapy and other less well-understood processes involving immune-mediated mucosal changes and altered sensory perception. In our sample, the symptoms (i.e. "*feeling of food remaining in the mouth*" and "*feeling of food remaining in the lower throat*", "*difficulty to start swallowing*") are consistent with objective findings. First, the BSE showed post-swallowing oral residue, and, moreover, flexible endoscopic examination revealed dry swallows and post-swallow pharyngo-laryngeal residue in 53% of cases and deficit of oral bolus propulsion in 21%. Similarly, Montesi et al. [7] found abnormalities in the oral and pharyngeal phases during videofluoroscopy (VFS) in SS patients. Using VFS, Russo et al. [24] demonstrated the presence of intraswallowing laryngeal penetration caused by altered epiglottal motility in 57.8%, and pooling of contrast agent in the valleculae and/or pyriform sinuses in 51.1%. Nevertheless, it cannot be excluded that upper esophageal sphincter dysfunction is possible in SS or secondary to GERD which may cause this finding.

Gastro-oesophageal reflux (GERD) occurs in over 50% of SS patients, which causes symptoms mimicking swallowing disorder. In our sample, 37% of patients had a diagnosis of GERD and almost 70% complained of symptoms suggestive of GERD (raclage, sensation of food backing up into the throat or mouth). Thonhofer et al. [25] found a high prevalence of oesophageal disease in asymptomatic patients. However, further research specifically oriented to clarify the role of GERD on dysphagia in SS is necessary. Forty-two percent of cases reported onset of cough during swallowing and one-third of patients had had at least one episode of pneumonia by bacterial infection. This may be correlated to the multifactorial increased risk of aspiration in SS. First, we found "poor" or "fair" performances of respiratory, phonatory and diadochokinesis in about 80% of patients. Normally, eating, swallowing and breathing are tightly coordinated; the coordination of breathing and swallowing reveals a well-timed pattern between physiological respiratory events and related swallowing events, and vocal fold closure might be part of a protective mechanism that involves swallowing apnoea [26]. Secondly, it is known that the physiologic breathing cycle is not simply repressed during swallowing; it is substituted by a different and well-controlled behaviour pattern [27] that is sensitive to variations in bolus volume [28-30] and viscosity [31,32]. Moreover, direct stimulation of the laryngeal vestibule produces a reflex apnoea with abrupt vocal fold closure. In about 60% of SS patients, we demonstrated a decrease of laryngeal mechanosensitivity, probably resulting from GERD [33] that impairs the perception of bolus characteristics, which may consequently alter these mechanisms of control and increase the risk of aspiration. Thirdly, decreased pharyngeal muscular performance as demonstrated by the high percentage (53%) of dry swallows and pharyngo-laryngeal residue contribute to a further increase of the risk of post-swallow aspiration. Nevertheless, the absence of signs of aspiration or penetration was probably because the residual strength was sufficient to ensure good control of the bolus.

Although swallowing alterations are common in the immunomediated population [6], as seen herein, they are often overlooked by patients as well (general distress was clinically significant in 26.3% of cases with a mean "mild" impact) probably because of the predominance of other discomforts. Nevertheless, both early diagnosis and treatment of swallowing alterations is an issue that must be considered carefully. Towards this objective, it is important to evaluate the scores from questionnaires not in absolute terms, but in relation to the clinical background. Moreover, the risk in underestimating dysphagia may be reduced by associating physical and instrumental assessment to characterise the abnormalities in the oral and pharyngeal stages of swallowing [34].

In conclusion, this study is the first to highlight the importance of a multidimensional approach in swallowing evaluation in SS patients, which should include subjective and objective evaluation (the latter by a speech pathologist and phoniatric consultant). It also demonstrates specific features

of swallowing alterations to consider when addressing the high impact of the upper dysphagia in SS.

Several limitations should be considered when interpreting this investigation. In addition to the cross-sectional nature of the study, a control group is also lacking. In this regard, this work should be seen as preliminary, but can increase awareness in taking an otorhinolaryngologic approach to dysphagia within the complex framework in patients with SS. It can also encourage ENT specialists and rheumatologists to consider oropharyngeal dysphagia in the evaluation of patients suffering from SS.

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