ISSN 0025-7680

ORIGINAL ARTICLE MEDICINA (Buenos Aires) 2005; 65: 117-120

SWALLOWING DISORDERS IN PATIENTS WITH BLEPHAROSPASM

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Abstract Blepharospasm is a focal dystonia characterized by involuntary eye closure due to abnormal contraction of orbicular eyelid muscles. When blepharospasm is associated to the presence of involuntary oromandibular movements, it is termed Meige syndrome. The aim of this study was to investigate the presence of deglutition alterations in patients with concurrent blepharospasm and Meige syndrome. Twenty consecutive patients were studied by video fluoroscopy using a barium technique. The 4 stages of deglutition were investigated. Ninety percent of patients (18 cases) presented deglutition disorders. The more commonly found alterations were premature food drop, 15 cases (83%) and vallecuale residuals, 14 cases (78%). Sixty seven percent of abnormal findings occurred in the third stage of deglutition. Eighty-nine percent of patients (16) presented more than one swallowing alteration. There was a positive and significant correlation between the number of alterations and patient´s age or disease duration. Prevalence of swallowing disorders in the healthy elderly population is reported to be 44%. In our series it reached 90%, suggesting that our findings might be related not only with age but also with a more widespread dystonia exceeding the orofacial muscles.

Key words: blepharospasm, swallow, dystonia, Meige's syndrome

Resumen ***Trastornos deglutorios en pacientes con blefaroespasmo.*** El blefaroespasmo es una distonía focal caracterizada por el cierre involuntario de los ojos debido a la contracción anormal de los músculos orbiculares de los párpados. Cuando el blefaroespasmo se asocia a la presencia de movimientos involuntarios oromandibulares se denomina síndrome de Meige. El objetivo de este estudio fue investigar la presencia de alteraciones deglutorias en pacientes con blefaroespasmo y síndrome de Meige. Se incluyeron 20 pacientes consecutivos que fueron estudiados mediante vídeo fluoroscopia con técnica de bario. Se investigaron las 4 etapas de la deglución. El 90% de los pacientes (18 casos) presentó trastornos en la deglución. Las alteraciones más comúnmente halladas fueron caída prematura del alimento, 15 casos (83%) y residuos valeculares, 14 casos (78%). El 67% de anormalidades se observó en la tercera etapa de la deglución. El 89% de los pacientes (16) presentó más de un trastorno deglutorio. Se observó una correlación positiva y estadísticamente significativa entre el número de hallazgos patológicos y la edad de los pacientes y la duración de la enfermedad. De acuerdo a lo publicado, la prevalencia de desórdenes de la deglución en pacientes sanos de edad avanzada es del 44%. En nuestra serie alcanzó el 90%, lo cual sugiere que nuestros hallazgos podrían estar relacionados no sólo con la edad, sino también con la posibilidad de que el compromiso distónico en pacientes con blefaroespasmo sea mayor de lo que se aprecia clínicamente extendiéndose más allá de los músculos orofaciales.

Palabras clave: blefarospasmo, deglución, distonía, síndrome de Meige

Blepharospasm (BS) is a focal dystonia characterized by sustained contraction of the eyelid orbicular muscles, which causes involuntary eye closure. It is more common in women and appears in adults after middle age. At onset, the picture may present only with increased blinking, but in time contractions become intense, frequent and sustained. Typically, situations such as stress, intense light or watching television worsen palpebral occlusion[1].

Very severe cases of BS may give rise to what is known as "functional blindness", which consists in the lack of sight as a result of involuntary, sustained eye closure, in the absence of any demonstrable lesions in optic pathways or visual organs.

Meige syndrome (MS) is manifest by the association of BS with the presence of dystonic movements of oromandibular muscles[2].

Alterations in deglutition may be found in diverse neurological afflictions such as parkinsonian syndromes[3], multiple lacunar infarcts[4], or Huntington's disease[5], among

Received: 14-IV-2004 Accepted: 19-I-2005

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others. However, the presence of swallowing disorders in patients with BS or MS has not yet been studied.

We currently run a large dystonia Clinic at the Buenos Aires University Hospital where the most prevalent forms of adult-onset dystonia are represented by BS and MS cases.

As some of our patients have occasionally manifested mild deglutition disorders, we decided to investigate how frequent these alterations might be. BS and MS patients were lumped together as both represent a restricted form of cranio facial dystonia. The objective of the study was to investigate the presence of deglutition alterations in a population of 20 patients with idiopathic BS/MS.

Materials and Methods

Twenty consecutive patients,15 women and 5 males, were selected from our dystonic clinic. Fourteen patients presented idiopathic BS and 6 MS. Mean age of patients was 68.5 ± 2.4 years (range 44 to 83 years) and a mean disease duration 8.36 ± 1.5 years (range 3 months to 27 years).

Out of the 20 patients, 5 (25%) reported dysphagia, while the remaining 15 were asymptomatic. Besides BS, 6 patients (33%) presented oromandibular dystonia (OMD) MS, and 4 patients had one of the following neurological alterations: postural tremor in upper limbs in two, and restless legs syndrome and cephalic tremor in one each (Table 1).

All patients were studied by video fluoroscopy by means of a modified barium technique, investigating a) the presence of abnormal movements of the buccal cavity, larynx and pharynx at rest, b) alterations in phonation and c) deglutition in each one of its 4 stages using liquid, semisolid and solid substances in combination with barium sulfate.

In all patients the swallowing study was carried out within 45 days following BS treatment with botulinum toxin (*Botox*, *Allergan*) 2.5U injected at five sites over the orbicularis oculi, so that at the time of examination all were BS-free. None of them received injections on other facial or masticatory muscles.

The number of abnormal findings observed during video fluoroscopy was correlated with patient age and with disease duration by means of Pearson's method.

The prevalence of swallowing abnormalities in our series was compared with figures from historical controls in a normal aging population by means of the exact test, based on a binomial distribution.

Results

Overall, 18 (90%) out of the 20 patients presented deglutition disorders, 5 of whom (28%) reported dysphagia, while none of the remaining 13 (72%) reported swallowing difficulties.

Besides, in 15/20 patients (75%) the presence of abnormal mouth and pharynx movements at rest was observed, while 2 (10%) exhibited phonation dysfunction

TABLE 1.– *Neurological abnormalities of the 20 patients*

Patient Nº	Age	Disease duration	Type of distonía	Nº of deglutition alterations	Other neurological manifestations
1	69	27	BS OMD MS	3	postural tremor in upper limbs
2	79	18	BS OMD MS	4	
3	59	10	BS OMD MS	3	restless legs syndrome
4	63	8	BS	2	
5	67	5	BS	3	
6	53	2	B	0	
7	51	6	BS	2	cephalic tremor
8	76	5	BS OMD MS	3	
9	44	3	BS	1	
10	83	16	BS	5	
11	65	1	BS	3	
12	77	15	BS OMD MS	3	
13	73	10	BS	3	
14	79	10	BS OMD MS	4	
15	78	8	BS	2	postural tremor in upper limbs
16	81	3 months	BS	3	
17	74	10	BS	4	
18	66	7	BS	2	
19	64	4	BS	1	
20	69	2	BS	0	

BS: blepharospasm, OMD : oromandibular dystonia, MS: Meige syndrome

due to a velopharyngeal movement disorder which was not evident clinically.

Deglutition alterations found among the 18 affected patients are listed in Table 2.

Overall, 67% of such abnormal findings occurred in the third stage of deglutition, 21% in the second, 9.4% in the fourth and 3.7% in the first.

Eighty nine percent of the affected patients presented more than one pathological finding at video fluoroscopy examination.

When comparing the results obtained in the deglutition study with liquids, semisolids and solids, it was observed that with liquids the evaluation was normal in 16 patients and pathological in 4, whereas with solids and with semisolids it was normal only in 2 cases and abnormal in the remaining 18. No difference between solids and semisolids was found (Table 3).

On analyzing different variables, a positive and statistically significant correlation was found between the number of alterations recorded and patient age ($r = 0.46$ and $p = 0.04$), and between the number of alterations and disease duration ($r = 0.44$ and $p = 0.05$). In contrast, patient age and disease duration failed to correlate.

TABLE 2.– *Abnormal findings in the affected cases*

Deglutition alteration	Patients (n= 18)	%
Premature fall of food	15	83
Residuals (vallecula, pyriform synusses)	14	78
Lack of pressure difference between 2º and 3º deglutition stages	8	44
Slow evacuation	4	22
Alteration in triggering deglutition reflex	3	17
Lack of pressure difference between 3º and 4º deglutition stages	3	17
Alteration in bolus preparation	2	11
Passage to rinopharinx	2	11
Pharyngeal lacuna	1	5.5
Aerophagia	1	5.5

TABLE. 3.– *Abnormal findings with solids, semisolids, and liquids*

Substance	Nº of patients with abnormal findings	%
Liquids	4	20
Semisolids	18	90
Solids	18	90

Patients with MS had more swallowing alterations (mean 3.3) than those with isolated blepharospasm (mean 2.2).

On comparing the prevalence of deglutition disorders in our cases (90%) with the prevalence of similar alterations in elderly subjects (44%)[6] a highly significant difference was found ($p < 0.0004$).

Discussion

While swallowing disorders in dystonia have been poorly delineated, their association to BS/MS has not yet been reported in the literature.

The normal swallowing sequence involves a complicated interplay of neural reflexes between the brainstem and the muscles of the mouth, pharynx, larynx and esophagus[7-12]. In addition to the neuromuscular control of these structures, a variety of external factors such as changes in head and neck posture, can affect swallowing physiology[10-12]. Movement disorders like Parkinson's disease, chorea and dystonia are apt to interfere with the swallowing process either by modifying the relative positioning of the oral cavity and pharynx as a result of postural changes, or directly, through damage to swallowing centers in the CNS[10].

Normal deglutition comprises oral preparation, as well as oral, pharyngeal and esophageal stages[10-13]. The pharyngeal stage, which was the most affected among our patients, comprises the following components: a) elevation and closure of the velopharyngeal structures, b) elevation of the larynx, c) closure of the top of the airway from the vocal folds, d) pharyngeal peristalsis, and e) opening of the upper esophageal sphincter[10, 12, 14].

In this study, 90% of examined patients with BS/MS presented deglutition disorders, which were asymptomatic in 72% of cases. The greatest number of alterations was observed in the third stage, and the most frequent patho-

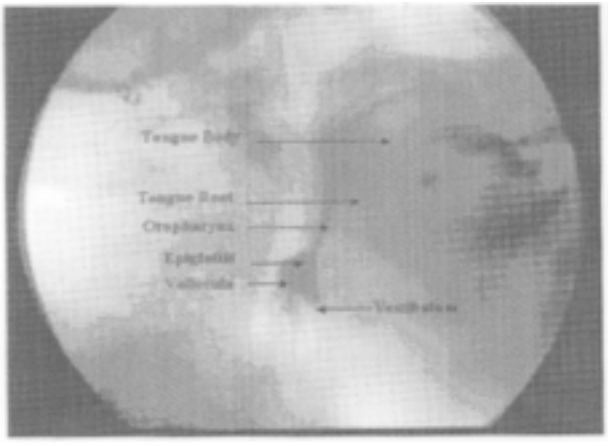

Fig. 1.– Lateral videofluoroscopy showing premature fall of food on the second swallowing stage with barium retention in vallecula, pharyngeal lacuna and laryngeal vestibula.

logical findings were premature fall of food and the presence of buccal residues (Table 3, Figure 1).

The analysis of our results indicates that disease duration and age positively correlate with more severe deglutition impairment. However, this positive correlation is only explained by age in a 46% (r = 0.46) and by disease duration in a 44% (r = 0.44) suggesting that dystonia is also responsible of the swallowing abnormalities observed in these patients.

The high prevalence of alterations observed in our group of patients suggests the possibility that BS/MS adds to age an additional risk factor for presenting deglutition alterations. In fact the prevalence of swallowing disturbances in normal elderly subjects has been reported to be 44%[6] whereas in our series it reached a significantly higher percentage ($p < 0.0004$) supporting a major role of dystonia rather than advanced age as the etiology of the deglutition disorder. Curiously enough, while swallowing difficulties in the normal aging population affects all phases of the swallow[6], our patients mainly showed involvement of the pharyngeal and esophageal phases. On the other hand, the study also shows that 75% of the patients had abnormal mouth and pharynx movements at rest which might worsen the swallowing disturbance. It could be argued that Botox may well have caused the swallowing disorder but this is highly improbable as our patients only received very low doses in the orbicularis oculi but not in the lower facial or masticatory muscles.

Our findings highlight that subclinical or even clinically evident swallowing alterations are frequent in patients with BS/MS, suggesting that these alterations may be part of the spectrum of cranial dystonia present in these patients.

Whether these alterations pose a risk of aspiration pneumonia and therefore early morbidity and mortality needs to be determined in a prospective study on patients with BS and MS.

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Above all else I have learned that there is no single path to creativity, not even within the stern halls of science. We are constrained not by the necessary discipline of rigor, but by the limits to our imaginations and to our intellectual daring.

Por encima de todo, he aprendido que no hay un solo camino hacia la creatividad, aun dentro de los austeros senderos de la ciencia. Estamos restringidos no por la indispensable disciplina del rigor, sino por los límites de nuestra imaginación y de nuestra audacia intelectual.

J. Michael Bishop

How to Win the Nobel Prize: An unexpected Life in Science. Cambridge MA: Harvard University Press, 2003, p 58