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LETTER TO THE EDITOR

Botulinum toxin and rehabilitation treatment in inclusion body myositis for severe oropharyngeal dysphagia

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Dear Editor,

Inclusion body myositis (IBM) is an idiopathic inflammatory myopathy considered the commonest acquired muscle disease among people over 50 years. IBM is characterized by a chronic progressive course, with proximal and distal muscular weakness [1].

Dysphagia has been previously reported in IBM, with an incidence of 65–86 % [1, 2], and it can be a presenting symptom. 10 % of subjects presents dysphagia at onset and 40 % complains of dysphagia at the time of diagnosis.

It is known that the presence of dysphagia leads to high risk of aspiration pneumonia and consequent poor prognosis. Dysphagia in IBM has a worse prognosis than in other myopathies; this could be attributed to poor response to therapy.

Diagnosis of IBM is difficult, and it often delayed. This is likely related to two reasons: IBM is an extremely rare disease, and the slow progression of muscle weakness is often attributed to aging. The gold standard to characterize IBM is the morphological, immunohystochemical, and immunopathological analysis of muscle biopsy [3]. Video-fluoroscopy and electromyography (EMG) studies demonstrate an abnormal pharyngeal phase, due to reduced cricopharyngeal (CP) muscle relaxation [3].

There is no definite cure for IBM, but several treatments have been suggested. Non-pharmacological interventions aim at avoiding non-use atrophy. Pharmacological treatments include immunosuppressants, tumor necrosis factor blocking agents, and lymphocyte depletion therapy, but these therapies do not maintain or improve muscle strength.

Therapeutic interventions for dysphagia in IBM include conservative measures (i.e., swallowing therapy, such as the Mendelsohn maneuver, and dietary modifications), pharyngoesophageal dilatation, and swallowing exercises. Cricopharyngeal myotomy is the surgical approach when conservative therapy fails. In this retrospective case-series study, we describe the effects of a multidisciplinary treatment consisting of rehabilitation combined with percutaneous Botulinum Neuro Toxin type A (BoNT/A) injection in the CP muscle in four subjects (3 females) (Table 1) with confirmed IBM and severe oropharyngeal dysphagia (OD). Among presenting symptoms, dysphagia was present in three subjects; all had a progressive weakness of the wrist and fingers, asymmetric in two subjects, and one subject presented also with an asymmetric quadriceps weakness. Clinical diagnosis was confirmed by biopsy, which showed inclusion bodies (3 plaque-like rounded bodies, 1 linear), rimmed vacuoles and in two cases reduction of type II muscle fibers. All underwent an unsuccessful trial of standard swallowing therapy; none of them had any previous balloon pharyngoesophageal dilatation or surgical procedure for treatment of upper esophagus (UES) dysfunction.

Swallowing function was evaluated with: VFS and/or fiberoptic endoscopic evaluation of swallowing (FEES) to determine CP dysfunction before and 1 month after the first BoNT/A injection; penetration–aspiration scale (PAS) to determine OD severity every 6 months during follow up (mean 4 years); electromyography (EMG) of CP and pharyngeal inferior constrictor (IC) muscles at diagnosis. EMG-guided bilateral CP muscle BoNT/A injection was

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Table 1 Characteristics of subjects and response

	Subject 1	Subject 2	Subject 3	Patient 4
Sex	Female	Female	Female	Male
Age at diagnosis of IBM (years)	73	74	72	82
Onset of dysphagia	At the time of IBM diagnosis	Few months after IBM diagnosis	At the time of IBM diagnosis	At the time of IBM diagnosis
BoNT/A injection (dose)	8 UI BTX-A	30 UI BTX-A (other type)	8 UI BTX-A	8 UI BTX-A
PAS pre-post (at 1st rehabilitation cycle conclusion)	5/2	6/2	7/2	7/7
Duration of response (months)	6	6	14	–
Follow-up (years)	5	3	5	3

performed if an electromyographic pattern of IC impoverished activation, and CP spasm was recorded. No adverse effects attributable to the BoNT/A injection were reported. Rehabilitation treatment with a speech therapist (40 min/day, 3 times/week for 1 month, performed after each injection) was started within the first week.

In three out of four subjects, post-injection VFS showed no sign of penetration/aspiration. Individuals reported a clinical benefit during the following months, and repeated BoNT/A injections every 6 months.

At follow-up (mean 4 years, range 3–5 years), they were still able to maintain a regular oral intake of viscous food with no aspiration or weight loss.

The fourth patient continued to complain of swallowing impairment after the first BoNT/A injection. VFS documented persistent aspiration. Two subsequent injections yielded no benefit. After 1-year surgical, CP myotomy was performed.

OD causes threatening complications due to the higher incidence of aspiration pneumonia in people with IBM. In these subjects, the synchronous activation of IC and CP muscles impairs the physiologic pattern of swallowing. Percutaneous injection of BoNT/A has been used to successfully treat CP muscle hyperactivity related to various neurologic diseases [3], but only one previous report [2] described satisfactory results after BoNT/A to upper esophagus sphincter in IBM. CP muscle spasm recorded by EMG predicts a favorable outcome of BoNT/A therapy. The improvement of swallowing follows soon after treatment and rehabilitation can be achieved early.

Other treatment strategies for dysphagia include muscle strengthening, pharyngoesophageal dilatation, intravenous immunoglobulin (IVIG), or CP myotomy.

The Mendelsohn maneuver aims at augmenting the extent and duration of laryngeal elevation, and consequently increase the duration of cricopharyngeal opening. To decrease CP muscle retraction, pharyngoesophageal dilatation (balloon dilation) has been suggested. However, dysphagia usually recurs. A trend toward improvement in dysphagia has been reported in a trial with intravenous immunoglobulin (IVIG) and in a small case series combining IVIG and steroids.

BoNT/A injection into the upper esophageal sphincter (UES) reportedly improves dysphagia in a heterogeneous population of individuals with cricopharyngeal muscle dysfunction [4]. The effectiveness of BoNT/A in IBM is still unknown. One study described improvement of dysphagia in two subjects affected by IBM after BoNT/A injection into the UES [5]. Conversely, no benefit after BoNT/A injection in another case report (2 subjects) has been reported.

One of our subjects did not respond to treatment possibly because of individual factors. This subject was the oldest one (82 years); we know that aging is independently associated with dysphagia, for which the definition of presbyphagia has been coined. In addition, older age often comes with a lower laryngeal position, especially in men. These characteristics could have made percutaneous treatment less accurate. In addition, tendency toward a more rapid decline in males than females and over the first 5 years after onset has been described, thus supporting the possibility of a more severe form in our male subject.

Here, we report a long-term improvement of swallowing function in IBM patients with OD after chemodenervation of the UES. This treatment could be suggested as first line treatment, alternative to more invasive procedures. Further studies on larger cohorts are required to confirm our results.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical standard Patient's anonymity was protected in this case report.

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