Botulinum toxin for the treatment of phonic tics in Tourette syndrome

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ABSTRACT

OBJECTIVE: To demonstrate the utility of botulinum neurotoxin (BoNT) injections in the treatment of phonic tics associated with Tourette syndrome (TS).

STUDY DESIGN: Case series at a single tertiary institution.

METHODS: Charts of patients less than 18 years old with TS and phonic tics who failed medical therapy and subsequently underwent unilateral submucosal thyroarytenoid (TA) muscle BoNT injections were reviewed. Duration of relief, effects on premonitory sensation, frequency of treatment, changes in symptoms and overall quality of life were assessed.

RESULTS: Four patients were treated with unilateral TA muscle BoNT injections. All patients experienced marked subjective improvement in phonic tics, echolalia, coprolalia as well as reduction in the premonitory urges associated with the vocal tics. The average number of treatments for each patient was 6.25, with 3 months in between each treatment.

CONCLUSIONS: We present a series of patients with TS who underwent TA muscle BoNT injections and experienced a reduction of symptoms of phonic tics and an improvement in quality of life. BoNT may interfere with a sensory reflex arc required to perpetuate tics and coprolalia in TS, possibly by reducing local buildup of tension or muscle contraction in affected areas. Further studies are required to determine exact mechanism.

INTRODUCTION

Tourette Syndrome (TS) is a complex, childhood-onset, neurobehavioral disorder characterized by chronic motor and phonic tics. Tics are sudden, rapid, recurrent, non-rhythmic motor movements (motor tics) or sounds (phonic tics). Phonic tics are motor tics that involve the oral, nasal, pharyngeal, laryngeal and respiratory musculature. Tics can be particularly disabling because it limits social activities and personal interactions. The etiology remains unknown, although evidence suggests a genetically determined dysfunction of the basal ganglia and limbic structures with varying clinical expressions in individuals.

A distinguishing feature of tics that separates them from other hyperkinetic movements is the presence of premonitory sensations, described as a generalized urge or local feeling of discomfort, tingling, or tension that precedes the tic. This consists of either localizable sensations or discomforts in the region of the tic, such as a feeling of “tension” or “tightness” in the neck or an uncomfortable feeling in the throat that is not relieved until the tic is executed. While medications such as dopamine-receptor blocking agents or dopamine depleters have been used to treat tics, these neuroleptics have some adverse effects such as tardive dyskinesia, hepatotoxicity, prolonged QT intervals, sedation, weight gain, depression.

Botulinum toxin (BoNT) has been used in the literature to treat conditions with excessive, abnormal involuntary movement. Local injections of BoNT are used routinely in the treatment of focal dystonia, hemifacial spasm, and other involuntary muscle spasms, including tics. We report a case series of patients with TS whose severe phonic tics who improved markedly have unilateral vocal fold injection of BoNT.

METHODS AND MATERIALS

Four patients with a tic disorder who fulfilled criteria for TS were included in this study. All subjects had received prior treatment with conventional neuroleptic drugs without improvement. Patients received injections of Botulinum Toxin (Botox, Allergan, Irvine, CA, USA) from 2008-2015 at the Otolaryngology –Head and Neck Surgery Clinic, Baylor College of Medicine, Houston, Texas.

TREATMENT: Treatment comprised of varying doses of BoNT injected into the unilateral thyroarytenoid muscle, under electromyographic guidance. Using a tekon coated injection, the needle was advanced through the cricothyroid space, and the vocal fold was entered. Position was confirmed with EMG signal on an /i/ vowel.

All patients had at least one follow-up visit. Data collected included (1) age at onset of tics; (2) subjective disability secondary to the TS; (4) frequency of treatments; (5) duration of relief; (6) subjective effect on premonitory sensation; (7) changes in symptoms; (8) duration of follow up; (8) overall quality of life; (9) presence of disabling or non-disabling complications.

RESULTS

4 patients were included in this study (all males).

- Mean age of TS diagnosis was 8.25 years (range 6-11). All patients had disabling coprolalia or echopraxia.
- Mean age at initial treatment was 14 years (range 12-16)
- Prior to BoNT injection, patients had experienced tics for a mean time of 5.75 years
- Average follow-up 25.2 months (range 12-40)

<table>
<thead>
<tr>
<th>Clinical variable</th>
<th>Results</th>
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<tbody>
<tr>
<td>Number of treatments</td>
<td>6.25 (4-10)</td>
</tr>
<tr>
<td>Dose per visit (Units)</td>
<td>17 (12-25)</td>
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<tr>
<td>Total dose (Units)</td>
<td>103 (75-182)</td>
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<tr>
<td>Time to response (number of days between injection and first signs of improvement)</td>
<td>7-21</td>
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<td>Total duration of benefit (weeks)</td>
<td>75</td>
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<tr>
<td>Time between injections (months)</td>
<td>3</td>
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<tr>
<td>Subjective decrease in premonitory sensation</td>
<td>All (n=4)</td>
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<tr>
<td>Complications</td>
<td>Dysphonia (n=3)</td>
</tr>
</tbody>
</table>

Table 1. Results of Botulinum Toxin injection

DISCUSSION

An interesting finding of this study was the amelioration of the premonitory sensory symptoms that often precede tics. The urge to produce vocalization was markedly reduced following BoNT treatment. The mechanism for the loss of premonitory sensation phenomena after BoNT injection is unknown, although interference with peripheral feedback mechanisms and a relief of local muscle contraction have been suggested to play an important role. It has also been suggested that premonitory sensations correlate with subclinical isometric muscle contraction, therefore the effect of BTX-A on muscle would explain its effect on such sensations.

Hoarseness was the only complication reported. This was not disabling to the patient, and resolution was noted within 1-3 weeks.

Limitations of this study include its retrospective nature. It is also a preliminary study that lacks the power to show significant differences in the measured variables. Additional patients and longer follow-up is needed to further evaluate the efficacy of BoNT in the treatment of tics.

REFERENCES