

## CASE REPORT

# Lingual Dystonia Treated with Botulinum Toxin - A Case Report

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### Summary:

*A 42-year-old female presented in Neurology Department of Sir Salimullah Medical College with gradually worsening difficulty in talking and eating for the last four months. Examination revealed dystonic tongue, macerated lips due to continuous drooling of saliva and aspirated lungs. She had no history of taking anti-parkinsonian, neuroleptics or any other drugs causing*

*dystonia. Chest X-ray revealed aspiration pneumonia corrected later by antibiotics. She was treated with botulinum toxin type-A. Twenty units of toxin was injected in six sites of the tongue. The dystonic tongue became normal by 24 hours. Subsequent 16 weeks follow up showed very good result and the patient now can talk and eat normally.*

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### Introduction:

In the early sixteenth century, Brueghel faced problem with open mouths and contracted facial muscles, similar to those we now associate with cranial dystonia<sup>1</sup>. In 1899, Gowers described conditions producing tonic and clonic jaw contractions. His differential diagnosis of tonic spasms included tetanus, trauma, hysteria, brainstem lesions and hypothermia. Convulsions, rigors, paralysis agitans, facial pain and chorea were recognized as causes of clonic spasms.

In 1910, Meige reported a syndrome of spasms of the eyelids in addition to contractions of the pharyngeal, jaw and tongue muscles. Characteristic of dystonia, these spasms were often provoked by voluntary action (e.g. talking, eating), or lessened by humming, singing, yawning or voluntarily opening the mouth. Some of the patients with Meige syndrome developed other signs of dystonia including torticollis or writer's cramp. In 1976, Marsden concluded that blepharospasm and oromandibular dystonia were adult-onset segmental

torsion dystonias<sup>4</sup>. Other reviews have supported this assertion<sup>2-3</sup>. Recently, the term 'oromandibular dystonia' (OMD) is used to mean the dystonia of the masticatory, lower facial and tongue muscles with resulting spasms and jaw deviation.

Involuntary lingual movements occur in a number of the conditions. The intrinsic tongue muscles are a complicated bundle of interwoven muscles with connective tissue septa. They change the shape of the tongue for speaking and swallowing. The extrinsic muscles also modify tongue shape, but, more specifically, they pull the tongue forward, upward, backward and downward. Hyoglossus flattens the dorsum of the tongue,

Systemic drugs have been the mainstay of therapy for dystonia, with anticholinergics, benzodiazepines or baclofen<sup>4</sup> being most effective. The combination of the dopamine-depleting agent tetrabenazine with lithium carbonate is quite helpful. However, in most patients, there is an inadequate response to pharmacotherapy or there are unacceptable side effects. Many authors have reported success in managing jaw closing OMD and tongue dystonia with local injections of botulinum toxin type A<sup>5-7</sup>.

After the emergence of botulinum toxin, there is a revolution in the management of dystonia. It is recommended that a neurologist, otolaryngologist and speech language pathologist should evaluate the patient before treatment, in order to confirm the diagnosis and assess other treatment options. A temporomandibular dysfunction specialist should be involved if there are signs or symptoms of temporomandibular joint dysfunction including joint pain and click or restricted jaw mobility.

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Treatment response could be assessed by rating scales. The disability rating scale-Columbia University Scale. This is a six-point rating scale. The second one is “The Global Clinical Rating Scale (percent of normal function scale)”. This is a linear scale, which assesses response to therapy graphically, especially when accompanied by the homework sheet. Ask the patient: ‘For the area of the body being treated, how would you rate your current level of function? Assume 0% represents fully disabled and no functional activity and 100% represents normal function.’ When used with the diary, it becomes a visual, graphic representation of the patient’s response to therapy and it can be enhanced by turning the diary counter-clockwise by 90 degree.

Correct muscle selection is the key to a good response to botulinum toxin treatment. Study the movements of the tongue to establish the muscles most likely to be involved and palpate them in different positions to determine which ones are contracting most actively.

Injection into orolingual structures requires a detailed knowledge of the local anatomy and of management of potential complications of therapy. Injections into the pterygoids need good experience or electromyographic (EMG) guidance as they can not be easily palpated. Other muscles could be injected by using anatomical knowledge and palpation but EMG guided injection gives more accuracy at the cost of more hazard. This is particularly important when performing follow-up injections. Gestures can help to target the toxin injection. They include opening the mouth when injecting the lateral pterygoid, and comparing opening the mouth versus protruding the tongue when injecting the digastric.

The dose should be adjusted according to the force of contractions, mass of muscle and weight of the patient.

All of the side effects resolved as muscle strength returned. There may be dysphagia in the tongue dystonia group and rarely which may be severe enough to need a change of diet.

#### **Case report:**

A 32-year-old right handed lady was admitted to Sir Salimullah Medical College Hospital in March 2005, complaining of gradual onset of difficulty in swallowing and talking for the last three months. Initially the problem was not severe and she could talk and swallow with some difficulty. The problem gradually became worsen. Drooling of saliva was added

to the previous problem, which made the life uncomfortable. Due to excessive salivation the lower lip became macerated. Recurrent chest infection was the common problem for the last one month. It was due to aspiration of saliva resulting from defective swallowing of saliva and liquid food. She did not give any history of taking levodopa, anticholinergic, chlorpromazine, phenytoin or antidepressant drugs. She also had no history suggestive of stroke or family history of similar disease. She was a normotensive and non-diabetic individual. On general examination, her vital parameters were normal. Her pulse was 80 beats/minute and blood pressure in right brachial artery in supine position was 120/80 mm of Hg. All the peripheral pulses including the carotids were well felt. On central nervous system examination, she was conscious, cooperative, well oriented in time, space and person. Examination of the motor system, sensations and deep tendon reflexes were normal except the dystonic tongue. The planter reflex was flexor in both the lower limbs. There were no cerebellar signs. Examination of the other systems was unremarkable.

Her haemogram, routine urine, stool examination and blood chemistry were normal. Her ECG was normal. Radiograph of her chest and skull were normal. Her computer tomography revealed no hypodense lesions.

*Treatment and follow up:* Initially, she was treated with benzodiazepines, anti-depressants and levodopa. She also visited abroad for this problem but no response was there. Rather the problems were increasing more and more with the passage of time. Finally she attended the “Botox Clinic” of SSMC where she was enrolled for botulinum toxin therapy.

Pre-therapy education was given to her. Different modalities of treatment of tongue dystonia and side effects of botulinum toxin were discussed with the patient. The patient decided to have botulinum toxin injection. Twenty mouse units of botulinum toxin was injected in six sites of hyoglossus and genioglossus.

Patient tolerated the procedure well and immediately after completion of injection she started to show improvement. Patient could swallow and talk. She became almost normal after twenty four hours. She was discharged from the hospital after two days. The disability of the patient was measured in Columbia University Scaling system. Initially her scale was six. After 24 hours the scale became three and after seven days which became one. Subsequent six weeks follow up showed excellent result.



**Fig.-1:** Before Botulinum toxin

#### Discussion:

The management of tongue dystonia has been revolutionized by the introduction of botulinum toxin<sup>8</sup>. Before the study of The Walton Centre for Neurology and Neurosurgery, Liverpool in 1984, there were no published guidelines for using Botox to treat focal dystonias<sup>5-6</sup>. In general, the approach should be empiric, beginning with small doses and titrating to the needs of the patient, selecting the muscles that on clinical examination had the greatest spasm.

The administration method as described by the Walton Centre for Neurology and Neurosurgery, Liverpool in 1984 was followed here and the result is consistent with those previously reported<sup>9</sup>. Smaller doses than Jankovic<sup>10</sup>, and adverse effects were less in this case. It is postulated that one can give a lower dose of toxin with significant benefit. EMG may be more important during follow-up, in order to avoid injecting into islands of muscle wasting.

In patients with tongue dystonia, the initial treatment typically gave adequate relief and needed no additional treatment in 16 weeks. After the initiation phase of treatment, 'boosters' should be discouraged because of concern about antibody development<sup>11</sup>. The patient stopped other drugs and continued to benefit from Botox injection for the last 16 weeks.

In 1990, botulinum toxin therapy was recognized by the American Academy of Neurology, the American Academy of Otolaryngology<sup>12</sup> and the National



**Fig.-2:** Seven days after Botulinum toxin

Institute of Health as safe and effective for many patients with dystonia, including those with jaw closing and tongue dystonias. Most experts now agree that botulinum toxin is safe and effective for other forms of jaw dystonia.

Botulinum toxin therapy for tongue dystonia is effective and there may be an unacceptable adverse effects including dysarthria, dysphagia, aspiration pneumonia, and therefore recommendation for injections into lingual muscles should be judicious.

Dystonia is one of the difficult neurological disorders usually not responding to pharmacotherapy. Botulinum toxin has made a big revolution in its management. Injections could be given in outpatients, except when patients who have severe jaw closing dystonia, need parenteral feeding until treatment permits resumption of oral feeding. Tongue dystonia is a very difficult situation and should be treated with 'Botox' if the patient is already unable to speak or swallow and can be very useful in the management of a protrusive tongue or painful spasms.

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