The Finding and Evaluation of EMG-Guided BOTOX Injection in Cervical Dystonia

Lian-Hui Lee[1], Wen-Neng Chang[1], and Chen-Sheng Chang[2]

Abstract- In this prospective study we report the results of EMG-guided BOTOX injections in a total of 15 cervical dystonia (CD) patients. Pre-treatment and post-treatment evaluations included physical examination results, Tsui ratings, and video recording. The dosage of BOTOX injection was determined by the EMG pattern, type of CD, and the degree of muscle hypertrophy. Seven patients underwent injections with and without EMG, and eight patients underwent injections with EMG-guidance only. The results showed that among the patients who underwent EMG-guided BOTOX injection there are: (1) fewer BOTOX-related side effects due to injection of the adequate dose of BOTOX to the accurate site of hyperactive muscles, (2) greater clinical improvement due to confirmation of hyperactivity in muscles in each type of cervical dystonia, (3) a better ability to reduce the amount of oral medication for treatment of muscle pain and spasms. We suggest that the use of EMG-guided BOTOX injections be considered for those CD patients with retrocollis, those who have had a sub-optimal treatment response to non EMG-guided BOTOX injections, and those with increased concern of side effects or a concomitant goal of reducing oral medications.

Key Words: BOTOX, Cervical dystonia, Dystonia ratio, EMG, Tsui rating scale

Acta Neurol Taiwan 2004;13:71-76

INTRODUCTION

Cervical dystonia (CD) is a neurological disorder characterized by involuntary contraction of neck and shoulder muscles associated with abnormal posture, head tremor, pain, and impaired voluntary volitional movement of the head. Cervical dystonia is difficult to treat with oral pharmacotherapy, although 40% of patients may achieve some degree of benefit from anticholinergics[1]. Therefore BOTOX injection is an important treatment for CD. Treatment with botulinum toxin A (BOTOX) is considered as a standard treatment for pain as well as the abnormal contraction of specific neck and shoulder muscles. Success in treatment of the postural abnormality requires successful identification of the involved muscles. Previous studies have demonstrated the benefits of EMG-guided BOTOX injections in CD patients when compared with those who received BOTOX injection based on clinical examination only[1-8]. The benefits of EMG guidance BOTOX injection have

Departments of Neurology, [1]Chang Gung Memorial Hospital-Kaohsiung, Kaohsiung, Taiwan; [2]E-DA Hospital, Kaohsiung, Taiwan.
Received December 12, 2003. Revised January 20, 2004.
Accepted March 15, 2004.

Reprint requests and correspondence to: Chen-Sheng Chang, MD, Department of Neurology, E-DA Hospital, I-Shou University Hospital, No. 1, E-Da Road, Kaohsiung, Taiwan.
E-mail: ed100234@edah.org.tw or jeansing@ms23.hinet.net

been reported previously[3-9], but BOTOX injection with EMG in conjunction with the use of the dystonia ratio[1,7] has produced detailed findings and is correlated with the spasmodic muscles and type of CD that has been rarely reported in the past. We conduct this study to confirm the difference in effects of BOTOX injection with and without EMG guidance in seven cases, and present the evaluation methods of EMG guidance as well as the major findings in each type of CD.

PATIENTS AND METHODS

Patient selection

Patients with cervical dystonia were eligible for the study. All patients underwent a complete neurological examination, and, when appropriate, neuroimaging studies (MRI or CT) were also performed. Eligible patients should have moderate to severe disability with a minimum Tsui score[1] of 11 prior to BOTOX treatment. We selected seven patients out of 20 who received previous BOTOX injection before without EMG guidance but with poor response. Eight remaining patients had not received BOTOX before. They joined this study without case selection due to the decision to treat all CD patients as patients that had received EMG guidance. Patients were treated with BOTOX injection with EMG guidance if 1) there is poor response without EMG guiding. 2) the injection sites of muscle can not be decided with palpation and the dystonia type. 3) the target spots are in the dorsal nuchal muscles in complicated type of cervical dystonia.

Evaluations

Patients underwent neurological examination at each visit (the initial evaluation, the day of injection, and weeks 2, 4, 8, 12 and 16 post-injection)[1]. The severity of muscle hypertrophy, muscle tightness as well as tenderness on palpation were measured[2]. Patients were evaluated by the Tsui scale[1]. Patients underwent digital videotaping while performing a series of head movements including head flexion and extension, rotation to the left and right, and tilting movement (attempted placement of the ear against the shoulder). Patients were videotaped using a standardized protocol[1,8], and the tapes were reviewed at the study conclusion. The patients were also asked to evaluate their quality of life, change in ease of voluntary head movement, posture, pain, muscle tightness and weakness (e.q. SMI or subjective movement improvement). These subjective results were recorded everyday in the first month, then recorded once a week from the second to the fourth month[1-8].

Selection of muscles for BOTOX injection without EMG

For those patients undergoing injections without EMG guidance, the muscles were selected based on palpation (areas of muscle hardness and tightness) and the clinical presentation of CD. Muscles were chosen based on to the possible causes of specific postural states[1,2]. The decision of shift to BOTOX injection with EMG guidance was made after two to three injections, based on clinical examination alone. Each BOTOX injection was performed around the end of the fourth month after previous injections.

Selection of muscles for BOTOX injection with EMG

Additional information was obtained from the assessment of muscle hyperactivity by needle EMG (the needle used was a Allergan 5980). The patient took a sitting position in a chair. The sites for the EMG-guided needle injection in the paraspinal muscle of the neck were divided into 4 to 6 points symmetrically distributed on each side. The needle was inserted from the superficial to the deepest muscle slowly. If spontaneous hyperactivity (at rest) was noted, the patient's head was turned in several directions to confirm the hyperactive muscle. Then maximal contraction was performed by the confirmed muscle. Other muscles than paraspinal muscles of neck (SCM, scalene complex, levator scapularis), were also checked with EMG. Muscles were evaluated both at rest and maximal voluntary contraction. Muscle hyperactivity was quantified based on the electrophysiological data. Dystonic muscle was differentiated from compensatory and co-contraction muscles by spontaneous and voluntary muscle activity (dystonia ratio)[1,7]. For each muscle a dystonia ratio was calculated based on the

number of turns of needle EMG activity at spontaneous contraction divided by that at maximal voluntary contraction. Abnormal EMG activity of the hyperactive muscle was defined as >100 turns/sec at rest[6].

BOTOX dosing determination

There are published guidelines for the typical BOTOX dose per muscle[1,6,7]. At Chang Gung Memorial Hospital Kaohsiung we have standardized our dosing regimens. The severity of dystonia, the relative mass of the muscle to be injected, and the risk, if any, of potential side effects associated with the injection into a specific muscle would influence the dose chosen. Typically each injection point receives 5 to 15 U of BOTOX in a volume of 0.10 to 0.30 cc. According to the size of the muscle there are usuall 1 to 5 injection points.

Adverse effects to BOTOX

Patients were asked to report any apparent side effects such as pain, tightness, weakness, muscle contraction and so on[1,8].

Additional treatment for CD

Medications included anticholinergic agents (trihexyphenidyl 2mg 1#/Tid) and benzodiazepines (diazepam 2mg or clonazepam 0.5mg I#/Tid) and muscle relaxants (baclofen 10mg 1#/Tid). All patients were instructed to complete a home-based daily exercise program of muscle stretching, massage and isometric exercise.

RESULTS

Fifteen patients of dystonia underwent injection and completed the study protocol. Seven were treated without EMG guidance (group B), and 15 were treated with EMG guidance (group BE). The clinical data of 14 assessable patients are listed in Table 1. In the B group, 3/7 reported 50% or more clinical improvement, and the Tsui score was improved by 50% or more in 5/7 patients. Among the patients in the BE group, 10/14 reported 50% or more clinical improvement, and the Tsui score improved by 50% or more in all but 1 patient (10/11) based on the video tapes and SMI. Despite EMG-guid-

Table 1. *Summary of the characteristics and responses to BOTOX treatment*

Patient # and Dystonia type	Baseline	B Post injection			BE Post injection		
	Tsui based on video	Tsui based on clinic	SMI	Able to reduce medication ?	Tsui based on video	SMI	Able to reduce medication ?
1. I (l't torsion)	11	5	30%	no	5	40%	no
2. I (r't torsion)	11				1	80%	yes
3. I with cranial (l't torsion)	11				4	40%	no
4. II (l't torsion)	11		70%	yes		60%	yes
5. II (l't torsion)	11	2	80%	yes	0	90%	yes
6. II (l't torsion)	12				4	50%	yes
7. II with cranial (r't torsion with tremor)	12	8	30%	no	6	40%	no
8. Iiaxial (r't torsion, l't tilt)	12				1	90%	yes
9. IIgeneralized (r't torsion with l't tilt)	14				8	50%	no
10. III (l't tilt)	11	4	50%	yes	2	65%	yes
11. IIIbrachial (r't tilt)	12					30%	no
12. IV+II (r't torsion)	12	6	30%	no	4	55%	yes
13. IV+II (r't torsion)	12	4	20%	no	1	90%	yes
14. Vgeneralized	12					50%	yes

Type I: Head rotated toward the side of shoulder elevation; Type II: Head rotation only; Type III: Head tilted toward the side of shoulder elevation; Type IV: Bilateral posterior cervical muscle spasm with elevation of the face (retrocollis); Type V: Bilateral anterior cervical muscle spasm with flexion of the face (antecollis). B: BOTOX without EMG; BE: BOTOX with EMG guidance; SMI: subjective movement improvement; ?: loss of follow up; IIgeneralized: Type II CD with generalized dystonia; IIaxial: Type II CD with axial dystonia (L truncal tilt); IIcranial: Type II cervical dystonia with cranial dystonia; III.brachial: Type III CD with l't arm dystonia; Vgen: Type V CD with generalized dystonia (patients of cerebral palsy patient). AED: antiepileptic drug

Table 2. *Summary of BOTOX doses and side effects*

Patient #	Dose of BOTOX (not EMG-guided)		Dose of BOTOX (EMG-guided)		Side effects (EMG-guided)
	# muscles	Total dose	# muscles	Total dose	
1	L't S. Cap.,Cerv. (70), L't L. Cap. (30),L't L.S (30), L't Trap. (20)	150U	L't S.Cap.,Cerv. (30/30), L't L. Cap.,Cerv. (20,20), L't L.S (15), L't S.C (15), Trap. (20)	150U	Weakness, pain
2			R't L.Cap.(40), R't Cap.,Cerv. (30,30), R't L.S (20), L't S.S (20).	150U	nil
3			L't S Cap. (30), L't S Cerv. (30), L't L. Cap. (30), Other (10). (50U for Cranial)	100U (+50U)	nil
4	L't S.Cap.,Cerv. (60), R't SCM (30), L't L (30), R't L.S (15), R't S.C (15)	150U	L't S.Cap., Cerv. (60), R't SCM (30),R't L.S (20), R't S.C (20), Trap. (20)	150U	Weakness, pain
5	L't S.Cap.Cerv. (80), R't SCM (30), R't S.S (25), R't L.S (15)	150U	L't S.Cap. (30),L't S.Cerv. (30), R't S.S Cerv. (30), R't L (20), R't L.S (15), Trap. (25)	150U	nil
6			R't S.S (30), R't S.C (15), R't SCM (30), L't S. Cap.,Cerv. (60)	150U	nil
7	R't S.Cap.,Cerv. (60), L't SCM (30), L't S.S (30)	120U	L't S.S Cap. (20), L't SCM (15), Bil. L.Cap. (30), R't S.S Cerv. (20), L't S.Cap.,Cerv. (20), R't SCM (15), (30U for cranial)	120U (+30U)	nil
8			L't S.S Cerv. (45), L't L.Cap (45), R't S.Cap.,Cerv. (45),	150U	nil
9			R't L't S.S (30),L't SCM (15), R't, L't. L (45,20), R't S.Cap. (30), R't S.Cerv. (20)	150U	nil
10	L't Trap. (50), L't L. S (30), L't S.S (30), L't S.Cap. (20), L't L (20)	150U	L't Trap. (45), L't L.S (30), L't L (45)	150U	nil
11			L't S.Cap. (30), R't L.Cerv. (40), R't SCM (30), R't S.S Cerv. (30), R't L.S (10)	150U	nil
12	Bil. S.S (40), Bil. L. Cap.,Cerv.(40), Bil. Subocci. (30), R't S.Cap. (20), Trap. (20)	150U	L't L.Cerv. (20), Bil. Subocci. (30), R't,L't S.S (20,40),R't L.Cerv. (30)	150U	nil
13	Bil S.S. Cap.,Cerv. (60), Bil. L. Cap.,Cerv. (60) Bil. Subocci (40)	150U	Bil.S.S (30,30),Bil. L.Cerv. (20,20), Bil.Subocci. (30), R't S. Cap. (20)	150U	nil
14			L't,R't SCM (30,40), Bil. S.C (20,20), L't S.Cap.Cerv. (20,20)	150U	nil
15			Bil. SCM (30,30), L't S. Cap. (20), R't S.C (20), L't S.C(20)	150U	nil

Bil.: bilateral; Crania: Cranial dystonia; L't: Left; R't: Right; L't L: L't longissimus; L.Cap.: Logissimus capitis; L.Cerv.: Longossimus cervicis; L.S: Levator scapulae; S.Cap.: Splenius capitis; S.Cerv.: Splenius cervicis; S.S:Semispinalis; Trap.:Trapezius; Subocci.: Suboccipitalis; S.C: Scalen complex; SCM: Sternocleidomastoid

ance, two patients with cranial dystonia (#3,7) patient with segmental CD (#11), CD (#1) and generalized CD (#9) responded poorly. 3/7 patients in the B group were able to reduce their medications, whereas, 9/14 patients in the BE group were able to reduce their medications (Table 1).

The BOTOX doses and side effects are reported in Table 2. Nuchal extensor weakness associated with local pain was reported in 2 female patients (#1,4) who had relatively small volume posterior neck muscles. However, the conditions resolved within 4-8 weeks. There were no reports of dysphagia or worsening of the CD.

The main abnormal activities of EMG were found in the semispinalis cervicis, longissimus (capitis, cervicis) and splenius (capitis, cervicis) muscles in type II, III, IV, and axial CD, and were found in the antecollis muscle in type II CD.

Table 3 shows the dystonia ratio of each patient. Calculation of the dystonia ratio helped to characterize the degree of abnormal EMG activity in specific neck and shoulder muscles. Abnormally hyperactivite muscle was defined by > 100 turns/s[1,4,6,7] at rest and the dystonia ratio[6] was given by the ratio between turns in needle EMG recording at spontaneous activity and maximal voluntary activation). In type I CD patients, torsion with ipsilateral shoulder elevation (#1,2,3), higher dystonia ratio was noted in the splenius capitis and splenius cervicis and ipsilateral longissimus muscles. In type II CD patients torsion only (#4,5,6,7,8,9,11), higher dystonia ratio was noted in ipsilateral splenius, and contralateral SCM, semispinalis, longissimus muscles. In type III CD patients (#10), neck tilting was usually ascribable to involvement of the scalene complex as well as the deeper postero-lateral neck muscles (semispinalis and longissimus), and shoulder elevation was effectively treated with injections into the levator scapulae and trapezius muscles. In type IV CD patients (#12,13), retrocollis was best treated with injections into deeper semispinalis, longissimus, suboccipitalis and bilateral splenius muscles. In type V CD patients (#14,15), there was antero-collis and higher dystonia ratio was noted in bilateral SCM and scalen complex muscles.

DISCCUSION

We evaluated patients with the Tsui rating scale[1], digital video, visual analog pain scale, and recordings of the patient's responses. Overall the mean change in the Tsui score was 58.31% in the injection group without EMG and 72.75% in the EMG-guided injection group. The mean response rate according to the patient (SMI: subjective movement improvement) was 44.29% in the injection group without EMG and 58.57% in the EMG-guided group. The patients undergoing EMG-guided injections obciously responses showed more favorable.

The use of EMG is especially helpful for cases of retrocollis (#13). Without EMG it is difficult to predict

Table 3. *Summary of dystonia ratios of selected muscles for each significantly improved patient after EMG guided BOTOX injection*

Patient # / CD type	Muscles and dystonia ratio of the main hyperactive muscles
2 / I	R't longissimus capitis (3), R't levator scapulae (3), R't splenius capitis and cervicis (3)
4 / II	L't splenius capitis (3), R't SCM (3), R't levator scapulae (3), R't scalene complex (3). Trapezius(2)
5 / II	L't splenius capitis (3), L't splenius cervicis (3), R't semispinalis cervicis (3). R't longissimus (2), R't levator scapulae (2)
8 / II axial	L't semispinalis cervicis (3), L't longissimus capitis (3), R't splenius capitis (3). R't longissimus capitis (3).
10 / III	L't trapezius (3), L't levator scapulae (3), L't longissimus (3).
12 / IV+ II	L't longissimus cevicis (3), Bil. suboccipitalis (3), Bil. semispinalis (3), R't longissimus cervicis (2). Trapezius (2)
13 / IV+ II	Bil. semispinalis (3), Bil. longissimus (3), R't splenius capitis (3). Bil. suboccipitalis(3)
15 / V generalized	Bil. SCM (3), L't splenius capitis (3), R't scalene complex (3), L't scalene complex (2).

All of above patients were improved more than 50% after EMG-guided BOTOX injection.

Type I: Head rotated toward side of shoulder elevation; Type II: Head rotation only; Type III: Head tilted toward side of shoulder elevation; Type IV: Bilateral posterior cervical muscle spasm with elevation of the face (retrocollis); Type V: Bilateral anterior cervical muscle spasm with flexion of the face (antecollis). IIgeneralized: Type II CD with generalized dystonia; IIaxial: Type II CD with axial dystonia (L truncal tilt); IIcranial: Type II cervical dystonia with cranial dystonia; III.brachial: Type III CD with l't arm dystonia; Vgen: Type V CD with generalized dystonia (cerebral palsy).

Dystonia ratio divided into 4 grades: Grade (0): no muscle activity, Grade (1): <50%, Grade (2): 50% to 75%, Grade (3): >75%.

SCM: Sternocleidomastoid, L't: left, R't: Right, Bil.: Bilateral.

accurately which muscle groups are involved. If there is rotation and anterocillis (#1) it is advisable to treat unilaterally (i.e. injection into the muscles ipsilateral to the side of rotation the splenius capitis and deeper longissimus on the same side). Unilateral injection the risk of over treatment, (head droop, etc). In female patients (#4,13) with relatively small muscle mass in the posterior neck, it is advisable to be particularly careful about the dose in the neck extensor muscles, because these patients seem to have a higher risk of head droop[7].

In type V CD (antecollis, #14,15) there was less EMG hyperactivity in the nuchal muscles. Injections into the scalene muscles and bilateral SCM may be more successful. It is also important to assess whether or not the prominent appearance of the SCMs[9] is due to the relative shortening of the SCMs (pure torticollis) rather than active dystonic contraction. EMG evaluation will help to clarify the situation.

In this study the EMG-guided BOTOX injection usually led to additional benefit in retrocollis (type IV), head tilting and shoulder elevation (type III) and most of type II cases. We experienced fewer positive results in patients with segmental dystonia (cranial plus cervical, axial, brachial dystonia) as well as generalized dystonia, although repeated injections at intervals of 3 to 4 months may lead to further improvement. Despite the complexity of type II patients with axial dystonia we were able to see excellent results of BOTOX in one such patient (#8). EMG demonstrated involvement of the left longissimus capitis, left semispinalis cervicis, right splenius capitis and right longissimus capitis muscles prior to BOTOX injection, and the dystonia ratio were >75-100% (grade 3) in all these muscles. Tsui score improved from a pretreatment score of 12 to a post-treatment score of 1. Subjective movement improvement was 90%. It is thought that the use of EMG guidance allowed us to identify the pattern of muscle activation and target our treatment more accurately and selectively.

This study suggests that EMG can help to 1) identify the hyperactive muscles in the posterior neck region (the depth of actively contracting muscle may be 30 mm or more); 2) differentiate dystonic muscles from the compensatory, co-contracting muscles; 3) enable selective imjection of BOTOX into those muscles with the most abnormal EMG activity and effectively avoid the non-involved muscle regions (so that there would be only minimal wastage of BOTOX) and minimal risk of side effects). Therefore, if the patient has a poor or sub-optimal response to BOTOX injection based on a muscle selection paradigm of palpation and a "guess" of the pattern of motor activation, EMG-guidance BOTOX injection is recommended.

REFERENCES

1. Ceballos-Baumann AO. Evidence-based medicine in botulinum toxin therapy for cervical dystonia. J Neurol 2001;248 (Suppl 1):14-20.
2. Brans JW, Aramideh M, Koelman JH, et al. Electromyography in cervical dystonia: changes after botulinum and trihexylphenidyl. Neurology 1998;51:815-9.
3. Klein AW, Mantell A. Electromyographic guidance in injecting botulinum toxin. Dermatol Surg 1998;24:1184-6.
4. Erdal J, Ostergaard L, Fuglsang-Frederiksen A, et al. Long-term botulinum toxin treatment of cervical dystonia- EMG changes in injected and noninjected muscles. Clin Neurophysiol 1999;110:1650-4.
5. Rollnik JD, Matzke M, Wohlfarth K, et al. Low-dose treatment of cervical dystonia, blepharospasm and facial hemispasm with albumin-diluted botulinum toxin type A under EMG guidance. Eur Neurol 2000;43:9-12.
6. Comella CL, Buchman AS, Tanner CM, et al. Botulinum toxin injection for spasmodic torticollis: increased magnitude of benefit with electromyographic assistance. Neurology 1992;42:878-82.
7. Fuglsang-Frederiksen A, Ostergaard L, Sjo O, et al. Quantitative electromyographical changes in cervical dystonia after treatment with botulinum toxin. Electromyogr Clin Neurophysiol 1998;38:75-9.
8. Richardson D, Sheean G, Werring D, et al. Evaluating the role of botulinum toxin in the management of focal hypertonia in adults. J Neurol Neurosurg Psychiatry 2000;69: 499-506.
9. Dressler D, Rothwell JC. Electromyographic quantification of the paralysing effect of botulinum toxin in the sternocleidomastoid muscle. Eur Neurol 2000;43:13-6.