

A Clinical Practice on Treatment of Spasmodic Torticollis, Meige's Syndrome and Craft Palsy

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Abstract

Objective: To study the therapeutic efficacy of botulinum toxin type A (LANTOX) for spasmodic torticollis, Meige's syndrome and craft palsy.

Method: 20 patients with spasmodic torticollis, 5 with complete type Meige's syndrome and 1 with craft palsy were treated by LANTOX, the therapeutic efficacy and side effect were observed.

Result: The Tsui scores showed a significant reduction after LANTOX injection for 20 patients with spasmodic torticollis, out of 5 patients with complete type Meige's syndrome, 4 were remarkably relieved, and 1 patient with craft palsy was completely cured. No allergic reaction and severe side effect were noted.

Conclusion: LANTOX local injection is an effective mean for spasmodic torticollis and other dystonia.

Key words: Botulinum toxin type A; Spasmodic torticollis; Meige's syndrome; Craft palsy

Dystonia or myodystonia is the disease or syndromes clinically characterized by involuntary, abnormal or excess contraction of some parts of muscle. The treatments include oral taken medicines, acupuncture and moxibustion, physiotherapy and surgery, etc, but the efficacies are unsatisfied or recurrent occurs. Since LANTOX has been applied for treatment of dystonia, the effects is encouraged. Especially for the idiopathic blepharospasm, facial spasm and other focal dystonia, the injection techniques have been very mature. However, since there are many differences between clinical characteristics of spasmodic torticollis, Meige's syndrome and craft palsy, the dosages and ways of LANTOX injection are different. In the past 4 years, we totally cured 200s patients (1000cases) with dystonia, and obtained obvious effects recently^[1]. The results of treatment of spasmodic torticollis, Meige's syndrome and craft palsy are reported as follows.

Information and Method

1. Clinical Information

During Feb 1998 – May 2002, our hospital had 20 cases of spasmodic torticollis

through neurology inpatient service, among them 8 were males and 12 were females, aged from 19 – 65 years, medical states varied from 1 month – 10 years, all conditions could not be controlled by medicines (e.g. Tegretol, Artane, Rivotril, Myonal, Haloperidol, Madopar, etc), acupuncture and moxibustion, physiotherapy, Chinese medicines, etc. 2 cases did the sternocleidomastoid muscle lysis surgery, no remission occurred after surgery; 1 case had rectum cancer and chemotherapy history; 10 cases processed cranial CT/MRI inspection, one of the case showed “cerebral infarction”; 12 cases did the routine cervical vertebrae radiography, one showed stenosis in cervical vertebrae duct, 4 cases merged with hyperosteoegeny in cervical vertebrae, one showed flexion deformity of cervical vertebrae, hyperosteoegeny in cervical vertebrae accompanied slight slippage. All patients of spasmodic torticollis had no family history and other special nervous system diseases. 5 cases with complete type Meige’s syndrome, one was male, 4 were females; aged from 45 – 61 years; medical states varied from 6 months – 5 years, one of the cases accompanied spasmodic torticollis, all conditions could not be controlled by oral taken medicines, 2 cases did the cranial CT inspection but no abnormality observed. One case was a 32-years-old male patient of craft palsy, processed spasm in left hand extensor muscle when studying hair dressing. No other location characteristics observed by nervous system checking. No abnormality found in serum copper, ceruloplasmin, serum calcium and liver functions upon checking. No improvement observed after taking various kinds of medicines.

2. Medicines and Injection Method

The LANTOX used is produced by Lanzhou Institute of Biological Product [(97) Drug Approval (Lan) S-01]. The formulation is a lyophilized toxin, there are 4 kinds of vials: 40U, 50U, 65U, 100U. It should be stored protect from light inside -5°C to -20°C fringe, diluted to 2.5U/0.1ml by 0.9% saline before use. Multipoint local injection was processed by using 1ml syringe, the dosage and injecting points were decided according to the size, amount and thickness of the spasmodic muscle. Generally speaking, in case of spasmodic torticollis: 6 – 12 points for sternocleidomastoid muscle, 3 – 5 points for levator muscle of scapula, 5 – 12 points for upper part of trapezius muscle, 4 – 6 points for splenius muscle of head and posterior deep cervical muscle, 7.5U for each point. Some patients needed extra injection of 4 – 6 points, each point 5U at group of scalene muscle. Total dosage for each patient was 100U – 300U. In case of Meige’s syndrome: generally refer to basic injecting points of facial spasm, increase or decrease the injecting points according to the location and level of spasm, injection point(s) between glabella, masseter and temporal muscle would be added when necessary, 2.5U for each point. In the case of

craft palsy, 6 points for the common extensors of left hand and extensor of index finger, 10U for each point, totally 60U.

3. Efficacy Standard and Evaluation

Scores were given according to Tsui *et al's*^[2] typing for spasmodic torticollis:

A: Degree of head obliqueness (0 – 9 marks)

Distortion (0 = nil, 1 < 150, 2 = 15 – 300, 3 > 300)

Inclination (0 = nil, 1 < 150, 2 = 15 – 300, 3 > 300)

Flexion (0 = nil, 1 = light, 2 = medium, 3 = severe)

A = sum of three

B: Time of head obliqueness (0 – 2 marks)

1 = interstitial; 2 = continual

C: Elevation of shoulder

1 = slightly interstitial; 2 = slightly continual or severe interstitial;

3 = severe continual

D: Shivering of Head or convulsion (0 – 4 marks)

Degree: 1 = light; 2 = severe

Time: 1 = sometime; 2 = continual

D = Degree X Time

Total marks = A X B + C + D, the highest score was 25, a decrease of 0 – 10% after treatment was defined as ineffective, 11 – 50% as partial remission, 51 – 80% as obvious remission, 81 – 100% as complete remission^[3]. Refer to the typing standard of blepharospasm and facial spasm^[1] for the diagnosis scoring of Meige's syndrome. The efficacy of craft palsy was evaluated according to the subjective sensation of patient and improvement of clinical characteristics.

4. Follow-up Visits

Visited inpatient after 2 – 3 weeks of LANTOX injection, observed and recorded efficacy and adverse responses, processed evaluation scoring, afterwards visited regularly via phone or during return visit.

Result

1. Spasmodic Torticollis

20 patients' Tsui's scores before injection were 7 – 18, with an average of 12.75 ± 4.23 ; after treatment the scores were 2 – 10, with an average of 5.00 ± 2.13 , there was obvious difference compared the two (t test, $P < 0.05$). Complete remission 3/20 cases, 15%; obvious remission 10/20 cases, 50%; partial remission 7/20 cases, 35%; none

was ineffective. All effective within 7 days after injection, 2 cases effective within 3 days after injection. Efficacy duration ranged from 11 weeks – 9 months, with an average of 14.45 ± 5.87 weeks, patients without recurrent and had not reached 3 months were still under observation. Recurrent characteristics of patients were all improved by different degrees when compared with conditions before treatment. The dosages were the same or reduced in second injection, efficacy did not decrease. In one case, the duration of efficacy was 12 weeks after the first treatment, and attained 9 months of duration after the repeated injection. In other 2 cases the patient took half dosage of Artane and Rivotril after injection, the control was good. No patients processed dysphagia, forceless in elevating head, anaphylactic reaction and general adverse effects during visiting periods.

2. Meige's Syndrome

Among the 5 cases of complete type Meige's syndrome, 4 with obvious remission and 1 with partial remission after LANTOX injection treatment. All effective within 3 days, duration of effect was 10 – 24 weeks. One of the cases had unclear vision after injection, and this symptom disappeared after about 3 weeks. The symptoms of recurrent patients were improved after treatment, and reinjection was effective.

3. Craft Palsy

For the one case of left hand extensors spasm, the LANTOX injection was effective at the 3rd day after treatment. The spasm symptoms were completely remised, but the straighten of left middle finger was not quite well, the degree was light, disappeared after 3 weeks, the efficacy have been maintaining for 5 months up till now, no recurrent, still being visited.

Discussion

Spasmodic torticollis is a common focal dystonia disease of neurology, patients are usually middle-aged, the reason is still not cleared. Symptoms in most patients are ache feeling or hypertrophy of superior and anterior muscle of neck, severity may increase upon exciting emotions, decrease when the head gains support, and disappear during sleeping. Clinically it is more commonly to involve sternocleidomastoid muscle, trapezius muscle and cervical splenius muscle. According to its clinical expression, it can be classified into five types: distortion, side dump, hypsokinesis, antexed and mixed. Distortion is the common type^[4]. Previous treatment includes oral-taken medicines, physiotherapy, acupuncture and moxibustion, Chinese medicines or surgical treatment, some can attain short-term efficacy, but recurrent is likely to occur or patients can not tolerate the medical side effects thus terminate

treatment. Local injection of LANTOX paralyzes muscles by inhibiting release of acetylcholine from presynaptic membrane, and have obtained certain efficacy on the treatment of spasmodic torticollis. However, the efficacy is relatively lower when compares with that of focal spasms (e.g. belpharospasm, facial spasm). It is because:

- i) There are many types of spasmodic torticollis, many muscles are involved and complicated;
- ii) Some of muscles contract actively whereas the others contract passively, and it is difficult to differentiate clinically;
- iii) The location of muscle of neck is deeper, the structure of the surrounding parts are complicated and with important functions;
- iv) Different individuals have different medical sensitivity towards LANTOX, thus make it difficult to localize the involved muscles accurately and decide suitable dosage.

Even under guidance of EMG, the results are often unsatisfactory. During the treatment, we noticed the followings: firstly the patients should be kept in a specific posture of torticollis. The hypertrophy muscles under spasmodic conditions were observed and touched; the dosage and injection points were decided preliminarily with reference to experience. For example, for sternocleidomastoid muscle, 8 – 10 points of injection are made, 7.5U for each point, each injection can be divided into deep and shallow, the total dosage is 50U – 100U. This can prevent suffers of patients and difficulties of operation caused by too many injection points, and also avoid the side effects of dysphagia and forceless of muscle of neck caused by high concentration of drugs. For patients of first injection, the dosage should be small, then increase gradually according to the efficacy of first treatment. Generally, the dosage for spasmodic torticollis is more suitable to be ranged from 100U – 300U. For patients that are fat or had unsatisfied effects for the first treatment, it is better to localize under EMG guidance. If there is no EMG guidance for localization, one can use the feeling of hands (e.g. toughness, resistance of injection) to make sure that the syringe had achieved the spasmodic muscle upon injection.

It should be noticed that for the symptomatic torticollis caused by the innate torticollis caused by innate abnormality of skeletal muscle (innate cervical deformity, fibrosis after haematoma of sternocleidomastoid muscle), local pain excitement (myositis of muscle of neck, cervical lymphadenitis) and the disturbance in balance of the eye muscle (e.g. paralysis of musculus obliques dorsalis); and hysterical torticollis, are generally not suitable for LANTOX local injection treatment. Thus one should strictly understand the diagnosis standard of spasmodic torticollis when diagnosing.

Meige's syndrome is also called Brueghel syndrome, which is blepharospasm with oromandibular dystonia, and is clinically divided into 3 types: (i) blepharospasm; (ii) blepharospasm combines with oromandibular dystonia; (iii) oromandibular dystonia. Among the types (ii) is complete type, (i) and (iii) are incomplete types. This syndrome mostly occurs at middle-old ages, usually expressed by weird and various movements such as tightly closing of eyes, frowning, angular retracting, mouth opening and mumping, contracting of lips, extending and twisting of tongue, grinning and gnashing, etc. Spasms usually initiated by speaking and chewing, can be reduced by touching of chin and compression of mentum, and will disappear during sleeping. In a few cases, the spasms can spread to neck, throat, pharynx, muscles of abdominal and trunk, thus express as disturbance in pronunciation, difficulties in breathing and torticollis. The efficacy has been quite unsatisfactory in many years. Recently, it is thought that usage of LANTOX in treating this syndrome is safe and convenience, the short term effect is satisfied. However, since complete type Meige's syndrome invades more muscles, especially muscles of pharynx, it is difficult for injection than that of idiopathic belfharospasm. In our treatment on 5 cases of complete Meige's syndrome, 4 cases had obvious remission, 1 case had partial remission. For the case that merged with disturbance in pronunciation, the spasm of throat muscle had not been treated. This kind of patients need a ear-nose-throat doctor to confirm and type by laryngeal mirror, inject into throat muscle under EMG guidance to achieve a more satisfactory results.

Craft palsy refers to the conditions when a person doing long time and delicate action by hands due to his job nature, his hands or includes forearms process spasms or dystonia, but normal when doing other activities, and do not have other positive characteristics of nervous system. It is likely to occur in adult, but the reason has not been known well. Usually oral taken medicines, massage, physiotherapy, acupuncture and moxibustion, surgical operation, etc. do not have good efficacy, but local injection of LANTOX has ideal effect. Our case of craft palsy had tried several kinds of medicine treatment for over a year but no remission, after only 3 days of LANTOX injection, the effects were obvious, but processed weaken of injected muscles, which remised spontaneous after about 3 weeks. The efficacy has been maintaining for over 5 months, and is still under observation.

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