Botulinum Toxin Type A in Treatment of Meige's Syndrome

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Meige's Syndrome, also called blepharospasm-oromandibular dystonia, is a primary local dystonia, the cause of disease and onset mechanism are still not cleared. In the past, patients were usually given the antispastic drugs and sedative, but the curative effects were usually unsatisfactory. In 1980, Scott^[1] successfully applied botulinum toxin type A in treatment of strabismus. In 1993, same kind of product was produced in China and started to be used in clinical treatment of local dystonia, the effects were good and safe^[2]. Our department applied botulinum toxin type A in treatment of 12 cases of Meige's Syndrome since March 1996 and processed follow visit for 8 – 20 months, the effects were better. The results are reported as follows.

Subject and Method

1. Subject

We treated 12 cases of Meige's Syndrome, among those there were 7 cases of blepharospasm type, 2 cases of blepharospasm-oromandibular type and 3 cases of oromandibular type: 5 cases of male, 7 cases of female, age ranged from 35-62 years old, with an average of 48.5 years; course of disease ranged from 10 months – 16 years. Before treatment, cranial CT examination in 7 cases all showed normal results, cranial MRI examination in 2 cases showed normal results, EEG examination in 12 cases were normal. In the past, the patients tried haloperidol, neostigmine, carbamazepine, phenytoin sodium, acupuncture and moxibustion, etc, and all of them do not have obvious effect.

2. Method

Before and after treatment, the spasmodic strength was classified into different levels^[3]. Level 0: no spasm; level I: slight spasm upon external stimulation; level II: slight spasm, observable jitter, no dysfunction; level III: medium, obvious spasm, with little dysfunction; level IV: serious, severe spasm and dysfunction, affect work and life.

Botulinum toxin type A for therapy produced by Lanzhou Institute of Biological Products was used. It should be stored in a -5° C $\sim -20^{\circ}$ C fringe, diluted to 25U/ml by saline when use. Multiple injections were processed. For blepharospasm, injection was processed at bilateral orbicular muscle, internal and external of upper and lower

eyelids, subcutaneous of outer canthus of eye (distant from eyelid for 0.5cm); for spasm of orbicular muscle of mouth, injection was processed for 3 ~ 5 points at a distance of 0.5cm from upper and lower lips and mouth angle; injection for other body parts was adjusted according to location, amount and size of spasmodic muscles, 2.5U for each point. For patients with remaining spasm, supplementary injection can be processed within a week, total dosage was 50 ~ 75U.

Result

In this study, botulinum toxin type A was used in treating 12 cases of Meige's Syndrome. Spasmodic strength of patients before treatment: 1 case of level II, 8 cases of level III, 3 cases of level IV. Spasmodic strength of patients after treatment: 2 cases of level 0, 6 cases of level I, 4 cases of level II. After Chi-square test, P<0.001, the difference is very significant. The treatment was effective in all cases, and efficacy was 100%, time for onset ranged from 1 to 7 days. There were 2 cases whose spasm temporarily emphasized in the 2 days after injection, afterwards decreased gradually, sustained for 8-24 weeks. Most recurrent cases were partial recurrence, still less severe than that before treatment. There were 6 cases of reinjection whose spasm could be relieved again, and the relief period was prolonged for 8-10 weeks compared to the first remission period. Side effect: after injection, there were 8 cases of slight myasthenia in the injected muscle, expressed as forceless in close of eyelid in 6 cases, slight drop of mouth angle in 2 cases and slight myasthenia of masseter in 2 cases. The effect mostly occurred after $1 \sim 6$ days of injection, all naturally relieved in $3 \sim 5$ weeks. No allergic reaction observed.

Discussion

Meige's Syndrome is a chronic progressive nerve muscle disease of extrapyramidal. It was firstly described by Henry Meige in 1910. Its clinical characteristic is blepharospasm and/or mouth and face part symmetrical irregular multiple contraction. Mostly onset was started by idiopathic spasmodic pumping, and the blepharospasm is mainly expressed as contraction of orbicular muscle of eye, the centre is located at glabella, the forehead and nose contract, the patients tightly close both two eyes, folding occurred in glabella.

For the patients with mandiblular spasm, except blepharospasm, nose side, surrounding of mouth and tongue muscle idiopathic contract to the inner part, accompanying frequent projection and recession of mouth, exposure of teeth, odontoprisis, grinding teeth, projection and recession of tongue, this involuntary action does not have rhythmicity. Tetanic spasm followed by idiopathic pumping. All

symptoms emphasized during emotional tension, and disappeared during sleeping. The cause of disease and onset mechanism is still not cleared, no clinically special treatment for that disease. Our department applied botulinum toxin type A to treat 12 cases of patients with that disease, and found it effective in all cases, short-term effect is good and safe, side effects are slight, can process repeated injection, and gained curative effect again.

Botulinum toxin type A is an exotoxin produced by fusiform *Clostridium botulinum* under anaerobic environment. After injection, it actions on local nerve muscle joint, inhibits release of acetylcholine by motor nerve endings presynaptic membrane, make muscles relax and paralysis, thus leads to improvement in local muscle spasm symptoms^[4,5]. The results of this study proved that application of botulinum toxin type A in treatment of Meige's Syndrome is highly effective, safe, with little side effects and can be completely recovered, no systemic side effects, easy to operate, easy to be used clinically, but recurrent occurs in most cases, repeated injections are needed and still have effects. Botulinum toxin type A local injection in treatment of Meige's Syndrome is a method that is worth to be promoted.

References

- 1. Scott AD. Botulinum toxin injection into extraocular muscles as an alternative to strabismus surgey. Ophthalmology, 1980, 87:1044.
- 2. Tang XF. Botulinum toxin type A in the treatment of focal dystonia and hemifacial spasm. Chinese Journal of Neurology, 1996, 29(6):111.
- Jankovic J, Kenneth S, Schwartz PA. Longitudinal experience with botulinum toxin injection for treatment of blepharospasm and cervical dystonia. Neurology, 1993, 43: 834
- 4. Schantz EJ, Johnsos EA. Properties and use of botulinum toxin and other microbial neurotoxins in medicine Microbiol Reviews 1992, 56: 80.
- 5. Olney RK, Aminoff MJ, Lowenstein DH. Neuromuscular effects distant from, the site of botulinum neurotoxin injection. Neurology 1977, 38:1780.

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