

CONGENITAL MUSCULAR TORTICOLLIS WITH SHORTENING OF RIGHT STERNOCLEIDOMASTOID MUSCLE. A CASE REPORT

Djohansjah Marzoeqi, Ferdinand

Department of Plastic Surgery

Airlangga University School of Medicine

Dr Soetomo Teaching Hospital, Surabaya

ABSTRACT

To date, the pathogenesis of Torticollis remains controversial. Torticollis may be caused by trauma, infection as well as congenital abnormality. Approximately 10% - 20% of torticollis patients belong to those in post-traumatic category. Torticollis treatment can be provided as medical therapy, physiotherapy and surgery. Surgical procedure is performed either as muscular incision or the incision of sternocleidomastoideus tendon using common or endoscopic incision. Another treatment for Torticollis is by means of intramuscular injection of Botulinum Toxin. This therapy is given particularly when the primary cause of Torticollis is muscular spasm. Botulinum Toxin acts by inhibiting synaptic neurotransmitter that results in transient muscular attrition. The shortage of Botulinum toxin therapy is the formation of Botulinum Toxin autoimmune, so that increased dose is needed to maintain the effects of therapy. An 8-year old male child visited the clinic with a complaint that his head turned leftside and leaned to the right. The patient was diagnosed with Torticollis and subjected to correction with tenotomy and musculotomy of right sternocleidomastoid muscle.

Keywords: *torticollis, musculotomy, tenotomy, botulinum toxin, sternocleidomastoideus*

Correspondence: Djohansjah Marzoeqi, Department of Plastic Surgery, Airlangga University School of Medicine, Dr Soetomo Teaching Hospital, Jl Mayjen Moestopo 6-8, Surabaya, Phone 62-31-5501316

INTRODUCTION

The word torticollis is originated from Latin, tortus and collum. Tortus refers to a rotated or inclined position, while collum means neck. Torticollis is a symptom occurring in neck where there is a contracture in one side of sternocleidomastoideus muscle. Thereby, unconsciously, the head is rotated to lesion side of neck muscles, and, conversely, the face and chin of the patient is rotated to the normal side. Torticollis is not a particular disease, but a symptom that occurs according to the pathophysiological process of the underlying disease (Azizkhan et al. 2003, Thompson 2004, Ross 2005).

Torticollis is differentiated into two major classifications, congenital muscular torticollis and acquired muscular torticollis. Several terms commonly used in explaining such condition are acutewryneck, rotational torticollis, idiopathic spasmodic torticollis, cervical dystonia, head and neck dystonia. In the classification of pediatric fibrous tumor, torticollis belongs to the group of fibromatosis colli (Holt 1940, Raffensperer 1990, Azizkhan et al. 2003, Thompson 2004, Ross 2005, Encyclopedia Index).

Torticollis can be caused by trauma, infection, as well as congenital abnormality. Approximately 10%-20% of torticollis patients belong to those with post-traumatic category. One of the commonest cause of the trauma was strenuous delivery process, such as breech delivery, delivery using forceps, narrow pelvis, and large-sized newborn and pre-delivery process, such as intrauterine fixed head position (Holt 1940, Raffensperer 1990, Skinner 1991, Cheng et al. 2001, Azizkhan et al. 2003, Thompson 2004).

There is an interesting story about the case of this disease. Apparently, Alexander the Great once suffered this disease in the third century BC. Antyllus ever performed tenotomies in the fourth century AD to treat this disease. Surgical procedure to sternocleidomastoideus muscle was done for the first time in Amsterdam by Minnius in 1641. Then, Heusinger in 1826 provided for the first time clear elaboration on torticollis (Azizkhan et al. 2003).

CASE REPORT

A male child of 8 years old visited Dr Soetomo Hospital, Graha Amerta, in October 4, 2005, with major complaint that his head leaned to the right. The

complaint was found in the last 6 months through the information of his sport teacher. According to the patient, his head had long been leaned to the left, although it had been realized for the last 6 months. There are no complaints of pain or lump found in the neck. Head was moving automatically without needed or acquired. Urinating, defecating and walking were normal. The patient was the second child of three children. He was delivered per vaginam without forceps and cried spontaneously with the help of a midwife.

General physical and laboratory examination revealed normality, GCS 456, vital signs were normal. Bodyweight was 23 kg. Local status examination in the neck showed findings as follows: head leaned to the right and chin to the left. There was a protrusion of right Sternokleidomastoideus muscle. Pressure pain, tremor and tic as well as tumor mass in the neck were not found. There was also no disorder in masticating, swallowing, and speaking. The patient was diagnosed with torticollis dextra.



Figure 1. The patient, pre-operatively. Shortened dextral sternocleidomatoid muscle is apparent.

The patient was subjected to musculotomy and tenotomy of the right sternocleidimastoid muscle.

Incision was performed in right mastoidal area for 1 cm and right supraclavicular in the orygo of right sternocleidimastoid muscle as long as 2 cm. Incision was deepened layer by layer until the sternocleido-mastoid muscle (in mastoid incision) and right sternocleidomastoid tendon are exposed. Musculotomy and tenotomy of right sternocleidomastoid muscle was carried out. Operation field was closed later by layer. After operation, Philadelphia collar brace was installed and maintained to 6 weeks. The patient was trained to turn his head to the right and lean his head to the left. Skin suture was removed at day-7 postoperatively.



Figure 2. The patient, two days after operation, wearing Philadelphia collar brace



A. Trace of operation (right)



Figure 3. The patient, seven days after operation. The difference of right (abnormal) and left (normal) sternocleidomastoid muscle stretching

DISCUSSION

The etiopathogenesis of torticollis remains controversial until today, although congenital muscular torticollis is suggested to result from local trauma in soft tissue of the neck during the delivery process, while the etiopathogenesis of acquired muscular torticollis depends on the underlying cause of the primary abnormality. The type of torticollis causes can be divided into torticollis caused by skeletal process (malformation of skull dan cervical bone), non-osseous process (muscle and soft tissue), and neurogenic process (Holt 1940, Brenda 2002, Azizkhan et al. 2003).

Most of pediatric acquired torticollis cases result from inflammatory process of neck muscles with the primary focus originated from upper respiratory tract and the presence of minor injury in neck muscle. Other serious manifestations of acquired torticollis are exerted from fossa posterior tumor or spinal cord. In general, torticollis is caused by trauma, infection and congenital infection. In trauma cases, incidence of less than 20%-30% is reported in breech delivery. Other conditions are strenuous delivery due to narrow hips, large-sized infant, delivery using forceps, intrauterine abnormal fixed fetal position or perinatal compartment syndrome characterized with torticollis symptom, hip dysplasia, Scoliosis, Club foot, Talipes, and hypoplastic lung (Holt 1940, Oski et al. 1987, Raffensperer 1990, Skinner

1991, Azizkhan et al. 2003, Othee 2004, Thompson 2004).



A. Before Operation



B. After Operation

Figure 4. Frontal view of the patient, seven days after operation. The stretching of right sternocleidomastoid muscle has disappeared. Head looks straight. Compare the picture before and after operation.

In strenuous delivery process, a snare may occur, resulting in blood flow reduction and neck muscle damage. The sternocleidomastoid muscle become stretched, pulled and torn, resulting in bruise and hematoma. During the development, the normal side of the neck becomes longer, and the abnormal side will have less elastic nature. The sternocleidomastoid muscle becomes shortened in abnormal side, thereby producing the symptom. This theory is applied to most of birthway trauma, but does not explain the torticollis in infants born with section or familial cases. This was likely due to fetal abnormal position in the uterus. Roemer in 1954

proposed a postulate that after the head passing sacral promontory in lateral position, hyperextension may result in rupture of sternocleido-mastoid muscle. Torticollis is also caused by several circumstances, such as cervical hemivertebrae, adenitis, fasciitis and ocular muscle imbalance (Osiki et al. 1987, Skinner 1991, Azizkhan et al. 2003, Encyclopedia Index, Joseph J 2005, Thompson 2004).

Congenital muscular torticollis begins to appear during delivery or several days-weeks after delivery. At the time of delivery, the neonate looks healthy without any abnormalities found. In the second to eighth week of age, clinical symptoms become more apparent. Each child has different symptoms. The head leans towards the abnormal side, while the face and chin turn towards the normal side. The neck movement is varied, starting from soft to hard movement, and becomes severe during standing, walking, or under full psychological stress. These symptoms are often found in torticollis patients in a frequency of 80% (Holt 1940, Osiki et al. 1987, Azizkhan et al. 2003, Encyclopedia Index, Othee 2004, Ross 2005).

Acquired muscular torticollis occurs after delivery period, childhood, adolescence, adult, and middle ages. In previous examinations, the patient is found in normal condition during and after delivery. Actually, the occurring clinical symptoms are similar to congenital muscular torticollis type. The most frequent clinical symptom in children is stiffness and tenseness of the neck. In children, there is a sudden symptom that present as very severe pain and stiffness of neck muscle in one side. However, the most common symptom is only the neck stiffness. This symptom may alleviate by itself from 2-3 days to 1-2 weeks. Such condition can be treated with muscle relaxant, analgesic, the use of cervical collar, or physiotherapy with massage, and sufficient rest. The types of drug used are nonsteroidal anti-inflammatory drugs (NSAIDs), benzodiazepines and other muscle relaxants, anticholinergics, local intramuscular injection of botulinum toxin as well as phenol (Allison 2001, Encyclopedia Index, Othee 2004, Ross 2005).

There is a report about complete healing of torticollis without treatment until the first six months of age, approximately 50%-70%, and after the first twelve month of age, there is about 10% of complaints, such as shortened sternocleidomastoid muscle and the fibrotic mass will be fixed. However, the final result of this abnormality cannot be easily determined. Thereby, early stretching training should be given (Azizkhan et al. 2003, Brenda 2002, Cheng et al. 2001, Twee 2006). The therapies of choice for torticollis are physical therapy (stretching training), massage, local heat, analgesics,

sensory biofeedback, transcutaneous electrical nerve stimulation (TENS) and intramuscular BOTOX (Botulinum Toxin) injection. Stretching training is carried out several times a day for 3-6 months and evaluated (Holt 1940, Osiki et al. 1987, Raffensperer 1990, Skinner 1991, Azizkhan et al. 2003, Encyclopedia Index, Othee 2004, Theresa et al. 2004).

Operation of torticollis is indicated for lesion that irresponsive to conservative therapy modality and the presence of facial hemihypoplasia development. The surgical procedure comprises sternocleidomastoid release or lengthening, selective denervation and dorsal cord stimulation (Osiki et al. 1987, Skinner 1991, Azizkhan et al. 2003, Othee 2004). The operation is contraindicated if the underlying disease has not been explored and non-surgical therapy has not been given to the patient (Othee 2004).

Another method of congenital torticollis treatment is by using Botulinum toxin. Botulinum toxin comprises several serotypes (A, B C, D, E, F and G). The most popular one is the botulinum toxin type A. The intramuscular injection of Botulinum toxin can be used as the primary therapy as well as adjuvant therapy besides physiotherapy to support stretching training. Botulinum toxin is used to control muscle stretch by inhibiting acetylcholine release in neuromuscular junction. The presenting clinical effects may manifest as transient weakness and the atrophy of striated muscle. Botulinum toxin A is often used for congenital torticollis due to cerebral palsy. The action effect of botulinum toxin A is lasting for 1-4 months. Repeated injection is needed to maintain the drug's effect. In several patients there are reports about the occurrence of botulinum toxin A immunity that required an increased dose (Allison 2001, Francis 2003, Theresa et al. 2004, Joseph 2005, Costa et al. 2006).

The required dose is ranging between 100 - 200 unit per injection. The selection of muscle that will be relaxed should be done carefully to prevent relaxation of unnecessary muscle. The most common side effect is dysphagia, which generally can be tolerated by the patient. Pain in injection site may last for 4 weeks after injection. In general, injection to sternocleidomastoid muscle is divided to three sites, in orygo, insertion and medial area, each of which receives a third of the dose. Dose increase is needed when there is botulinum toxin resistance or autoimmune. The dose can be increased to 200 units per injection and further increase beyond 200 units will not provide more advantages. If there is immunity against botulinum toxin type A, botulinum toxin with other serotype can be used. However, studies using serotypes B, C, D, E, F and G for congenital muscular torticollis remain rare (Allison 2001, Francis

2003, Joseph 2005, Costa et al. 2006, Twee 2006). In this patient, botulinum toxin injection therapy was not given due to long distance of the patient's residence from health center, preventing him to undergo repeated injection process, to financial problems and to the late and unfeasible physiotherapy modality.

In general, serious complications are relatively not found in surgical procedure. The most frequent complication was hematoma in incision or bleeding site. Other reported complications are injury in pars spinalis accessory nerve; adjacent blood vessels, such as jugular veins and carotid artery; neck muscle atrophy, loss of muscle control, instability, variable numbness/sensory loss, pain and neck deformity. There is a report about second operation due to the attachment of sternocleidomastoid muscle to the fascia during wound healing process or detached muscle in the adjoining site (Azizkhan et al. 2003, Othee 2004).

Most of torticollis patients could undergo non-surgical therapy, and, with reliable follow-up, they could obtain maximum neck movement and healing from fibrotic mass. A complete remission is reported among 12%-21% of torticollis patients, particularly among young children. In congenital muscular torticollis patients, it is reported that about 90% of the patients showed good response in the first one year with non-surgical treatment. The best outcome will be obtained if surgical therapy is applied in one or two years, and followed subsequently with physiotherapy. Torticollis may lead to permanent facial deformity if treatment is not immediately given in the first one year. Recurrence rate in surgical therapy is less than 3% (Skinner 1991, Azizkhan et al. 2003, Encyclopedia Index, Othee 2004, Ross 2005,).

CONCLUSION

We reported a case of congenital muscular torticollis, characterized with shortening of the right sternocleidomastoid muscle, with head leaning to the right and turning to the left. The patient was treated with torticollis therapy with musculotomy and tenotomy of right sternocleidomastoid muscle.

REFERENCES

Allison, B 2001, 'The botulinum Toxin in the Treatment of Cervical Dystonia', *Seminars in Neurology*, vol. 21, no. 1.

- Azizkhan, RG et al. 2003, 'Head and Neck Lesion' in *Operative Paediatrics*, International Edition, vol. I, Mc Graw-Hill Companies, Inc. USA, pp. 232-234
- Brenda, LL 2002, 'Congenital muscular torticollis', *Orthopaedic Nursing*, vol. 21, no. 3, p. 21.
- Cheng, et al. 2001, 'Clinical determinants of the outcome of manual stretching in the treatment of congenital torticollis in infant', *Journal of Bone and Joint Surgery*, vol. 83-A, pp. 676-867.
- Costa, J et al. 2006, 'Botulinum toxin type A therapy for cervical dystonia (Review)', in *The Cochrane Library*, issue 3.
- Encyclopedia Index, *Torticollis*. Retrievable from <http://www.drhull.com/EncyMaster/T/Torticollis.html>
- Francis, OW 2003, 'Botulinum toxin therapy for cervical dystonia', *Phys Med Rehabil Clin N Am*, vol. 14, pp. 749-766.
- Holt, E 1940, 'Disease of the Muscle, Torticollis', in *Disease of Infancy and Childhood*, 11th edn, Appleton-Century Company Inc., New York, pp. 1065-066
- Joseph, J 2005, *An Update on Cervical Dystonia*, Department of Neurology, Baylor College of Medicine, Houston, Texas.
- Oski, FA et al. 1987, 'Bone, Joint and Muscle Problems, Torticollis' in *Principles and Practice of Pediatrics*. 2nd edn, JB Lippincott Company, Philadelphia, pp. 1162-1165
- Othee, GS 2004, *Torticollis*. Retrieved from <http://www.emedicine.com/Otrhoped/topic542.htm>
- Raffensperer, JG 1990, 'Soft Tissue tumors', in *Swenson's Pediatrics Surgery*, 5th edn, Appleton & Lange, Norwalk, Connecticut, pp. 438-439.
- Ross, MJ 2005, *Torticollis*. Retrieved from <http://www.emedicine.com/emerg/topic597.htm>
- Skinner, SR 1991, 'Orthopaedic Problem in Childhood, Congenital Muscular Torticollis' in *Rdolphs Pediatrics*, 19th edn, Appleton & Lange, Norwalk, p. 1927
- Theresa, AZ et al. 2004, 'Botulinum toxin A for the treatment of cervical dystonia', in Ashley Publication Ltd.
- Thompson, GH 2004, 'The Neck, Torticollis', in *Nelson Text Book of Pediatrics*, 17th edn, Saunders, pp. 2288-2289
- Twee, TD 2006, 'Congenital muscular torticollis: current concepts and review of treatment', *Curr Opin Pediatr*, vol. 18, pp. 26-29.