Case report

Neglected Congenital Muscular Torticollis Treated with Bipolar Release: A Case Series

Pierre Alexander[1], Jainal Arifin[2], Muhammad Ihsan Kitta[3], Rheza Setiawan Budiharto[1]

Authors' affiliations: [1]Resident, [2]Consultant, Dept. of Orthopaedic & Traumatology, Hasanuddin University, Makassar; [3]Consultant, Dept. of Orthopaedic & Traumatology, Muhammadiyah University, Makassar
Correspondence: Rheza Setiawan Budiharto Email: rhezsetiawan@gmail.com, Mob. No. +6281233383606

INTRODUCTION

Congenital muscular torticollis (CMT) is a thickening and/or tightness of the unilateral sternocleidomastoid muscle (SCM) characterized by fibrosis, resulting in shortening of the SCM and consequent limited neck motion.[1, 2] Torticollis in Latin means twisted and and in the 1912, Tubby first defined it as a "deformity, congenital or acquired, characterized by lateral inclincation of the head to the shoulder, with torsion of the neck and deviation of the face."[3]

Original written references to this deformity appeared in Plutarch's classic description of the Macedonian king Alexander the Great.[4] CMT itself had a reported incidence of 0.3-1.9% and various theories have been proposed while the true etiology of torticollis remains uncertain.[3]

The differential diagnoses of torticollis is extensive and is as shown in **Table 1**.[3]

Table 1. Differential diagnoses of torticollis

Congenital :
- Muscular
- Vertebral Anomalies; failure of formation, segmentation or both
- Ocular
Acquired :
- Tumoral; eosinophilic granuloma, osteoma/osteoblastoma
- Traumatic; C1 fracture
- Inflammatory; Juevnile rheumatoid arthritis, respiratory tract infection, cervical adenitis
- Hysterical
- Paroxysmal torticollis of infancy
- Associated with ligamentous laxity; Down syndrome

As such, a thorough investigation is sometimes needed to identify any accompanying disorder. Historically, the treatment of the CMT itself is primarily conservative and surgical release is need only in a small rate that conservative treatment fails, or in certain neglected cases. [5, 6]

Previous studies have demonstrated that the best outcome is obtained if the surgery is performed between the ages of 1 and 4 years and as such, neglected case is stated for those who did not undergo operative treatment within adequate time. [1]

We present a case series of neglected congenital muscular torticollis treated by bipolar SCM muscle release at the age of 12 years and 20 years followed by active physiotherapy and exercises. The lack of the previous treatment together with advanced age and marked deformity that significantly improved after surgery warrant the report of the outcome.

CASE 1

We report a case of a 12-year-old girl affected with neglected congenital muscular torticollis involving the left SCM (**Figure 1**). The patient was firstborn and had no family history for muscular torticolis. The diagnosis itself was made at the age of 12 when her parents were concerned about the possible progressivity of the condition. She had previously undergone physical therapy of active neck stretching exercise for several months but was not fruitful. Detailed medical history revealed a history of shoulder dystocia and significant manipulation was needed to deliver her right shoulder.

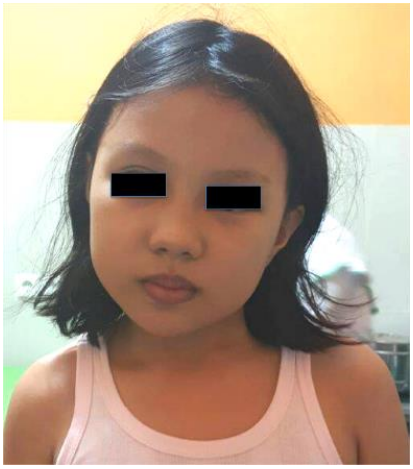

Figure 1. Preoperative frontal photograph (reproduced with the patient's permission)

Maxillofacial region examination revealed an inclined head to the left, raised chin, face rotated to the op-

posite direction. All other facial features and dental examinations were within normal limits. Range of motion of the cervical region (neck rotation and lateral flexion) was limited on one side with short, tight, firm, left SCM.

Preoperative cervical radiographs revealed no evidence of cervical vertebra abnormalities besides the increased cervicomandibular angle (CMA) as measured from AP plain radiographs of the cervical spine. The CMA was defined as the angle between the line along the upper border of C7 spine and the lines connecting the lower borders of both mandibles. The preoperative CMA value was 30°.

Figure 2. Postoperative clinical photograph showing excellent cosmesis and improved range of motion at 4-months follow-up (reproduced with the patient's permission)

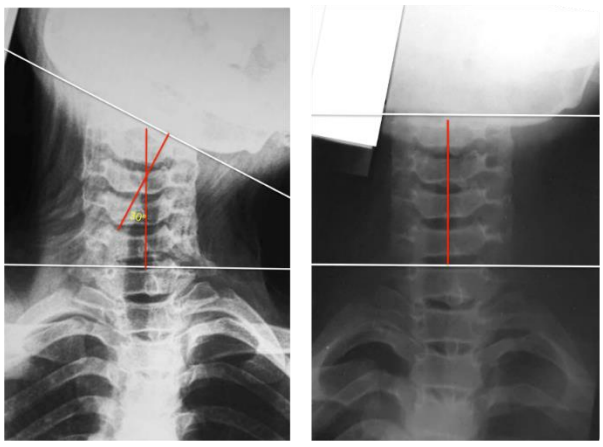

Fig 3. Radiographic evaluation during the preoperative and 4-months postoperative period

We then formulated a treatment plan of bipolar surgical release of the left SCM followed by postoperative utilization of a cervical brace, aggressive physical therapy along with postural exercises.

A short transverse proximal incision behind the ear was made and the SCM insertion was divided just distal to the tip of the mastoid process. Another distal incision of 4-5 cm long was made a fingerbreadth proximal to the medial end of the clavicle and sternal notch after which the clavicular portion of the muscle was cut transversely. The subcutaneous tissue and skin were closed primarily in two layers and the patient was extubated uneventfully.

The patient was placed on a non-rigid cervical brace during the first week postoperative period and aggressive physiotherapy which include neck strengthening and extension exercises was started early at second postoperative day. At 4-month-postoperative follow-up, patient was able to extend the neck and perform rotation to the opposite site and there was only a moderate amount of scar tissue formed at the surgical site. Final radiographic examination also reveal excellent postoperative CMA value of 0°. Functional and cosmetic result was excellent with improved cervical range of motion and centered head position (**Figure 2**). (**Figure 3**).

CASE 2

We report a case of an 20-year-old girl affected with neglected congenital muscular torticollis involving the left SCM (**Figure 4**). The patient was second child of 3 siblings and had no family history for muscular torticolis. Her condition was recognized by her parents since she was 4 years old, but they did not seek any medical assistance. But now, she were concerned about the possible progressivity of the condition. She had previously undergone physical therapy of active neck stretching exercise for several years but was not fruitful.

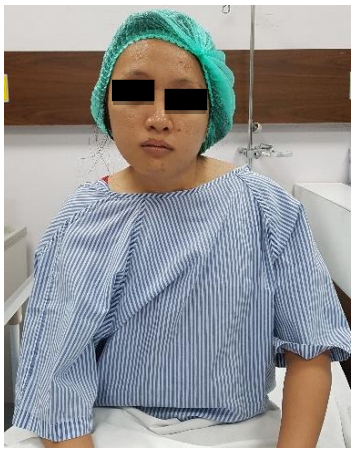

Figure 4. Preoperative frontal photograph (reproduced with the patient's permission)

Maxillofacial region examination revealed an inclined head to the left, raised chin, face rotated to the opposite direction. All other facial features and dental examinations were within normal limits. Range of motion of the cervical region (neck rotation and lateral flexion) was limited on one side with short, tight, firm, left SCM.

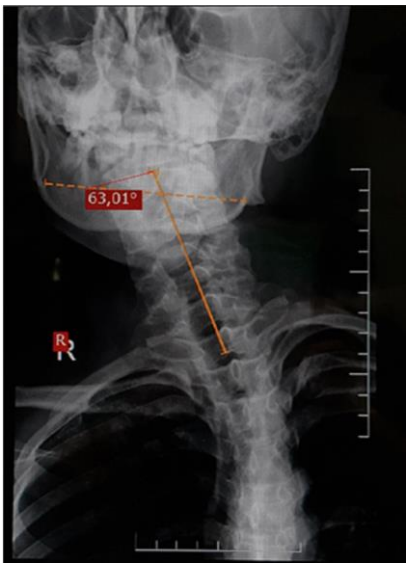

Figure 5. Radiographic evaluation during the preoperative

Figure 6. Postoperative clinical photograph showing excellent cosmesis and improved range of motion at 2-months follow-up (reproduced with the patient's permission

Preoperative cervical radiographs revealed no evidence of cervical vertebra abnormalities besides the increased cervicomandibular angle (CMA) as measured from AP plain radiographs of the cervical spine. The preoperative CMA value was 63°.(**Figure 5**).

We then formulated a treatment plan of bipolar surgical release of the left SCM followed by postoperative utilization of a cervical brace, aggressive physical therapy along with postural exercises.

With patient in a supine position, maximum tension of the affected muscle was achieved by placing the neck in a hyperextended position along with rotation of the head to the opposite direction. A short transverse proximal incision behind the ear was made and the SCM insertion was divided just distal to the tip of the mastoid process. Another distal incision of 4-5 cm long was made a fingerbreadth proximal to the medial end of the clavicle and sternal notch after which the clavicular portion of the muscle was cut transversely. The subcutaneous tissue and skin were closed primarily in two layers and the patient was extubated uneventfully.

The patient was placed on a non-rigid cervical brace during the first week postoperative period and aggressive physiotherapy which include neck strengthening and extension exercises was started early at second postoperative day. At 2-month-postoperative follow-up, patient was able to extend the neck and perform rotation to the opposite site and there was only a moderate amount of scar tissue formed at the surgical site. Final radiographic examination also reveal excellent postoperative CMA value of 10°. Functional and cosmetic result was excellent with improved cervical range of motion and centered head position (**Figure 6**).

DISCUSSION

Muscular torticollis is the end result of shortening of the SCM muscle resulting in limitation of neck motion. [1, 2]

A male to female predominance of 3:2 has been reported and it is generally more common on the right side. The CMT case we found from a female patient was left-sided. Muscle involvement may be diffuse, but more often it is localized near the clavicular attachment of the muscle. [2]

CMT itself is often associated with other congenital deformities such as Developmental Dysplasia of the Hip (DDH) with a coexistence rate estimated as high as 14.9%. We however did not find any other congenital deformities in our patient. Other coincident lesions less frequently recorded include cervical scoliosis, tibial torsion, clubfoot, calcaneovalgus foot,

flexible pes planus, metatarsus adductus, and hallux valgus. [1, 2, 5, 7]

In 1969, the CMT was further divided into three groups by MacDonald. The first group is the SCM tumor group (42.7%) which is characterized by a palpable mass that is hard and movable within the substance of the SCM. The second group is the muscular torticollis group (35.2%) and it consists of those with tightness of SCM but without clinical "tumor." The third group is the postural torticollis (POST) group (22.1%) which clinical features of CMT but with no demonstrable tightness nor tumor of the SCM. [6, 8]

CMT has an unclear etiology although it is postulated that fetal position abnormalities, intrauterine or perinatal compartment syndrome and birth trauma ensuing a difficulty delivery embody the main causes. There was a previous history of shoulder dystosia during the childbirth period which may initiate the pathologic changes of the involved SCM. Other causes may include hereditary and venous or arterial occlusion which may create fibrous tissue within the SCM. Other findings in the muscle are presence of muscle giant cells, loss of transverse striations, vacuolization, and disruption of endomysial sheaths. [2, 4, 5, 6, 7]

Contrary to past view that the craniofacial asymmetry is the result of either positional moulding of open cranial sutures arising from the tilt of the head, or deformation from the pull of the shortened muscle, it has been suggested recently that it is this fascial contraction that is responsible for craniofacial distortion. [4]

The diagnosis of the condition itself is mainly based on past medical history and clinical examination of the patient although several objective masurement methods have been proposed such as the cervicomandibular angle, lateral shift of the head and cobb angle of the involved spine segment. [1, 5, 6]

The main approach to the condition remains a trial of conservative treatment consisting of stretching maneuvers, although surgical release of the affected SCM is recommended for resistant cases. [1, 7]

Manual stretching is most effective if performed before the age of 12 months. The technique involves placing one hand on the child's head and the ipsilateral shoulder, while with the other places the head on a lateral flexion together with rotation towards the opposite side. At lease two times a day, 10-15 stretches are performed. This stretching technique can also be combined with Botulinum toxin A injections. [2, 5, 9]

Botulinom toxin A injections have also been used by some authors to decrease spasticity of the involved muscle hence enabling the manual stretching. [5, 9]

Surgery itself is highly recommended when a severe restriction of movement is present, as well as in cases complicated with deformities of facial bones. We had opt to perform surgery in our patient as a trial of nonoperative measures had previously failed and restoration of functional and cosmetic became more challenging. Parameters such as residual head tilt, scar formation, craniofacial asymmetry and age at the time of surgery play an important role in the outcome after surgery.

A potential complication of the surgical approach is an injury of the accessory nerve with the rate of relapse of up to 1.2%. [5, 7, 10]

Although there are various surgical procedures for CMT, unipolar and bipolar release are the most popular. Eventhough it was reported that bipolar release combined with Z-plasty can preserve the normal v-contour of the SCM in the neckline, several authors have reported that no loss of normal v-contour was seen in bipolar releases without Z-plasty as seen in our patient. [3]

Bipolar release remains a very viable option for correction of neglected and relapsed congenital muscular torticollis although after the age of five, the form and efficacy of treatment are controversial. [3, 9, 11,12]

Regardless of the surgical procedure, a postoperative regimen of intensive physiotherapy including passive range of motion and active strengthening exercises for at least 3 months is of utmost importance to maintain the effects of surgery. [7, 8]

Without adequate treatment, the limitation in neck range of motion may lead to complications such as pain, spinal deformites and craniofacial abnormalities. [1, 2]

CONCLUSION

There has not been a clear consensus regarding which surgical technique provides the best chance at restoring a near normal function and cosmesis for neglected cases of CMT. Bipolar release is still a viable option and the role of well planned physiotherapy cannot be underestimed in the treatment plan. As some deformational change is most resistant to remodelling after puberty, early recognition and treatment of this condition is most likely beneficial.

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