

NEGLECTED CONGENITAL MUSCULAR TORTICOLLIS TREATED WITH BIPOLAR RELEASE: A CASE REPORT AND OVERVIEW

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ABSTRAK

Introduction Torticollis a symptom of cervical spine abnormality. The most common form of congenital painless torticollis is congenital muscular torticollis (CMT), or wry neck. This is a case of neglected congenital muscular torticollis treated by bipolar sternocleidomastoid (SCM) muscle release at the age of 12 years followed by active physiotherapy and exercises. **Case report** A 12-year-old girl affected with neglected congenital muscular torticollis involving the left SCM with no evidence of cervical vertebra abnormalities besides the increased cervicomandibular angle. A treatment plan of bipolar surgical release of the left SCM was formulated, followed by postoperative utilization of a cervical brace, aggressive physical therapy along with postural exercises. Functional and cosmetic result was excellent with improved cervical range of motion and centered head position. At 4 months postoperative, 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. Radiographic examination also revealed excellent postoperative CMA value. **Discussion** CMT has an unclear aetiology although it is postulated that foetal position abnormalities, intrauterine or perinatal compartment syndrome and birth trauma ensuing a difficulty delivery embody the main causes. The main approach to the condition remains a trial of conservative treatment consisting of stretching manoeuvres, although surgical release of the affected SCM is recommended for resistant cases. Manual stretching is most effective if performed before the age of 12 months. This stretching technique can also be combined with Botulinum toxin A injections. Surgery itself is highly recommended when a restriction of movement up to thirty degrees is present, as well in cases complicated with deformities of facial bones. **Conclusion** No clear consensus regarding which surgical technique provides the best chance at restoring a near normal function and cosmesis for neglected cases of CMT.

Keywords : torticollis, congenital torticollis, muscular torticollis

INTRODUCTION

Torticollis (from the Latin meaning “twisted neck”) is a symptom of cervical spine abnormality. The most common form of congenital painless torticollis is congenital muscular torticollis (CMT), or wry neck. The condition itself is a thickening and/or tightness of the unilateral sternocleidomastoid muscle (SCM) characterized by fibrosis, result in shortening of the SCM and consequent limited neck motion. The child typically presents with head tilted toward the involved fibrotic SCM muscle, and the chin is rotated toward the contralateral shoulder, thus producing the “cock robin” appearance

(Kyung-Jay Min et al, 2016; Anil & Indreshwar, 2011; Herring, 2014).

CMT itself had a reported incidence of 0.3-1.9% and various theories have been proposed while the true aetiology of torticollis remains uncertain. The differential diagnoses of torticollis are extensive and can be simplified by determining whether the deformity was present at birth (congenital) or was acquired and whether the deformity is painful or nonpainful. The complete list is as shown in Table I (Farzad,2008; Herring, 2014).

Table 1. Differential diagnoses of torticollis

Congenital – Nonpainful	Acquired – Painful	Acquired – Painful or Nonpainful
<ul style="list-style-type: none"> • Congenital muscular torticollis • Cerebral anomalies • Failure of segmentation <ul style="list-style-type: none"> - Klippel-Feil syndrome - Occipitalization of C1 • Failure of formation <ul style="list-style-type: none"> - Congenital hemiatlas • Combined failure of segmentation and formation • Ocular torticollis 	<ul style="list-style-type: none"> • Traumatic <ul style="list-style-type: none"> - Atlantoaxial rotatory displacement - Os odontoideum - C1 fracture • Inflammatory torticollis <ul style="list-style-type: none"> - Atlantoaxial rotatory displacement (Grisel syndrome) - Juvenile rheumatoid arthritis - Diskitis or osteomyelitis - Other infection in neck • Tumors <ul style="list-style-type: none"> - Eosinophilic granuloma - Osteoid osteoma or osteoblastoma • Calcified cervical disk • Sandifer syndrome 	<ul style="list-style-type: none"> • Paroxysmal torticollis of infancy • Tumors of the central nervous system <ul style="list-style-type: none"> - Posterior fossa - Cervical spinal cord - Acoustic neuroma • Syringomyelia • Hysterical torticollis • Oculogyric crisis (phenothiazine toxicity) • Associated with ligamentous laxity <ul style="list-style-type: none"> - Down syndrome - Spondyloepiphyseal dysplasia or mucopolysaccharidosis

Historically, the treatment of the CMT itself is primarily conservative and surgical release is needed only in a small rate when conservative treatment fails, or in certain neglected cases. Previous studies have demonstrated that the best outcome was 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 (Kyung-Jay Min et al, 2016; Antonios et al, 2013; Jaiswal et al,2005).

We present a case of neglected congenital muscular torticollis treated by bipolar SCM muscle release at the age of 12 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 REPORT

We reported a case of a 12-year-old girl affected with neglected congenital muscular torticollis involving the left SCM (Fig. 1). The patient was firstborn and had no positive family history for muscular torticollis. The diagnosis itself had been 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.



Fig 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 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.

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°.

We then formulate 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.

Functional and cosmetic result was excellent with improved cervical range of motion and centered head position (Fig. 2). Aggressive physiotherapy which include neck strengthening and extension exercises was started early. At 4 months postoperatively 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. Radiographic examination also reveal excellent postoperative CMA value of 0° (Fig. 3). 12 months follow up after surgery was showing an excellent result with minimal scar tissue at the surgical site.



Fig. 2. Postoperative clinical photograph showing excellent cosmesis and improved

range of motion at 4-months follow-up (reproduced with the patient's permission)

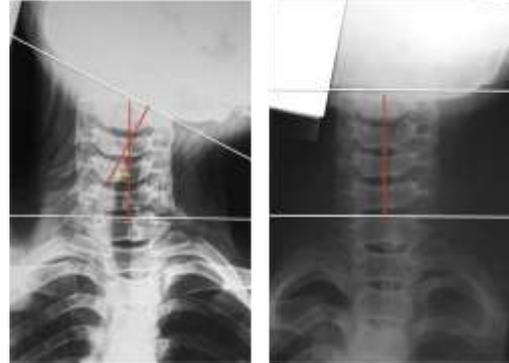


Fig 3. Radiographic evaluation during the preoperative and 4-months postoperative period.



Fig. 4. Postoperative clinical photograph showing excellent cosmesis at 12-months follow-up (reproduced with the patient's permission)

DISCUSSION

Muscular torticollis is the end result of shortening of the SCM muscle resulting in limitation of neck motion. A male to female predominance of 3:2 has been reported and it is generally more common on the right side. Muscle involvement may be diffuse, but more often it is localized near the clavicular attachment of the muscle (Kyung-Jay Min et al, 2016; Anil & Indreshwar, 2011).

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%. Other coincident lesions less frequently recorded include cervical scoliosis, tibial torsion, clubfoot,

calcaneovalgus foot, flexible pes planus, metatarsus adductus, and hallux valgus (Kyung-Jay Min et al, 2016; Cheng et al, 2000; Anil & Indreshwar, 2011).

In 1969, the CMT was further divided into three groups by MacDonald. The first group is the SCM tumour 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 "tumour." The third group is the postural torticollis (POST) group (22.1%) which clinical features of CMT but with no demonstrable tightness nor tumour of the SCM (Ta & Krishnan, 2012; Jaiswal et al, 2005).

CMT has an unclear aetiology although it is postulated that foetal position abnormalities, intrauterine or perinatal compartment syndrome and birth trauma ensuing a difficulty delivery embody the main causes. 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. 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 (Antonios et al, 2013; Jaiswal et al, 2005; Anil & Indreshwar, 2011).

The clinical presentation usually varies from a simple head tilt with slight rotation and minimal restriction of motion to a more severe plagiocephaly, which may be exacerbated by the position of infant during sleep. Flattening of the face on the ipsilateral side of the lesion can be worsened by prone

sleeping position. If the infant was placed supine while sleeping, reverse modelling of the contralateral side of the skull can occur. The diagnosis of the condition itself is mainly based on past medical history and clinical examination of the patient although several objective measurement methods have been proposed such as the cervicomandibular angle, lateral shift of the head and Cobb angle of the involved spine segment (Herring, 2014).

The main approach to the condition remains a trial of conservative treatment consisting of stretching manoeuvres, although surgical release of the affected SCM is recommended for resistant cases. 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 least two times a day, 10-15 stretches are performed. This stretching technique can also be combined with Botulinum toxin A injections. Botulinum toxin A injections have also been used by some authors to decrease spasticity of the involved muscle, hence enabling the manual stretching (Do T.T, 2006).

Surgery itself is highly recommended when a restriction of movement up to thirty degrees is present, as well in cases complicated with deformities of facial bones. 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%^{5,8,10}. (Patwardhan et al, 2011).

Although there are various surgical procedures for CMT, unipolar and bipolar release are the most popular. Even though 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. 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 (Chang et al, 2013).

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. Without adequate treatment, the limitation in neck range of motion may lead to complications such as pain, spinal deformities and craniofacial abnormalities (Anil & Indreshwar, 2011).

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 underestimated 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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