

Congenital Muscular Torticollis – Case Series & Review of Literature

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Abstract: The term torticollis means “twisted neck” is defined as a congenital or acquired deformity, characterised by lateral inclination of the head to the shoulder, with torsion of the neck and deviation of the face. If conservative treatment is started early, with a regimen of passive stretching exercises and active strengthening of the contralateral muscle, about 95% of patients achieve an acceptable range of neck movement. The aim of treatment is to provide a long-term, cosmetic and functional restoration of neck mobility while minimizing the development of craniofacial deformity and upper cervical scoliosis. The surgical management of patients who do not respond to physiotherapy remains controversial. This paper provides a review of literature and highlights treatment protocol we follow in our patients with Congenital Muscular Torticollis and therapy achieved good esthetic and functional outcome.

Keywords: Congenital Muscular Torticollis , Unipolar & Bipolar release of SCM , Physiotherapy

I. Introduction

The term torticollis means “twisted neck” comes from Latin roots ‘tortus’ meaning twisted and ‘collum’ meaning neck.¹ It is defined as a congenital or acquired deformity, characterised by lateral inclination of the head to the shoulder, with torsion of the neck and deviation of the face.² Plutarch’s written references were one of the first to describe the special characteristics of emperor Alexander the great , the King of Macedonia who is believed to have this deformity.³ The differential diagnosis is extensive (Table 1), with muscular torticollis being the most common form.⁴ Muscular torticollis results from shortening or excessive contraction of the sternocleidomastoid (SCM) muscle.¹

The SCM muscle comprises of four distinct anatomical elements, each with a different action. This complexity is reflected in the clinical features of torticollis, which in part depend on the site of the disease within the muscle.⁵ The characteristic signs of torticollis include orbital dystopia (the eye on the unaffected side is higher up), deviation of the chin to the affected side, considerable restriction in rotation of the neck, associated with a tight sternocleidomastoid muscle, tilt of the head, and dental malocclusion (the lower mid line is deviated to the affected side, causing a unilateral crossbite, postnormal Class II molar relation on the affected side, and prenatal Class III molar occlusion on the unaffected side).⁶ The basic abnormality of Congenital Muscular Torticollis (CMT) is endomysial fibrosis with deposition of collagen and the aggregation of fibroblasts around individual muscle fibres, which then undergo atrophy.¹ The diagnosis of CMT is based on the clinical palpation of a firm mass or fibrous band within SCM muscle.¹

Table 1 – Differential diagnosis of torticollis

- A. Non-osseous:
 - Muscular
 - Ocular (muscle palsy)
 - Neurogenic tumours:
 - Cerebellar
 - Spinal cord
 - Syndromal: Sandifer syndrome (torticollis and gastro-oesophageal reflux)
 - Spasmodic
 - Neurological (brachial plexus injury)
- B. Osseous:
 - Congenital cervical spine malformations:
 - Occipitocervical invagination,
 - Atlas malformation
 - Klippel–Feil syndrome
 - Rotatory fixation (C1–C2):
 - Trauma
 - Respiratory tract infection
 - Cervical adenitis

II. Case reports:

In this case reviews , we report 5 patients with diagnosis of congenital muscular torticollis . 4 out of these patients were adults between age group of 18-25 years and one belonged to pediatric age group (8 years). All 5 patients were female. 4 out of these patients had CMT involving the right SCM and one patient had CMT involving the left SCM. Two of these patient’s had more advanced form of CMT where the entire length of SCM was cord like.

No.	Age of pt.(years)	Gender	Side involving	H/o trauma	Type of surgery	H/o previous physiotherapy
Patient 1.	22	F	Left	Present	Bipolar	Absent
Patient 2.	18	F	Right	Absent	Unipolar	Present
Patient 3.	20	F	Right	Absent	Unipolar	Absent
Patient 4.	08	F	Right	Absent	Unipolar	Absent
Patient 5.	24	F	Right	Present	Bipolar	Present

None of these patients had h/o any abnormal obstretic presentation at birth , any infection or previous head & neck surgery. There were no other congenital abnormality noted in any of these patients. All patients underwent a ophthalmological , ENT & orthopaedic evaluation to rule out any gross deformity.Cervical & Chest radiograph of 22 year old (patient 1) who had CMT involving the left side revealed degenerative changes in the cervical spine. Cervical & chest radiograph of patient 2 revealed cervical & thoracic scoliosis (S-shaped).

On extraoral examination , all patients had disproportionate physique due to raised shoulder & shortened neck on the involved side. Head was tilted towards the side of torticollis , raised chin , face rotated to opposite / normal side. Facial asymmetry was present in all patients except the child due to cranio – orbito zygomatic flattening on the involved side. Two patients with advanced CMT had cord like SCM with pronounced plagiocephaly on the involved side.

Extent of craniofacial deformity was less in the 8 year old patient compared to adults. This could be attributed to the fact that the growth is not complete & hence structures are continuously remodelling & adapting. All the 5 patient’s underwent surgical excision of the fibrous band under general anaesthesia followed by early aggressive physiotherapy in the post operative period.

Surgical Procedure:

1) Unipolar release:

Patient was placed in the supine position . Endotracheal intubation was difficult because of stiffness and rigidity of neck. Head was rotated towards the uninvolved side and neck was hyperextended to achieve maximum stretch and tension of the affected SCM muscle. Transverse incision of 1 inch length was placed in the supraclavicular region. The subcutaneous tissue and platysma were divided to expose the sternoclavicular head of SCM. Sternal head & clavicular head were palpated for thick fibrous band. Blunt dissection was carried out around the insertion of SCM. Muscular fibrous band was detached from the clavicular end without disturbing the articular portion of the sternoclavicular joint. Transverse cervical artery spotted under the muscle was preserved. Betadine gauge placed inside the wound.

2) Bipolar release:

Two of the patients with advanced CMT underwent bipolar release. After the completion of excision of fibrous band at the sternocleido attachment excision of the mastoid attachment was carried out. Vertical incision was placed in the mastoid region. Blunt dissection was done to expose the elongated mastoid process. Mastoid process was found elongated due to muscle pull. Layered dissection was done to expose the SCM muscle. Few muscle fibres were released from the mastoid process. Total release of all muscle fibres was not carried out in view of disfigured anatomy and to prevent injury to facial nerve.

Elongation of neck obtained and full range of free neck movements were achieved intraoperatively in all the patients. Subcutaneous layer and skin were closed primarily in layers . Physiotherapy was started from the second post – operative day which included neck positioning , strengthening and extension exercises. All patients had a tendency to place the neck in the pre operative position hence , cervical collar was placed to keep the neck in normal anatomic position. A custom made orthopaedic appliance was delivered to patient 1. The patient 1 wore this appliance upto 6 months post operative period. This appliance along with physiotherapy helped the patient to gain normal anatomic position of the neck with full range of movements. At 2 months postoperative follow up patient were able to move their neck to the opposite side.



Pre operative picture of Right side CMT



Post operative picture

III. Discussion

The aftermath of torticollis is shortening or excessive contraction of SCM muscle.² Incidence ranges from 0.3% to 2% and it is more commonly seen in male patients with male to female ratio of 3:2.¹ Right side of SCM muscle is most frequently involved than the left side and there is no sex predilection.⁷ Various theories have been proposed as a etiology but exact etiology is still not known.^{1,5,7} Some authors believe the hypothesis that there is a hematoma formation in the SCM from an intrauterine vascular disturbance, intrauterine malposition of the head, or due to compartment syndrome where the SCM shortens.⁸ Birth injury, infections, and hereditary theories have also been discussed.⁸ The diagnosis of congenital muscular torticollis is based on clinical palpation of firm fibrous band within the SCM muscle.¹ Ultrasonography is the imaging modality of choice for the evaluation of Congenital muscular torticollis, which must be clearly differentiated from other congenital and acquired types of torticollis, such as congenital cervical vertebral anomalies, post-traumatic conditions, infections and inflammations of adjacent structures, tumours, ocular torticollis, hearing deficit and miscellaneous neurological structural and functional causes.¹ Initial management of muscular torticollis in an infant is always conservative, and consists of exercises conducted by parents and physiotherapists two to four times per day. At four months of age the infant would undergo a re-evaluation.⁸ If the infant's head tilt is equal to or greater than six degrees, then a cervical, tubular brace/collar is used⁸ during the infant's waking hours. Some authors also recommend Botulinium Toxin as an adjunctive therapy in infants who fail to respond to 3 months of physiotherapy. Surgical intervention may be recommended if this approach does not resolve the torticollis within one year.⁸

Two types of surgical procedures may be performed. The first is a surgical division of the SCM muscle and the other is the surgical resection of the spinal accessory nerve and/or the anterior and posterior divisions of the first three cervical motor roots.⁸ The surgical options for the affected sternocleidomastoid muscle include unipolar release at the sternoclavicular origin, bipolar open tenotomy bipolar release, transection of the middle of the muscle, Z-plasties on the attachments of the sternal muscle, and complete excision of the muscle.⁶

Many authors have reported subjective improvement with varying degrees of success, in both facial asymmetry and function after the release of the affected thick fibrous SCM depending on the age of the patient at the time of operation.⁶ Some have suggested that the best results are when the operation is done either before the age of 1 or up to the age of 7 although it has been reported that good results can be achieved as late as the age of 12.⁶ However, the results are worst when the operation is done after puberty, because of the loss of potential for growth and remodelling.⁶ Two of our patients (patient 1 and 5) with advanced CMT showed

obvious facial deformity after the surgery although full range of neck movements & function were restored. This was also the reason why patient 4 was operated at such an early age of 8 years to avoid residual facial deformity at a later stage.

Daily, non-invasive approaches performed by the parents may contribute to the success of intervention such as feeding the infant in the prone position on both the right and left sides while the patient is awake.⁸ In another study of 452 patients with CMT with pseudotumor, 8% of patients receiving manual stretching of the SCM experienced a sudden giving-way or snapping of the SCM.⁹ In a study by Hyman, she discusses a case of a three month old child in whom a marked reduction of CMT and facial asymmetry was achieved after 6 weeks of chiropractic care. These results were attributed to the correction of vertebral subluxation.¹⁰

In another case report by Moore, she focuses on a pediatric torticollis case secondary to a fall. The four year old boy who fell head first off a bed, presented with left lateral head tilt and mild right head rotation. This position is typical for an atlantoaxial rotator fixation where usually the presenting sign is torticollis. The patient was unable to move his head around properly without pain and muscle guarding. Light massage and trigger point therapy were performed on the paraspinal musculature followed by a Diversified spinal adjustment to C3/4. Two weeks after the treatment, the patient had no clinical signs or symptoms of the fixation or torticollis.¹¹ In a report of three cases of acute cervical torticollis in adults Bolton describes the identification of upper cervical subluxation in all three cases. All cases were managed utilizing Palmer Upper Cervical Specific technique and all three cases resolved.¹²

Guttman describes a syndrome involving subluxation resulting in a blocked Atlantal nerve in infants and small children. He describes a clinical picture of central motor impairment, impairments of vegetative regulatory functions, and lowered resistance to infections. He reviews three case studies of children with the diagnosis including one with torticollis that resolved following two adjustments to reduce upper cervical subluxation.¹³

In a study Jae lee and authors recommended that the operative technique involving complete fibrous band release and resection, combined with intensive postoperative physical therapy and application of a soft neck collar, provides good functional and cosmetic results.¹ Results obtained by following this protocol has provided our patients with good functional & esthetic outcome.

In a paper stassen describe a technique that combines subperiosteal lengthening of the sternocleidomastoid muscle at its mastoid insertion, and division of lower fibrotic bands with minimal postoperative fibrosis. As the sternomastoid muscle is reattached lower down on the mastoid process, the lengthening of the muscle is stable, because the tendency to fibrosis and shortening is minimized. Comparison of the results with previous series shows that this technique provides immediate benefit and good long-term results.⁵

In a paper sanjeev dutta describes a new surgical technique for the treatment of torticollis. Transaxillary subcutaneous endoscopy offers a simple and effective way to surgically manage persistent torticollis while avoiding the potential for poor cosmesis from neck Scars.¹⁴

In a paper Savas Demirbelik et al treated fifty-seven infants and children for sternomastoid tumors and CMT. They treated patients conservatively with passive and active stretching exercises (PSE and ASE). It was concluded that PSE and ASE are highly effective for the treatment of congenital muscular torticollis. The success rate of conservative treatment is primarily dependent on the patients' age at the initiation of exercises.¹⁵

In a paper Yong – Taek Lee et al concluded that risk factors for intrauterine constraint appear to be associated with ultrasonographically detected severe fibrosis involving the entire SCM muscle in early presenting CMT.¹⁶

In paper Bharatendu Swain described a new technique for surgical correction of CMT by transaxillary endoscopic release of restricting bands. He concluded that this technique provides direct and quick access, perpendicular to the line of the fibrotic bands, avoids injury to neurovascular structures and does not leave visible neck scars.¹⁷

IV. Conclusion:

The functional & esthetic problems on the face & neck can have a lifelong impact on one's self esteem. It is in the patient's best interest to release SCM completely.

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