Congenital Muscular Torticollis-Case Report and an Effective Treatment Plan

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Abstract Congenital muscular torticollis results from shortening or excessive contraction of the sternocleidomastoid (SCM) muscle. The reported incidence varies between 0.4 and 1.9%. Various theories have been proposed, but its true aetiology remains obscure. The deformity is characterized by a practically painless, contracted cordlike SCM muscle, which pulls the head toward the side affected, narrows and draws the shoulder upward, forcing the chin in the opposite direction. Torticollis of congenital origin is a deformity not commonly met with in the practice of maxillofacial surgery. The rarity, lack of, or inadequacy of the previous treatment, together with the advanced age and marked deformity appear to warrant an investigation and report of the outcome. A case of congenital muscular torticollis is presented who reported at the age of 18 years and has been successfully treated by unipolar SCM release.

Introduction

The word torticollis comes from a Latin word which means twisted neck [1]. By definition it is a deformity, congenital or acquired, characterised by lateral inclination of the head to the shoulder, with torsion of the neck and deviation of the face [2]. The differential diagnosis is extensive (Table 1), with muscular torticollis being most common form [3]. Muscular torticollis results from shortening or excessive contraction of the sternocleidomastoid (SCM) muscle. Original written references to this deformity

appeared in Plutarch's classic description of the Macedonian king Alexander the Great [4, 5]. Subsequent to this historical mention, this rare deformity has become a better defined entity from histological studies and magnetic resonance imaging (MRI), showing muscle atrophy and interstitial fibrosis [6].

Torticollis being the cradle of birth is a deformity not commonly met with in the practice of maxillofacial surgery. A case of congenital muscular torticollis is presented who reported at the age of 18 years and has been successfully treated by unipolar SCM muscle release followed by use of readymade cervical brace, active physiotherapy and postural exercises. The lack of, or inadequacy of the previous treatment, together with the advanced age and marked deformity appear to warrant an investigation and report of the outcome.

Case Report

The patient affected with congenital muscular torticollis was an 18 year old adult female (Fig. 1) involving the right SCM muscle. The patient was first-born child with no positive family history for muscular torticollis. Congenital muscular torticollis was diagnosed in the first year of life. She had undergone physical therapy of active neck stretching exercise at the age of three as part of the rehabilitation process in Mumbai, India. Detailed medical history revealed no events of abnormal obstetric presentation during birth or episodes of gross trauma, unusual or prolonged infection, prior head and neck surgery, or associated pain in the neck. No other congenital abnormality existed.

Patient was moderately built with disproportionate physique especially due to raised shoulder and shortened neck on the right side. Maxillofacial region showed

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Table 1 Differential diagnosis of torticollis

Non-osseous:

Muscular

Ocular (muscle palsy)

Neurogenic tumours:

Cerebellar

Spinal cord

Syndromal:

Sandifer syndrome (torticollis and gastro-oesophageal reflux)

Spasmodic

Neurological (brachial plexus injury)

Osseous:

Congenital cervical spine malformations:

Occipitocervical invagination,

Atlas malformation

Klippel–Feil syndrome

Rotatory fixation (C1–C2):

Trauma

Respiratory tract infection

Cervical adenitis

Fig. 1 Preoperative frontal photograph of the wry-necked patient (reproduced with the patient's permission)

inclination of head to right side, raised chin, face rotated to the opposite direction of tilt and facial asymmetry (plagiocephaly) due to right cranio-orbito-zygomatic flattening and deviated nasal septum to the right. Her dental occlusion revealed slight shift in the dental lower midline to the right with angles class I molar relation bilaterally. Range of motion of the cervical region (neck rotation and lateral flexion) was deficit along with short, tight, firm and fibrous nontender right SCM muscle.

Ophthalmological and ENT evaluation including visual field tracking and response to sound was checked pre-operatively to rule out gross ophthalmologic and auditory

causes. Additionally, a complete neurologic examination revealed a normal study.

Pre-operative cervical and chest radiographs revealed cervical and thoracic scoliosis (S-shaped) with no evidence of maldevelopment or subluxation of the cervical vertebrae.

To this end we found our way of treatment plan by unipolar surgical release of the right SCM muscle and postoperative use of a cervical brace, aggressive physical therapy and postural exercises.

With the patient in a supine position, maximum stretch and tension of the affected SCM muscle was achieved by hyperextension of the neck and rotation of the head to the opposite shoulder. A simple technique of ipsilateral sternomastoid release at its inferior insertion was done by placing a transverse incision 1.5 cms superior to the right clavicle. The subcutaneous tissue and platysma were divided to expose the thick fibrotic cord-like SCM. Blunt dissection was carried out around the insertion of SCM. Division of the clavicular attachment of the muscle was done (Fig. 2) without disturbing the articular portion of the

Fig. 2 The taut band (a) was released in full at its clavicular attachment (b)

Fig. 3 Three months postoperative frontal photograph (reproduced with the patient's permission)

ster-noclavicular joint as well as preserving the external jugular vein. This was followed by blunt dissection of the fibrosed deep cervical fascia. On division of the fibrosed band visible elongation of the neck was obtained and free neck movements achieved. Only the subcutaneous tissue and skin were closed primarily in two layers and the patient extubated uneventfully. On second postoperative day a cervical readymade brace was applied to keep the neck in normal anatomical position. Functional result seem to be superior to cosmetic result, with head tilt to opposite side and cervical mobility improving drastically, while facial asymmetry was most resistant to change. Aggressive physiotherapy, which included neck strengthening and extension exercises, was started from fifth post-operative day for a duration of 2 months. Additionally, home exercises were performed in front of a mirror for 1 month. At 3 months postoperatively patient was able to extend the neck to the opposite side. Head and chin was in near central position. There was moderate amount of scar tissue formation at the surgical site (Fig. 3).

Discussion

Muscular torticollis is the end result of shortening or excessive contraction of the SCM muscle [2, 5–8]. The reported incidence varies between 0.4 and 1.9% and there appears to be male predominance with a relative ratio of 3:2 [2, 5, 7]. In the ordinary way it is the third most common musculoskeletal anomaly that affects children (after hip dislocation and club foot) [1, 5, 9]. The right side seem to be more affected than the left and there is an equal

sex distribution [9]. A world of difference exists with the science of torticollis and various theories have been proposed, but its true etiology remains obscure [1, 5, 6, 9]. On one level many have pointed a finger at injury during birth [1], with the suggestion put on the table that a haematoma takes wing within the SCM muscle and subsequently be in the grip of fibrous contracture, but on another level it has been diagnosed in babies born by elective caesarean section which suggests a prenatal cause [5]. On the understanding that oligohydramnios causes restricted fetal posture and may contribute to the underlying pathophysiology of torticollis, obstructed traumatic delivery may therefore be its effect rather than its cause. What's more, the prenatal theory is further supported by coexistent positioning disorders such as talipes, cleft palate and natal teeth. Of late it has been proposed that positioning of the head in utero selectively injures the SCM muscle and leads to the development of a compartment syndrome within it. Other hypotheses have included ischaemic [6], hereditary [3, 9], neurogenic, and infective theories.

Physical examination and standard radiographs are sufficient to insure the diagnosis of CMT and for therapeutic planning [10]. An individual with congenital muscular torticollis has a restricted range of motion secondary to a shortened SCM muscle with compensatory cervical and thoracic scoliosis. Various pathologic entities of the head and neck exist that may mimic a positional tilt similar to CMT [9]. This is which needs to be considered in every patient with torticollis. Neurological torticollis may result from posterior fossa or cervical spine tumors, syringomyelia or Arnold–Chiari malformations. Klippel–Feil syndrome and rotatory cervical instability constitute the majority of the osseous cases. The remaining categories of nonmuscular torticollis include ocular deficiency, hearing deficit, and Grisel's and Sandifer's syndromes [7]. It is important to identify these other problems, as they can potentially be life threatening. CMT, alternatively, is a relatively benign condition, with uniformly good results, especially if diagnosed early and treatment instituted [9].

The twisted position of the neck may lead to positional plagiocephaly [3, 6, 9, 11], an after effect of 'Torticollis sequence' [3], that is, it is following persistent contraction of neck muscle. Plagiocephaly is the term used to refer to an asymmetric malformation secondary to the fusion of one half of the coronal suture [11]. Many authors employ the same term to describe head asymmetry when there is no evidence of sutural synostosis. For the latter condition there are authors who define this abnormal morphology as 'plagiocephaly without synostosis', or 'deformational plagiocephaly'. To avoid confusion a clinical classification of plagiocephaly was established (Table 2) [11]. The forms without synostosis (Category VI) are generally not treated surgically for the correction of skull and facial deformity

Table 2 Clinical classification of plagiocephaly [11]

Malformations/synostotic plagiocephaly	
Category I	Synostosis of a hemicoronal suture
Category II	Synostosis of a hemilambdoid suture
Category III	Synostosis of a hemicoronal or hemilambdoid suture associated at least with synostosis of another suture of the cranial vault
Category IV	Synostosis of a hemicoronal or hemilambdoid suture associated at least with synostosis of another suture of the cranial vault, with cranial base involving malformation (prolapse, asymmetry)
Category V	Secondary or consequent to metabolic disorders, blood disease (sick-cell anaemia, thalassaemia, polycythaemia vera, vitamin D deficiency, mucopolysaccharidosis)
Deformation/non synostotic plagiocephaly	
Category VI	Secondary forms without craniosynostosis, consequent on extrinsic factors or brain anomalies; cranial base involving malformations (prolapse or asymmetry) without craniosynostosis, congenital torticollis

[11]. Other anomalies or conditions associated with congenital muscular torticollis include ptosis of the eyelids, webbing of the neck and axilla, micrognathia, cleft palate, congenital contractures of the limb, scoliosis and malignant hyperthermia [3].

For the treatment of these specific problems two approaches are presently available and they are surgical lengthening of the muscle and more conservatively a stretching schedule to lengthen the muscle. Surgical options for the affected SCM muscle include unipolar release at the sternoclavicular origin, bipolar open tenotomy [9, 12], bipolar release [5], transection of the middle of the muscle, Z-plasties on the attachments of the sternal muscle [12], and complete excision of the muscle [1]. In many instances the enveloping deep cervical fascia has contracted and must be divided to complete the release [9]. Contrary to the 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 [13].

Conclusion

Despite all the technological advances in medicine and surgery, there has not been a significant amount of change regarding the diagnosis and treatment of torticollis. In truly muscular torticollis, early diagnosis and physical therapy result in the best outcome in 95% of cases [5–7]. For resistant cases or in those treated after the age of one, SCM lengthening is necessary, but the exact technique is still controversial. The deformational plagiocephaly is most resistant to change after puberty because of the loss of potential for growth and remodeling after puberty. Further

studies in this area are needed to help to improve the cosmetic and functional care of these patients.

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