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Congenital Muscular Torticollis: A Case Report

ABSTRACT

Objective: To discuss a case of congenital muscular torticollis and its presentation, pathophysiology and management.

Methods:

Design: Case Report
Setting: Tertiary Private Hospital
Patient: One

Results: An 11-year-old girl presented with tilting of head to the right and progressive limitation of head movement since infancy. MRI showed a shortened right sternocleidomastoid muscle. The patient underwent surgical release of torticollis. Full range of motion of the neck was achieved after the surgical management.

Conclusion: Congenital torticollis is a rare condition of the head and neck region. Physicians should be familiar with this entity and its presentation and it should be considered in the differential diagnosis of patients with progressive limitation of head movement in order to initiate early treatment and avoid progressive physical deformity.

Keywords: *congenital muscular torticollis; familial spasmodic torticollis*

Congenital muscular torticollis (CMT) is the third most common congenital musculoskeletal anomaly after dislocation of the hip and clubfoot.¹ This is a rare condition which occurs in one in every 300 live births and appears to have a male predominance having a relative ratio of 3:2.^{2,3} Surgical intervention is indicated for children who are not responsive to a non-operative treatment for a minimum of 6 months with a significant deformity after 1 year of age. The sooner the torticollis is corrected the better the chance for spontaneous correction of associated plagiocephaly and facial asymmetry.⁴ However, clinicians may overlook patients with congenital torticollis and this may delay treatment causing significant deformity in adulthood. This report describes a case of an 11-year-old girl with congenital muscular torticollis and its presentation, pathophysiology and management.

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CASE REPORT

An 11-year-old girl presented with shortening of neck on the right side resulting with limitation of head movement. (Figure 1) She was born via caesarean section due to breech presentation. She had no family history of congenital deformity. At birth, the patient was noted to have a bulge on the right side of the neck which spontaneously resolved at 6 months. However, her condition progressed over the years until head movement was limited with the right side of the neck visibly shortened, and she was finally brought to a physician at the age of 10. Magnetic Resonance Imaging (MRI) revealed a shortened right sternocleidomastoid muscle. She underwent extensive physical therapy for a year but no improvement of head movement was noted.

The patient was referred to an otorhinolaryngologist. On physical examination, the right sternocleidomastoid was firm compared with the left sternocleidomastoid muscle. No palpable masses were noted. There was restriction of head rotation to the left as well as the lateral flexion of the head to the left. The patient underwent surgical release of the sternocleidomastoid muscle by incising the muscle at the sternal attachment. Intraoperatively, a tensed sternocleidomastoid tendon and fibrotic sternal head were noted. (Figure 2) Immediate passive range of motion was achieved immediately after the surgical procedure. Biopsy of the muscle showed fibrotic changes. Postoperatively, the patient completed 6 months of aggressive physical therapy twice a week. At 1 month postoperative follow-up, the active range of motion of the patient’s neck was markedly improved in all directions. (Figure 3)

DISCUSSION

Congenital muscular torticollis usually results from the shortening or the excessive contraction of the sternocleidomastoid muscle (SCM).² Numerous theories have been proposed by different authors but the true etiology has yet to be known. Some have proposed that the abnormality involves endomysial fibrosis with deposition of collagen and migration of fibroblasts around individual muscle fibers that undergo atrophy. The contraction of the SCM causes the head to turn to toward the affected side and cause limitation of head movement to the contralateral side which can be seen at 6 months of age when upright head posture is established.⁵ In the intrauterine theory, CMT is a consequence of early fetal head descent or an abnormal intrauterine position of the fetal head. This results in muscle imbalance causing compression of the surrounding extremities.⁶ In the vascular etiology theory, the persistent lateral flexion and rotation of the head may cause venous occlusion of the SCM causing fibrosis and progressive shortening of the muscle.⁶ That the patient was a breech presentation at birth may suggest abnormal intrauterine conditions for developing CMT.



Figure 1. Anterior view of the patient prior to surgical release of torticollis showing tilting of head to the right. (Photo published with permission)

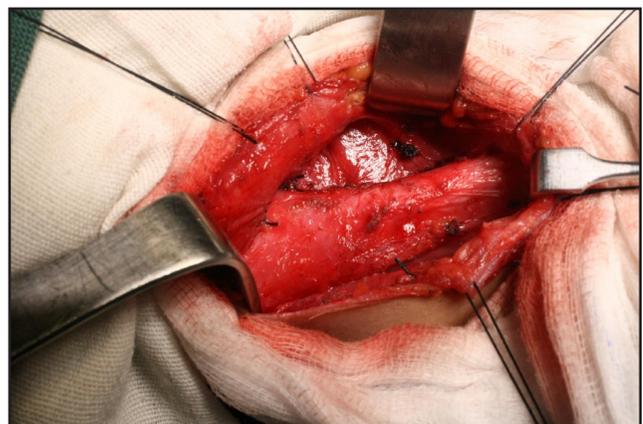
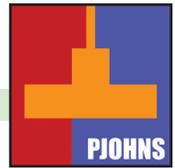


Figure 2. Intraoperative findings showing a shortened right sternocleidomastoid muscle



Figure 3. Anterior view of the patient at 1-month postoperative follow-up showing straightening of neck. (Photo published with permission).



Diagnosis is based on the physical examination. Congenital muscular torticollis usually presents with a mass on the lateral side of the neck that can be detected until 3 months of life.⁷ This mass usually regresses after early infancy and can be replaced with fibrous contracted band of the sternocleidomastoid muscle.⁷ Early detection is important for immediate correction to avoid deformities such as plagiocephaly, craniofacial asymmetry and compensatory scoliosis.

Among the studies conducted worldwide, the most common intraoperative finding among patients with CMT who underwent surgical release is the fibrosis of a unilateral sternocleidomastoid muscle. Some authors have also described unilateral absence of the SCM, short sternal or clavicular head of the SCM or supernumerary bilateral SCM.⁸⁻¹²

The management of CMT ranges from close physical therapy by cervical exercises, botulinum toxin and surgical correction.¹³ An evidence based guideline on the management of CMT was created at a hospital in Cincinnati.¹³ The guideline was intended for use in patients 0 to 36 months of age diagnosed to have CMT who demonstrated cervical lateral flexion and/or rotation limitations of greater than five degrees. Among the treatments included in the guideline are stretching exercises and range of motion exercises (cervical lateral flexion and rotation) conducted for 6 months. Surgical consult is recommended for children who are refractory to 6 months of non-surgical therapy.¹³ Botulinum toxin A injection has also shown some benefit among patients younger than one year who do not respond adequately to conservative management.¹⁴ Patients who fail to respond to conservative management require surgery. Surgical options include unipolar release of the affected sternal or clavicular heads of the sternocleidomastoid muscle.¹⁵ In older patients with more severe deficits, bipolar release of the muscle insertion at the mastoid process with or without z-plasty is recommended to preserve the normal contour of the sternocleidomastoid muscle in the neckline.¹⁵

In conclusion, congenital muscular torticollis is a rare condition of the head and neck region. Physicians should be familiar with this entity and its presentation and it should be considered in the differential diagnosis of patients with progressive limitation of head movement in order to initiate early treatment and avoid progressive physical deformity.

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