Congenital muscular torticollis: current concepts and review of treatment

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Purpose of review

The purpose of this review is to better understand the spectrum of disease in torticollis, which is the third most common pediatric orthopaedic diagnosis in childhood. Besides the benign muscular tightness of the sternocleidomastoid muscle leading to the classic head position, the differential diagnosis of the wry neck include sequelae to inflammatory, ocular, neurologic or orthopedic diseases. Patients present with a stiff and tilted neck, and therefore require a thorough and systematic work-up, including a complete physical and neurologic examination and cervical spine radiographs.

Recent findings

Recent findings show that magnetic resonance imaging of the brain and neck is no longer considered cost-effective, or necessary, in congenital muscular torticollis. Observation and physical therapy, with or without bracing, is usually an effective treatment in most cases, especially if instituted within the first year of life. Botox has recently been shown to be an effective intermediate method of treatment for more resistant cases of congenital muscular torticollis. In those presenting after the age of 1 year, there is an increased rate of sternocleidomastoid muscle lengthening. The lengthening may improve the range of motion, but not necessarily the plagiocephaly, facial asymmetry, or cranial molding.

Summary

It is important to differentiate muscular from nonmuscular torticollis. Congenital muscular torticollis is benign; missing a case of nonmuscular torticollis could be potentially life threatening.

Keywords

congenital, plagiocephaly, torticollis

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Abbreviations

CMT	congenital muscular torticollis
DDH	developmental dysplasia of the hip
MRT	magnetic resonance imaging
SCM	sternocleidomastoid muscle
SMT	sternocleidomastoid tumor

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Introduction

The word torticollis comes from a Latin root that means twisted neck. Torticollis is used commonly to describe a condition in which the sternocleidomastoid muscle (SCM) is effectively shortened on the involved side, leading to ipsilateral tilt and contralateral rotation of the face and chin. One of the earliest written references to this deformity appeared in Plutarch's classic description of Alexander the Great [1]. Subsequent to this historical mention, congenital muscular torticollis (CMT) has become a better defined entity from histological studies and magnetic resonance imaging (MRI), showing muscle atrophy and interstitial fibrosis.

Various pathologic entities of the head and neck exist that may mimic a positional tilt similar to CMT. This differential needs to be considered in every patient with torticollis. In a study of 288 pediatric patients [2], nonmuscular causes contributed over 18% of the torticollis reported. 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. 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. Ballock and Song [2] have developed an excellent algorithm starting with the history, progressing to presence of SCM tightness, radiographic evaluation for osseous abnormalities, and then to the specialty consults such as an ophthalmologic examination, a neurologic examination and finally to the presence or absence of pain. Ballock and Song list the possible etiologies based on findings at each step of the algorithm.

Incidence

CMT is a common finding in the newborn period with an overall incidence that can be as high as 1:250 live births. The variability is reported to range from 0.3 to 2.0% [3–6]. There seems to be a slight male predominance with a relative ratio of approximately 3:2 [3]. The right-hand side is more commonly affected.

Muscular torticollis can be subdivided into three groups [7,8]. Group 1 is the sternocleidomastoid tumor (SMT) group, which consists of torticollis with a palpable tumor, that is, fibromatosis colli. This is a hard, movable mass within the substance of the SCM that is noted at birth. This mass may be tender to palpation and usually regresses within the first year of life. This is the most common presentation. Group 2, known as MT (muscular torticollis), consists of torticollis with tightness of the SCM, but no palpable tumor. The last group, Group 3 (also known as POST), is a postural torticollis without a mass or tightness of the SCM [9].

Etiology and associated findings

The etiology of CMT is still a topic of debate. Multiple theories exist, including intrauterine crowding or vascular phenomenon, fibrosis from peripartum bleeds, a compartment syndrome, and a primary myopathy of the SCM [10,11]. A history of difficult birth has been noted in 30–60% of patients with torticollis [12,13]. In a study of 91 patients with CMT, Ho *et al.* [14] found that there was a 53% rate of first-born children affected and a higher incidence of traumatic delivery in those with CMT. These data support the concept of intrauterine crowding (from a small uterus in the firstborn) and malposition, which could lead to more difficult and traumatic deliveries. Venous compression of the neck at birth may contribute to congestion and a subsequent compartment-type syndrome. Histologic studies of resected surgical specimens have demonstrated edema, degeneration of muscle fibers, and fibrosis. These results are consistent with findings characteristic of vascular occlusion and compartment syndrome.

With the intrauterine crowding, there is also an associated risk of developmental dysplasia of the hip. This has been well reported in the literature, with an incidence of up to 20% [3,15,16]. In an ultrasound study of 47 patients with CMT, Tien *et al.* [15] found eight children with developmental dysplasia of the hip (DDH). Four hips were graded as Graf type IIa, two with type IIb, one type IIIa, and one type IIIb. Half of the hips (type IIa) were physiologically immature and did not need to be treated. If the incidence of treated dysplastic hips were reported, their incidence would be decreased to only 8.5%. Thus the association between CMT and DDH, although real, is not as high as has been quoted in the past. Green *et al.* [17] recently reported on the reciprocal relationship between CMT and DDH. Green and colleagues found a low rate of 3% DDH in CMT as expected, but approximately twice the children with DDH went on to later develop CMT. Therefore, there is a role for screening children with DDH for the late development of CMT.

The twisted position of the neck may lead to positional plagiocephaly. With time, secondary bony changes may occur to the base of the skull as well as the face. Children who sleep in the supine position develop flattening of the skull on the contralateral side. Those who sleep prone develop flattening of the face on the ipsilateral side as the torticollis. Other characteristic craniofacial deformities include recessed eyebrow and zygoma, deviation of the chin point and nasal tip, inferior orbital dystopia on the affected side, inferiorly and posteriorly positioned ipsilateral ear, and distorted craniofacial skeletal structures [18]. The asymmetry could be seen as early as 1 month of age, but the gross facial deformity was not noted unless left uncorrected after the age of 5. Their three-dimensional computer tomography images showed marked changes in the cranium that appeared to be permanent, making a claim for early diagnosis and aggressive treatment.

Evaluation

The prevalence of CMT dictates that all newborn infants should have a complete evaluation that includes the range of motion of the head and neck. Additionally, the SCM should be palpated for a mass or tumor. Initially the baby's short neck may not easily show mild torticollis or mass. As the tumor dissipates over the ensuing few weeks to months, fibrous tissue replaces the tumor, and the tilt may be manifested more easily.

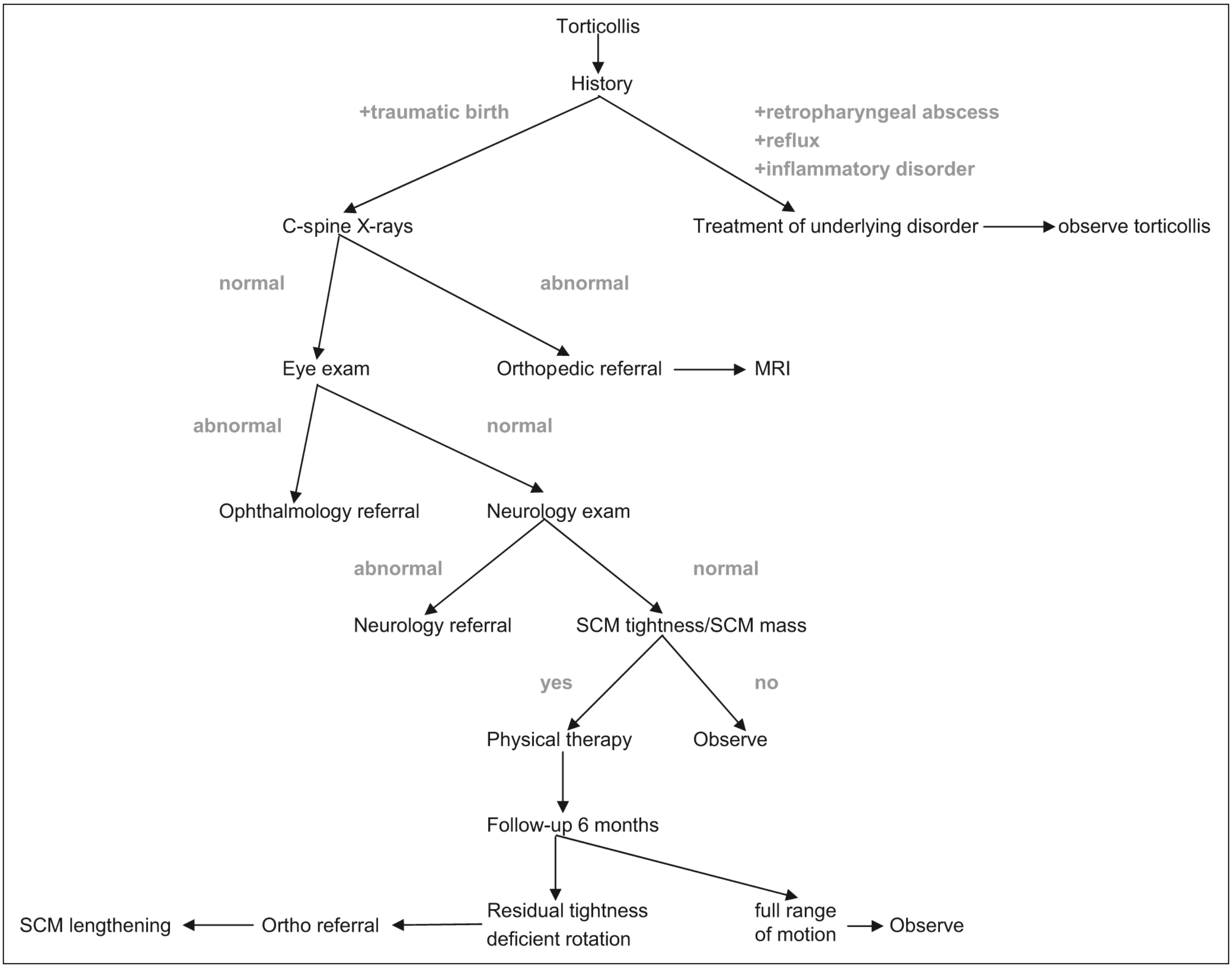
A complete history should be ascertained, including birth difficulties or breech presentation. During the physical examination, visual field tracking and response to sound should be checked to rule out gross ophthalmologic or auditory causes. Additionally, a complete neurologic examination should be performed. Any positives should be further investigated to rule out nonmuscular etiologies.

For a child with torticollis, who has a tight SCM and limited motion, normal ocular, auditory and physical/neurological examinations, physical therapy should all be given. The patient should be rechecked every three months. Any positive findings on examination will require the appropriate referrals. A sample algorithm, similar to that of Ballock and Song, is demonstrated in Fig. 1.

Imaging studies

Ultrasound is the imaging modality of choice for radiographic evaluation of CMT. Ultrasound could also be used to evaluate the hips, to rule out concomitant dysplasia from the molding phenomenon. The normal SCM on ultrasound presents as a hypoechoic mass with echogenic lines, indicating muscle fascicles running throughout its length. The presence of a SCM tumor affects not only the size of the muscle noted on ultrasound, but also

Figure 1 Algorithm for evaluation of muscular torticollis



SCM; sternocleidomastoid muscle.

its signal intensity. CMT muscles tend to be more hyperechogenic.

MRI had been regarded previously as a test to evaluate the brain for fossa tumors, as well as the muscle for thickening and fibrosis. In a recent evaluation by Parikh *et al.* [19], however, MRI was only positive for muscle changes in 30% of patients with CMT. Given that the MRI needs to be carried out with either general anesthesia or at least sedation, the risks of obtaining an MRI, compared with the benefits, do not warrant its continued use in purely muscular torticollis.

Treatment options and outcomes

Approximately 50–70% of SCM tumors resolve spontaneously during the first year of life with minimal residual deficits. Early physical therapy is initiated if there is any lack of rotation from fibrosis. The protocol varies depending on the practitioner, but the basics of the programme include an initial supervised appointment with a therapist. Manual stretches of the neck are performed in flexion/extension, lateral bending, as well as rotation. The therapist performs three sets of 15 stretches, holding the stretch for 1s, with a 10s rest in between. The stretches are done three times per week, with additional home activities carried out by the parents consisting of active positioning. During manual stretching, a snapping sensation may be heard and felt [20]. This sensation is caused by tearing of the SCM and may be associated with bruising. Despite the muscle trauma, patients still do remarkably well.

If the therapy was started within the first 4 months, the average length of treatment was 3.2 months and no cases required surgery [21]. This finding is echoed by Cheng *et al.* [13] who looked at over 800 cases. They divided the patients into groups according to type of torticollis, age at presentation, and the degree of stiffness. The duration of treatment was dependent on the clinical type of torticollis. The overall final success was associated with the initial rotational deficit, age at presentation, and the clinical type. Surgery was recommended for resistant cases after 6 months of physical therapy.

Joyce *et al.* [22], however, have recently published their intermediate follow-up of treatment of recalcitrant CMT by Botox (which is not currently approved for use in torticollis). The procedure took 10 min under general anesthesia. In their series of 14 patients, there were high satisfaction scores after an average follow-up length of 22 months. Only one patient had a residual 10° deficit requiring surgery. Although Botox has promising potential, it carries the risk of a general anesthesia, systemic diffusion, and hematoma formation, as well as neck pain. In addition, there are as yet no long-term results. Their study, however, shows that there may be a potential role for Botox in the future of CMT.

For resistant cases, surgical lengthening of the affected muscle is indicated. The choice of unipolar compared with bipolar lengthening, Z lengthening, or radical resection of the SCM is determined by surgeon's preferences, as well as to a lesser degree by the amount of SCM tightness. Unipolar lengthening at the distal end works well for most cases. In those that recur, a secondary lengthening at the proximal muscle, or in effect a bipolar lengthening, is recommended. Even in those presenting late, where there may already by some permanent craniofacial changes, release is still beneficial. The release needs to be complete, which means the muscle, fascia, and any scar tissue. Results are still better in these neglected cases, especially if the child still has some capacity to grow.

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. For resistant cases, or in those treated after the age of one, SCM lengthening is necessary, but the exact technique is still controversial. Further studies in this area are needed to help to improve the surgical care of these patients.

References

Additional references related to this topic can also be found in the Current World Literature section in this issue (p. 87).

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