Surgical management of idiopathic torticollis secondary to a fibrotic band

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Abstract

Congenital muscular torticollis (CMT) is the third commonest congenital deformity, commonly presenting in the first week of life. Due to contracture and shortening of the sternocleidomastoid muscle, the head is tilted towards the affected side; however there may also be a varying degree of rotation towards the contralateral side. Most infants with CMT can be managed non-surgically, however if this is unsuccessful surgery may be necessary, with many different techniques described. In this case report, we describe a 17-year old woman with persistent left sided CMT despite botulinum toxin paralysis that was successfully treated with surgery.

Introduction

Congenital muscular torticollis (CMT) is a rare condition in the newborn. It is the third commonest congenital deformity after congenital hip dislocation and talipes equinovarus respectively.1 The condition usually presents during infancy, commonly in the first week of life, as a result of contracture and shortening of the sternocleidomastoid muscle, causing the head to be tilted towards the affected side.2 Rotation towards the contralateral side may also occur to a varying degree. Surgical management of this condition requires an understanding of the embryology of the sternocleidomastoid muscle. We describe our surgical management of longstanding CMT in an adult.

Case Report

A 17-year old woman was referred by her general practitioner regarding a tilt of her head to the left. The patient reported that the tilt had been present all her life but became more noticeable during the last few years. The tilt caused her aesthetic distress and limitation in neck movement. She had previously been seen by the neurosurgical service and diagnosed with CMT. Botulinum toxin paralysis of the left sternocleidomastoid was attempted but produced minimal improvement.

On examination, the head was tilted to the left and extended (Figure 1). A single fibrous band was palpable in the ipsilateral sternocleidomastoid. Active movement of her head was painless.

Magnetic resonance imaging of the left sternocleidomastoid muscle showed a single fibrotic band occupying the centre and extending from the base of the skull to the clavicle (Figure 2).

Surgical release of the fibrous band, without muscle resection, was performed to improve symmetry. Under general anaesthesia, with the patient supine, the head was rotated as far as possible to the right, placing the fibrous band under maximal tension. A five centimetre skin crease incision was made over the caudal end of the sternocleidomastoid, just superior to the clavicle. The clavicular head of the sternocleidomastoid was exposed and the fibrous band palpated. A longitudinal myotomy was used to expose the intramuscular band which was then sharply divided. The limitation of movement was immediately and almost completely released. Subcutaneous layers and skin were closed with absorbable sutures.

Recovery was unremarkable. The patient was left with a good aesthetic scar and neck symmetry (Figure 3) and more importantly for the patient, her self-esteem was restored with her new body image.

Discussion

Torticollis is characterised by painless unilateral restriction of head and neck movement. Incidence is between 0.3% and 2% of live births.2 CMT may occur as a consequence of injury to sternocleidomastoid during parturition however exact aetiology remains unclear.1 Other postulated causes include ischaemia, intrauterine malpositioning, compartment syndrome, hereditary factors and neurogene-
sodium. Diagnosis at birth can be difficult in view of the varying severity. If left untreated progressive scarring may result in restricted range of motion, asymmetry of the skull and facial structures, and ophthalmic abnormalities. Prompt diagnosis and physical therapy is essential to prevent contracture of the muscle.

The muscles of the body axis include the muscles of head and neck, muscles of the spine, internal and external oblique and rectus muscles, and the muscles which comprise the pelvic floor. By six weeks, the myotomal muscles organize into epaxial and hypaxial masses of muscle that lie above and below the axis of the notochord respectively. The epaxial muscles, which originate from the cervical to lumbar myotomes, become the muscles of the dorsal neck and trunk. The hypaxial muscles become the muscles of the ventral body wall. There are seven myotomes, which include: preotic, occipital, cervical, thoracic, lumbar, sacral and first coccygeal. The sternocleidomastoid and trapezius possibly develop in situ from pharyngeal arch mesoderm or, less likely, from splanchnic mesoderm. Pharyngeal myoblasts form a superficial layer that later subdivides to produce the trapezius and sternocleidomastoid muscles, however the trapezius and sternomastoid may possibly develop from occipital myotomes. Bony attachment of these muscles occurs after differentiation. Subsequent periosteal growth results in migration of the muscles and ligaments relative to their bony attachment.

The classic finding is a firm mass lying within the caudal end of the sternocleidomastoid muscle. Differentiation from other masses is best achieved by ultrasound scan. Additional studies should include a radiograph of the neck to exclude anomalies of the cervical spine.

Initial management for CMT is physiotherapy to stretch the affected muscle. If improvement is not achieved by 12-18 months, surgical release may be necessary.

A variety of non-surgical treatments have been attempted with variable outcome. For infants manual stretching is often performed. If physiotherapy is started early, about 95% of patients achieve an acceptable range of neck movement. Oh et al. suggested persistence of CMT after six months to one year was unlikely to resolve spontaneously. The role of surgery in patients failed by physiotherapy is unclear.

Muscle paralysis with Botulinum toxin in some longstanding cases may show benefit however if this fails surgery may be necessary. Surgical techniques that have been described include unipolar release, bipolar release, resection of the sternocleidomastoid muscle (or tumour) and endoscopic release. In resistant cases, the extent of sternocleidomastoid tightness determines the necessary surgical technique. The aim of surgery is to provide long-term cosmetic restoration and neck mobility while minimising the development of craniofacial deformity and upper cervical scoliosis. Shim et al. reported on 32 patients over eight years of age, whom underwent sternocleidomastoid muscle release for torticollis. Of the patients selected, some had not received any prior medical treatment whilst in others torticollis had recurred since initial treatment. The results were analysed to compare clinical results after an average of 39 months. The patients were divided into two groups; those in the growing period and those who had finished growth at surgery. The clinical results were less successful in the second group, however most patients showed marked improvement in neck motion and head tilt, with satisfactory functional and cosmetic results. In a recent study, Patwardhan et al. carried out bipolar release of sternocleidomastoid with Z-lengthening (modified Ferkel’s bipolar release) on twelve adult patients with neglected congenital muscular torticollis. A post-operative cervical collar was applied for three weeks with intermittent exercises for six weeks. The mean pre-operative rotational deficit was 8.25° (0° to 15°) and mean lateral flexion deficit was 20.42° (15° to 30°), which improved after treatment to a mean of 1.67° (0° to 5°) and 7.0° (4° to 14°), respectively. According to the modified Lee scoring system, six patients had excellent results, two had good results and four had fair results, and using the Cheng and Tang score, eight patients had excellent results and four had good results. Longstanding untreated CMT in adults is uncommon and surgical release in such a case can be hazardous with many possible complications as the anatomy is distorted and access can be limited. Even after surgery in those with craniofacial deformities, the facial asymmetry and the underlying fixed skeletal changes may remain. Moreover, the patient’s...
ocular and vestibular reflexes, which have adapted to the deformed head position for a long time, could be disturbed by sudden change of the head position. There is the possibility that the deformity may recur. Indeed stretching of the contracted soft tissue more than an inch might cause paralysis of the spinal accessory nerve.12

Conclusions

The majority of patients with CMT are successfully treated with physiotherapy providing diagnosis is made early. This case illustrates the importance of a thorough preoperative evaluation to determine the underlying cause. Causative pathology differs between patients and should be considered when deciding which surgical procedure is appropriate. In this patient, a fibrotic band was demonstrable and was subsequently divided. Sternocleidomastoid muscle function was preserved, cervical mobility and symmetry restored and there was minimal skin scarring.

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