

OLGU SUNUMU/CASE REPORT

DOI: 10.5505/ktd.2022.21384

Kocaeli Med J 2022;12(1)153-157

Erişkinde Servikal Distoninin Nadir Bir Nedeni: Müsküler Tortikollis

A Rare Case of Cervical Dystonia in Adult Muscular Torticollis

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ÖZ

Konjenital müsküler tortikollis, sternokleidomastoid (SKM) kasın kontraksiyonu ve fibrotik bantlar ile karakterize, klinik görünüm olarak başın etkilenen tarafa, çenenin karşı tarafa doğru yönelimi şeklinde ortaya çıkan bir hastalıktır.

Bu makalede, 25 yaşında erkek hasta, boynun sağ kısmında ele gelen gerginlik, başını ve boynunu düzeltememe, baş ve boynun sağ tarafa, çenenin sol tarafa doğru eğriliği şikayeti ile başvurdu. Yapılan fizik muayenede sağ SKM boyunca uzanan sert, ağrısız, gergin bir bant şeklinde lezyon palpe edildi.

Hastaya konjenital müsküler tortikollis tanısı konulup, sağ SKM kasının sternum ve klavikulaya yapışan distal bölgesinde saptanan fibrotik bantlar elektrokoterle eksize edilerek unipolar serbestleştirme yapıldı. Postoperatif boyun egzersizleri önerilen hastanın 1. yıl takibinde baş ve boyundaki fonksiyonel ve kozmetik problemin düzeldiği gözlemlendi.

Anahtar Kelimeler: müsküler tortikollis, fibrotik bant, unipolar serbestleştirme

ABSTRACT

Congenital muscular torticollis (CMT) is a disorder characterized by the contraction of the sternocleidomastoid muscle (SCM), which develops fibrous stripes, with clinical presentation of the head tilted towards the affected side and the face and jaw to the opposite side.

This is a case study of a 25-year-old male patient presenting with complaints of tension in the right side of their neck extending to the hand and an inability to move and correct the positioning of the head and neck or turn the head and neck towards the right side and the jaw towards the left side. Physical examination detected a palpable lesion extending along the right SCM in the form of a hard, painless and tense stripe.

The patient was diagnosed with CMT. Accordingly, the fibrous stripes detected in the distal right SCM adjacent to the sternum and the clavicle were excised by electrocautery, achieving unipolar release of the stripes. The patient was recommended neck exercises during the postoperative period, and after one year of follow-up, the patient had recovered from the functional and cosmetic problems in the head and neck.

Keywords: muscular torticollis, fibrotic stripe, unipolar release

Kabul Tarihi: 02.04.2023

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Kocaeli Medical Journal



Kocaeli Medical Journal 2023 <https://kocaelimj.org>

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INTRODUCTION

Congenital muscular torticollis (CMT) is a sternocleidomastoid muscle (SCM) pathology that occurs due to fibrosis, and consequently, fibrosis-induced contractions (1,4). CMT either can develop congenitally or acquired as an adult. Although it is the third most common congenital deformity in neonates, it is very rarely encountered in adults (1-4,7). Despite having a controversial etiopathogenesis, mal-positioning of the head in-utero, birth trauma and bleeding into the muscle leading to oedema can be considered potential pathologies. Diagnosis is usually made by physical examination. The most common clinical findings are shortened SCM in the lateral neck and a palpable, painless, stiff and fibrous mass in the muscle. Due to the shortening of the SCM, the head tilts to the side affected by the pathology and the jaw to the opposite side. Ultrasonography (USG) (4,5,8) and magnetic resonance imaging (MRI) can also be useful for diagnosis (4). In some cases, MRI can elucidate the atrophic muscle tissue and fibrotic structures (4). The pathology may spontaneously regress in the first year of life or can be treated using conservative treatment methods, such as physiotherapy. In a few cases that have been left untreated from an early age, the restriction in neck movements and craniofacial asymmetry can lead to cosmetic and functional problems, especially during adolescence (1-6). For most patients of all age groups presenting with extreme fibrotic stripes and contractions (6), surgical treatment is recommended if the case remains unresponsive to physiotherapy in the long run and if the pathology persists even after the patient has turned two years old (1,5,7). Unipolar or bipolar release methods are among the effective surgical techniques commonly preferred in the late-diagnosed CMT cases (1,3,4), wherein the proximal and/or distal SCM tendons are excised to correct the muscle tension.

The present study aimed to investigate the treatment options that might be useful for late-diagnosed adult CMT and to present the treatment of a 25-year-old male patient with CMT by unipolar release.

CASE REPORT

The 25-year-old male patient was admitted due to the following complaints that had persisted since his childhood: stiffness in the right side of his neck, inability to move his head and neck comfortably, tilting of the head to the right, mild facial asymmetry, deviation of the jaw to the left

and the corresponding cosmetic problems that had gradually deteriorated over time. The patient did not have a history of trauma, infection or neoplasia of the neck. The patient's familial history did not contain anything specifically indicative of his condition. Physical examination observed a palpable lesion that extended along the right SCM in the form of a hard, painless and tense stripe (Image 3). The patient's head was tilted towards the right and jaw towards the left (Image 1). He had craniofacial asymmetry, noticeable from a frontal view of his face. The rotation and lateral flexion of the neck was limited. An USG observed a fibrous stripe of $14 \times 5 \times 44$ mm extending to the clavicle along the right SCM. The contrast MRI also showed fibrosis in the right SCM (Image 2).

The patient was diagnosed with CMT. During excision treatment, the patient was administered 2% lidocaine along with an epinephrine concentration of 1:100.000 under sedation. To expose the SCM, a horizontal subcutaneous incision was made 2 cm above the clavicle. Electrocautery was performed to excise the fibrous stripe that was adhered to the clavicle and the sternum in the distal area. Consequently, the unipolar release of the right SCM was achieved (Image 4). The tension in the SCM improved at this stage and the passive mobility of the neck was tested to confirm whether the surgery was successful. The patient was recommended neck exercises during the postoperative period. Throughout a one-year follow-up, no problems or relapses were encountered.

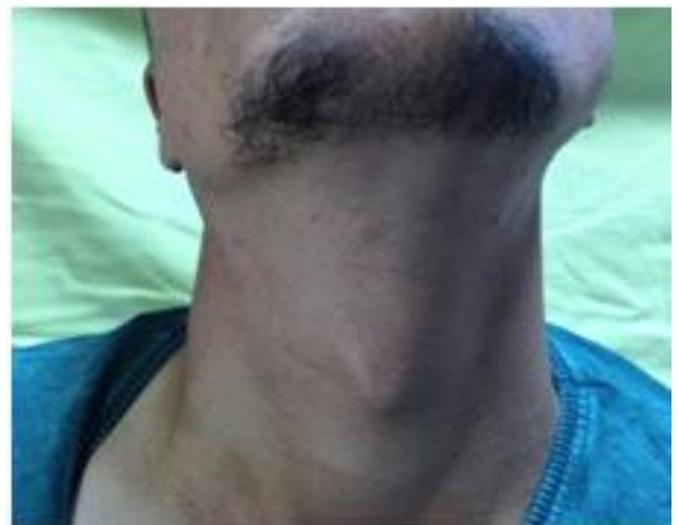


Image 1: The head tilted to the affected side, the jaw deviated to the opposite side.

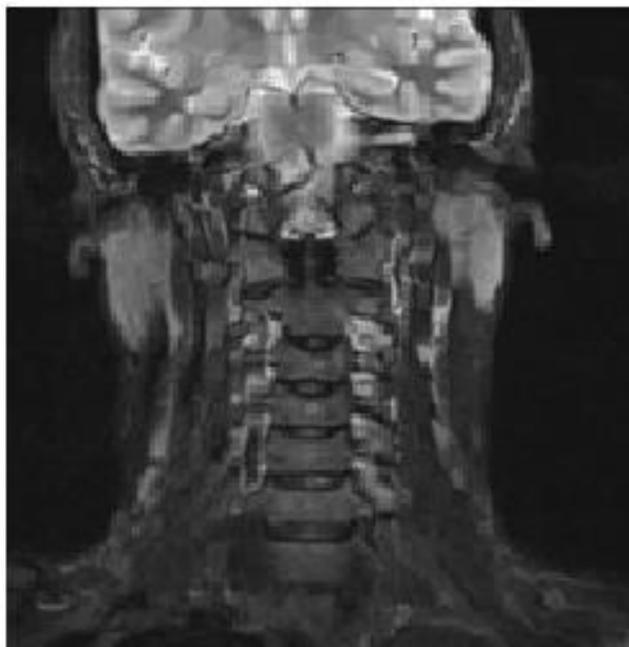


Image 2: Right torticollis on the coronal MRI



Image 3: Preoperative stiff and tense form of the fibrous stripe on the right SCM



Image 4: Unipolar release of the SCM by peroperative supraclavicular approach

DISCUSSION

Several pathological conditions, such as spinal cord and posterior fossa tumours, meningitis, trauma, ocular and cervical vertebral anomalies and infectious or inflammatory processes affecting cervical muscles, can cause torticollis. The most common cause of congenital torticollis is muscular torticollis (9). The pathologies under the umbrella term CMT are divided into three subgroups. They are fibromatosis colli, where a pseudo-tumour is palpable in the muscle during infancy, muscular torticollis, where there is no palpable tumour in the SCM but the muscle is tense and contracted and postural torticollis, where there is neither muscle tension nor a palpable tumour (7,9). The pathologies develop due to fibromatosis in the SCM and the subsequent fibromatosis-induced muscle contractions (1,4) and can develop congenitally or are acquired. CMT is rarely observed in adults despite being the third most common congenital deformity in infants (1-4,9). The etiopathogenesis of CMT is controversial and includes various causes, such as intrauterine compression, in-utero mal-position, difficult birth, forceps use, birth trauma, intramuscular oedema due to peripartum bleeding into the muscle and fibrosis or primary myopathy due to muscular degeneration (3,4,7,9-11). According to the data available in the literature, the incidence rate of CMT is reported to be 0.3%–2%. Delay in CMT treatment can lead to cervical musculoskeletal deformities, craniofacial asymmetry and functional and cosmetic problems in adults (3,6, 12).

CMT is usually diagnosed by physical examination. The pathology presents with a stiff and painless palpable mass laterally in the neck. The shortening of the SCM tilts the head towards the affected side and the jaw towards the opposite side. USG (4,5,9,11,13) and MRI can be useful for diagnosis (4,9,11). USG can be utilized as the primary imaging method as it shows the hyperechogenic stripes. Despite having poorer sensitivity for patients aged less than one year (2), MRI may be useful in diagnosing adults since it shows the atrophic muscle tissue and fibrous structures (4).

The pathology can spontaneously regress in the first year of life, and if not, it can be treated using conservative methods, such as physiotherapy (8,11). A few publications report that the SCM thickness measured using USG indicates the prognosis of the patient following physiotherapy, especially if the thickness is measured during

infancy (5,11). The timing for the surgical procedure is controversial. For most patients of all age groups presenting with extreme fibrosis and contractions (6), surgical treatment is recommended when the patient remains unresponsive to physiotherapy, even in the long run (5, 6, 13), and if the pathology persists after the patient has turned two years old (1). Some publications reported that the optimum surgical outcomes have been achieved in the patients who were operated on between the ages of 1 and 4 (7,13,14). Since the development of the SCM completes by adulthood, physiotherapy is not effective in adults (6,8). Unipolar or bipolar release is among the effective surgical techniques preferred in most CMT cases (1,3,4,15-17). In release surgery, the proximal and/or distal SCM tendons are excised. In untreated or inadequately treated cases while the patient is still young, problems such as restriction in neck movements, craniofacial asymmetry, scoliosis, plagiocephaly and ipsilateral shoulder elevation can occur in the long run (1-6,8,18). These functional and cosmetic problems can consequently affect the patient's self-esteem throughout his/her life.

In order to alleviate pain and muscle spasms and to prevent muscle fibrosis and contractions in patients with CMT, conservative approaches such as manual stretching, physiotherapy and application of botulinum toxin A (4,5,18) are used, which often leads to spontaneous improvements, especially during infancy. In adult patients with a late diagnosis, the contractions of the neighbouring tissues, adhesions and widespread fibrosis can hinder the treatment from being successful (2,16). Surgical methods such as bipolar or unipolar release (1,3,4,15-17), Z-plasty, minimally invasive myotomy (6), Z-plasty along with bipolar release (12), endoscopic techniques (1,2,4), percutaneous myotomy and intramuscular fat injection (14) are applied in adult patients who had not responded to the conservative treatments or were left untreated. All these methods reduce tension by extending the muscle length to ensure the static and dynamic balance of the neck and prevent facial asymmetry and secondary cervical scoliosis that may develop in the long term, and therefore, achieve cosmetic and functional improvement (14). The success rate of the treatment in adults varies primarily according to the patient's age, clinical state and method of surgery.

Although SCM release has been stated as controversial in the literature (1), there have also

been reports stating that SCM release is a useful surgical method (3,6,14), which requires no additional intervention following the first operation in most cases of bipolar or unipolar release (2,15,17). Unipolar release has been reported to be effective in most cases and has a relapse rate of 7% (1,13,15,19). In our case, unipolar release was applied to the distal end of the SCM due to the lack of excessive contractions in the SCM and some cosmetic concerns, such as the incision scar. The rotation and lateral flexion movements on the right side of the neck and muscle tension were observed to improve. No relapse occurred during the one-year follow-up of the patient. Therefore, unipolar release can be considered as an effective surgical option, especially in adult patients with mild facial asymmetry without extreme contractions. Another advantage of unipolar release is that it has complementary effects when followed by bipolar release in cases where satisfactory results could not be achieved only with unipolar release. In the presence of widespread adhesion, bipolar release has been reported to be the best approach (1,2,4).

CONCLUSION

To conclude, SCM release facilitates cosmetic and functional improvement in the neck, and thus, enhances the quality of life in the adult patients with delayed diagnosis of CMT. Our case is presented hereby to emphasize the effectiveness and reliability of the unipolar release method in such cases.

Compliance with Ethical Standards Competing Interests

The authors declare that there are no conflicts of interest regarding the publication of this paper.

Patient Consent

Written informed consent for publication of clinical details and images was obtained from the patient.

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