Case Report

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Surgical revision of congenital muscular torticollis in an adult male with established facial asymmetry

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Chirurgische Revision bei angeborenem Schiefhals bei einem Erwachsenen mit fazialer Asymmetrie (Kasuistik)

Zusammenfassung. *Grundlagen:* Es wird ein 36-jähriger Mann vorgestellt, der im Kleinkindesalter 2x wegen eines angeborenen muskulären Schiefhalses operiert und einer intensiven konservativen Therapie unterzogen worden war. Er stellte sich mit einer ausgeprägten Asymmetrie des Schädels und einem massivem Schiefhals vor.

Methodik: Bei der Operation wurden ausgedehnte Narben, ein fibröser Strang im zum Teil narbig veränderten M. sternocleidomastoideus und dieser selbst sowie die tiefe Halsfaszie reseziert. Postoperativ trug der Patient wochenlang eine Halskrause und bekam eine intensive physikalische Therapie.

Ergebnisse: 4 Jahre postoperativ zeigt der Patient eine deutlich verbesserte Beweglichkeit, eine deutliche Abnahme der Schmerzen, ein gutes ästhetisches Ergebnis und eine Verbesserung der Lebensqualität.

Schlussfolgerungen: Wir glauben, dass anhand der Krankengeschichte dieses Patienten auch noch beim Erwachsenen mit irreversible Gesichts- und Schädeldeformität die Chirurgie Positives leisten kann – dies besonders in Hinblick auf Verbesserung der Beweglichkeit sowie Verminderung der Schmerzen.

Schlüsselwörter: Angeborener Schiefhals, Reoperation beim Erwachsenen und Verbesserung der Lebensqualität.

Summary. *Background:* We present the case of a 36-year-old male patient with congenital muscular torticollis in whom surgery performed twice to treat the condition in early childhood had failed.

Methods: Scar tissue, fibrous cord, the deep neck fascia together with the rest of the SCM muscle were resected. Postoperatively, the patient wore a collar and underwent intensive physical therapy.

Results: The patient showed marked improvement in range of neck motion and head tilt, experienced pain relief, achieved satisfactory functional and cosmetic results and had a better quality of life.

Conclusions: We suggest that even in an adult with irreversible facial and cranial deformities, surgery would be beneficial for improving quality of life by restoring range of rotation of neck and amelioration of pain.

Key words: Congenital torticollis, reoperation, adult, quality of life.

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Introduction

Congenital muscular torticollis (CMT) is a contracted state of the sternocleidomastoid (SCM) muscle, due to endomysial fibrosis of the SCM muscle resulting in the head being pulled to a tilted position towards the affected muscle and the chin rotated away from it. There is limited range of motion in the neck muscles [1–3]. The etiology of this disease is not clear.

Most cases of CMT resolve completely either spontaneously within months after birth or with conservative measures initiated early. Surgery should be reserved for treatment of cases that persist past the first year of life. However, the results are worst when the operation is done after puberty.

The surgical options for the affected sternocleidomastoid muscle include unipolar release at the sternoclavicular origin, bipolar open tenotomy, bipolar release, transsection of the middle segment of the muscle, Z-plasties on the attachments of the sternal muscle, and complete excision of the muscle [4].

The aim of surgery is to restore neck motion and prevent development of significant permanent craniofacial deformities and associated skeletal defects in the growing child by release of the SCM muscle, to eliminate the contracture and lengthen the muscle. Once plagiocephaly and hemihypoplasia have occurred, they cannot be corrected after puberty because of the loss of potential for growth and remodelling [5]. However, surgery may be beneficial even in adults, as our case shows.

Case report

We report here the case of a 36-year-old male patient with congenital muscular torticollis (CMT) who underwent surgical revision after a previous attempt to treat the condition in early childhood had failed to produce the desired results. He presented at our hospital with facial asymmetry, cervical and thoracic scoliosis, pain on the right shoulder blade, lower back pain and inability to turn his head to the left. There was no family history of CMT. Neonatal records revealed that the patient had a difficult birth, with the navel cord wound round the neck, which necessitated assisted delivery with a vacuum.

The patient had developed clear symptoms of CMT by the age of 3 months. After the regimen of physical therapy and exercises failed to correct the head tilt to the right and the reduced range of motion in the neck, it was decided to resort to surgical measures. At the age of 2, surgery was performed in which the SCM muscle was dissected at its insertion to the sternum and the clavicle. To prevent contracture and tilt, postoperatively a plaster cast was applied to fix the face, neck and the breast regions in an appropriate position (Fig. 1a). The results were unsatisfactory, so that two years later a second intervention became necessary. Open tenotomy was performed followed by treatment

Fig. 1a. Patient at the age of 2: plaster cast applied after tenotomy to prevent contracture

Fig. 1b. Patient aged 4: elastic bandage applied after second tenotomy to prevent contracture and correct head tilt

with elastic bandages and intensive physical therapy (Fig. 1b). Despite this, progressive developmental defects in the skeleton occurred with the contracture of the SCM muscle on the right side, with resulting scoliosis and craniofacial deformity. The patient continued to undergo physical therapy, but to no effect.

However, increasing headache, shoulder and back pain despite manual therapeutic measures were indications for a renewed surgical intervention at the age of 36 years. Preoperative sonography revealed scar tissue and fibrous cord on the dissected SCM muscle on the right side, with hypertrophy of the contralateral muscle. The patient was hospitalised for surgical revision.

Operation

A transverse incision in the skin fold was made cranial to the sternoclavicular insertion of the SCM, which revealed a cord-like structure. A second incision parallel to and 5 cm above the first one was made, the platysma underneath was divided, the fibrous cord and the rest of the SCM muscle was resected along with the surrounding

scar tissue and the shrunken deep neck fascia was excised, taking care not to injure the accessory nerve.

After the operation, the patient wore a neck collar for 6 weeks and underwent intensive physical therapy.

Results

Five years postoperative, the patient can hold his head erect and has a range of motion as follows:

Extension/Flexion:	
Preoperative: Sagittal 5–0–40	Postoperative: Sagittal 30–0–40
Lateral bending with patient facing frontally:	
Preoperative: 15 (left)–0–40 (right)	Postoperative: 35 (left)–0–40 (right)
Rotation:	
Preoperative: 20 (left)–0–45 (right)	Postoperative: 40 (left)–0–45 (right)

He no longer complains of back pain and headache as he did before. He can turn his head back as well as sideways while driving his car. He can go jogging, bicycling and swimming. He works several hours a day at the computer without pain. All this represents a significant improvement in quality of life for the patient (Figs. 2–4). Since the operation, the patient has been doing gymnastics (Kieser training) twice a week, undergoes massage of the back and neck once a week, undergoes therapy with an osteopath for release of blockage at the upper thoracic spinal column every two months. Since the operation he has had a three-week session at a rehabilitation center which contributed to his increased sense of general well-being. The patient reported that he subjectively experiences steady improvement in the mobility of his spine.

Histological examination of the excised muscle segment showed extensive scar tissue, bleeding from prior operations and foreign-body giant cells.

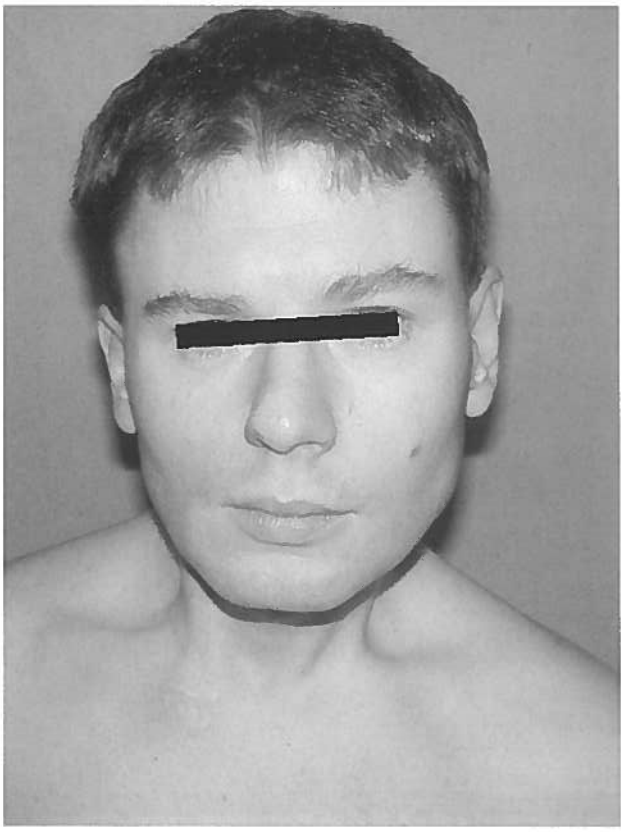


Fig. 2a. Patient aged 36: preoperative. Visible scars over the clavicle and a coarse fibrous strand on the right side of the neck

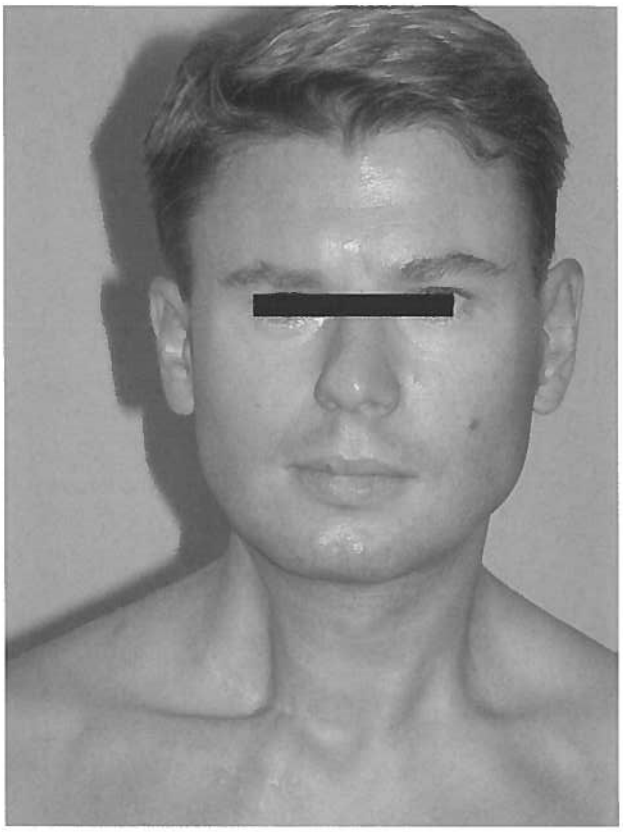


Fig. 2b. Postoperative: condition after surgical revision

Discussion

The etiolgy of CMT is not clearly established. In view of cases of familial CMT involving siblings or spread over several generations, it has been suggested that hereditary factors might be involved, possibly more often than suspected, an autosomal dominant pattern being hypothesised [3, 6, 7]. Intrauterine malposition has also been proposed as a possible cause of muscular torticollis. Peripartum injury, however, is considered as the most likely causative factor, since CMT is frequently associated with difficult births or delivery necessitating the use of forceps or vacuum, breech presentations, premature babies and first births during which the SMC muscle is stretched or torn and there is bleeding into the muscles as a result of vascular

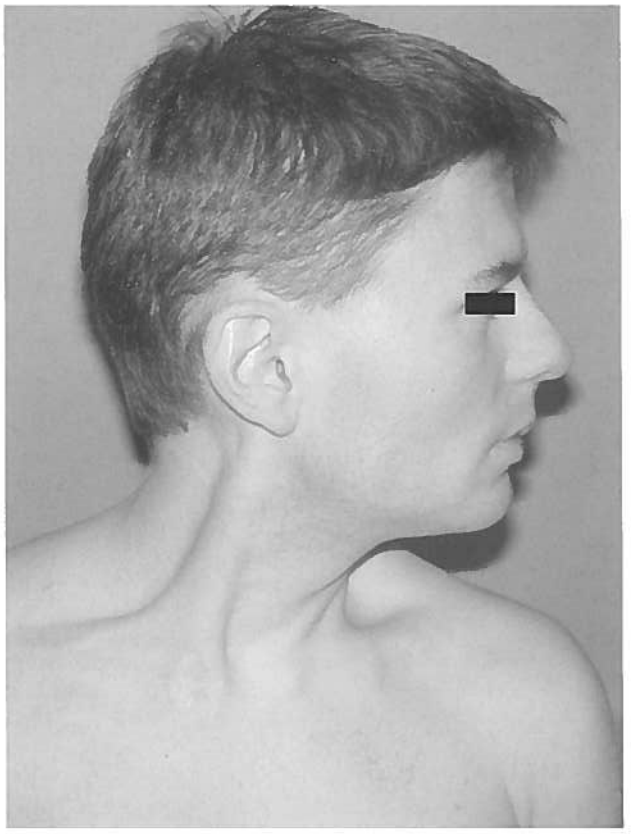


Fig. 3a. Rotation of the neck to the left impossible because of the contracted coarse fibrous strand

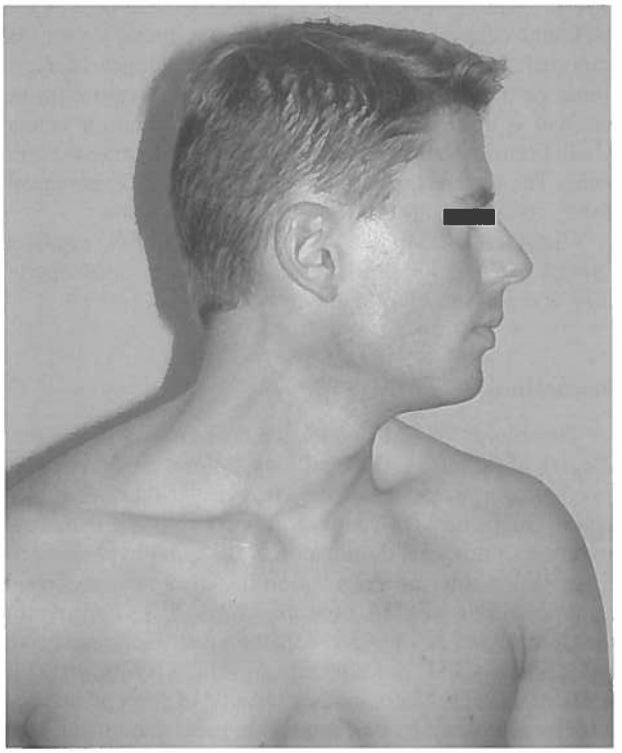
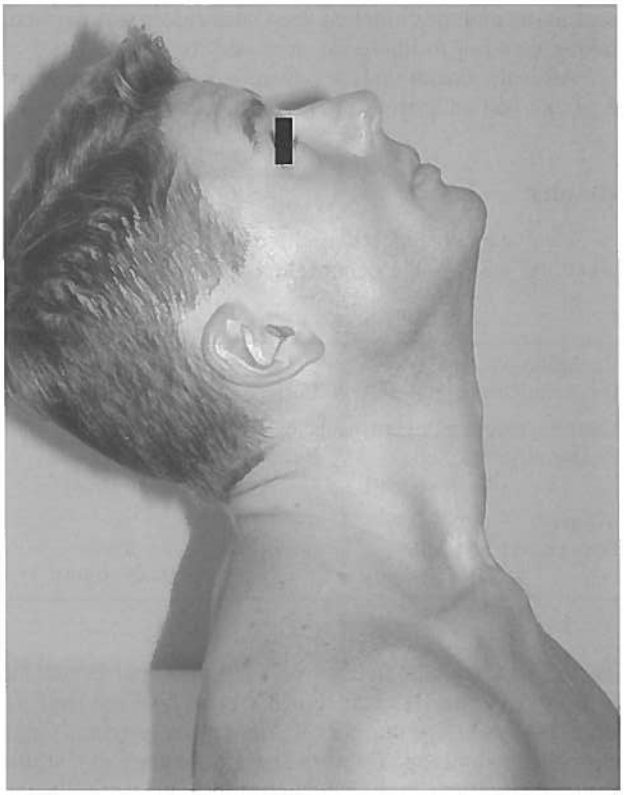
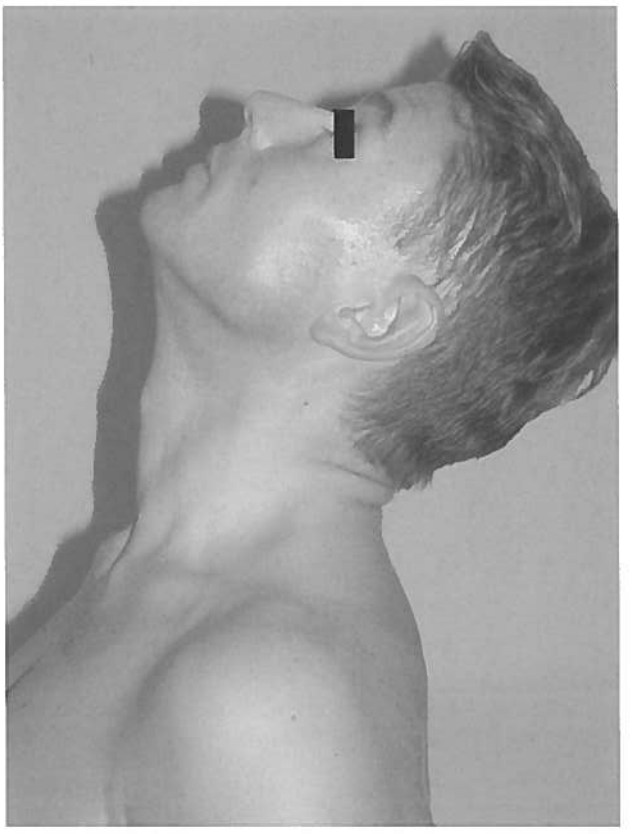


Fig. 3b. Postoperative: patient can rotate his neck to the left



a



b

Fig. 4a and b. Hyperextension and rotation of the neck possible. Unavoidable visible scars

injury. Scar tissue that replaces the muscle transforms the muscle into a fibrous cord, which cannot lengthen with the growing neck in a child, resulting in a shortened muscle [8, 9]. Craniofacial changes are often associated with CMT [5].

Most cases of CMT resolve completely either spontaneously within months after birth or with conservative measures initiated early such as gentle controlled passive manual stretching exercises on the affected side and active muscle-strengthening exercises on the contralateral side, which eliminate the initial deficit in the rotation of the neck and correct the head tilt. Celayir [10] reports that in all 45 infants, 15–120 days old at first presentation, with whom intensive passive stretching exercises were carried out, symptoms completely resolved and there was no need for surgery. According to Porter and Blount [8], more than 70 percent of patients will respond to a conservative approach [2]. If there is significant facial plagiocephaly, especially involving the frontal and orbital areas, a concomitant moulding helmet is often prescribed.

If the patient is refractory to these measures in terms of achieving full range of motion of the neck and having significant facial plagiocephaly, some improvement may be achieved by endoscopic release [11, 12] of the affected sternocleidomastoid muscle before age 1. However, there are some reports of good results in children as old as 12 [13].

In most cases, surgery will result in achieving acceptable range of motion [9]. Most surgical corrections reported in the literature refer to children [14], with very few such interventions in adults. Cheng et al. [2] report that for patients who were 10 years or older at the time of surgery, 63.6% had excellent results and 81.8% had good to excellent results, indicating the benefit of surgery even in the late cases. The oldest reported case we know of is in a patient 22 years old [14]. As our case shows, even if facial asymmetry can no longer be corrected, surgery can help in improving quality of life by restoring range of motion of the neck and possibly preventing further progressive skeletal deterioration.

If left untreated, the consequences are craniofacial deformity with ipsilateral flattening of the face, plagiocephaly, with one side of the face being flat, the skull oblong and not round, and hemihypoplasia, with the face becoming shorter from above downwards and wider from side to side on the affected side [5]. Scoliosis, upper cervical and thoracic, resulting from contracture of the cervical fascia and the scalene muscle and atrophy of the ipsilateral trapezius muscle may also occur. Hip dysplasia is also often present [15].

The standard surgical technique to deal with CMT is tenotomy. Stassen and Kerawala [16] report that the technique they used, which combines subperiosteal lengthening of the sternocleidomastoid muscle at its mastoid insertion, and division of lower fibrotic bands, will result in minimal postoperative fibrosis. They claim that as the sternocleidomastoid muscle is reattached lower down on the mastoid process, the lengthening of the muscle is stable, because the tendency to fibrosis and shortening is minimized and that comparison of the results with previous series shows that this technique provides immediate benefit and good long-term results.

The incision chosen must, as far as possible, be one that leaves no unsightly hypertrophic scars, but one that permits at the same time adequate access to and proper visualisation of the muscle and neurovascular structures in the region so as to avoid causing iatrogenic injuries, primarily to the spinal accessory nerve. Skin incisions immediately adjacent to clavicle are not often chosen because ugly scars may be caused. Inferior open tenotomy of the SCM is one possible approach, in which a single transverse incision is made along the skin folds of the neck 1.5 cm proximal to the clavicle, the portion of the distal sternocleidomastoid muscle is released from its attachments to the sternal and clavicular heads and fascia and any other structures that may be restricting motion are also resected. Such an approach offers adequate visualization and results in imperceptible scars. Uncommonly, distal resection is insufficient and proximal release of the SCM muscle is needed. The other possibility is bipolar Z plasty to release and lengthen the SCM muscle. Here 2 incisions are made, one below the ear on the affected side and another along the collarbone. SCM muscle is cut apart in a Z cut and then reattached.

To avoid noticeable scars, endoscopic surgery can be considered. It has been reported that complete muscular release of the SCM muscle can be achieved with this technique, while at the same time the spinal accessory nerve can be preserved because of adequate exposure of the region [11, 12].

Conclusion

Surgical sectioning of the SCM muscle should be considered even in adults with irreversible facial and skeletal deformities, in order to improve quality of life by restoring some degree of range of rotation of the neck, even if postoperative residual head tilt may persist. Postoperatively, it is advised to place the patient in traction for 24 hours and to wear soft cervical collar or braces for the neck for several months. At the same time, physical therapy and appropriate exercises must be carried out. In pre-puberty patients, long-term follow up is required because of the possibility of recurrence or reappearance of the disease during periods of heightened growth.

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