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Case Report

Triple intervention in congenital wry neck: case report

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ABSTRACT

Wry neck or congenital muscular torticollis is consequent to shortened or contracted sternocleidomastoid muscle (SCM). A painless, retracted sternocleidomastoid muscle is the characteristic feature which results in tugging the neck towards affected side, and draws the shoulder upward and forces the chin in the opposite direction. The incidence is between 0.4 to 1.9%. A 17 years old lady with torticollis since childhood is being discussed in detail with the steps of surgical intervention; at three sites to attain a sternocleidomastoid release and resistance free neck and thus a free head movement. This was followed by application of cervical collar, active physiotherapy and postural exercises. The ignorance and improper advice during childhood had led to this marked deformity at adolescence.

Keywords: Wry neck, Torticollis, Congenital, Z plasty, Bipolar, Tenotomy

INTRODUCTION

Muscular torticollis or wry neck is a rare condition with an incidence between 0.4 to 1.9%. A congenital or acquired entity, manifesting as a typical cervico-facial deformity with a head tilt towards the ipsilateral shoulder tip and- a neck twist with facial deviation towards the contra lateral side.[1] Imaging and histopathological analysis has supplemented the clinical diagnosis of this deformity. Muscular atrophy of the sterno-cleido-mastoid muscle heads and interstitial fibrosis is noted on MRI and histopathology respectively.[2]

A lady with congenital muscular torticollis presented at the age of 17 years and was treated by sternocleidomastoid muscle release followed by Z-pasty, anti-inflammatory medication and other measures.

CASE REPORT

A 17 years old young lady presented with inability to look towards the right shoulder. On examination she was moderately built with a change of physique due to shortening of the neck and with a raised shoulder on right side. She showed inclination of the head towards right side with a raised chin and face rotated towards opposite left side. The clavicular head was prominent, shortened, firm and taut on palpation, and thereby the range of neck movement was markedly reduced (Figure 1 and 2).

A diagnosis of congenital muscular torticollis or wry neck was made during her childhood and she had undergone multiple physical therapy sessions as a part of rehabilitation program, without much improvement. Ignorance and improper guidance were the reason behind delay in surgical intervention. Detailed history revealed

no other congenital abnormality at the time of birth, nor any history of any infection.

Figure 1: Pre-op limited neck movements.

Figure 2: Pre-op taut clavicle head of sternocleidomastoid muscle.

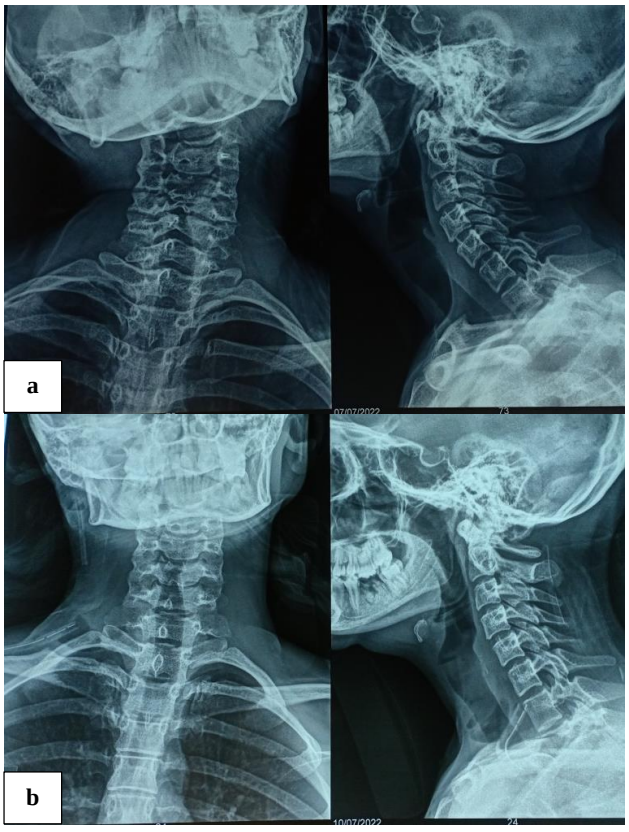

Figure 3: (a) Pre-op radiographs neck; (b) post-op radiographs neck.

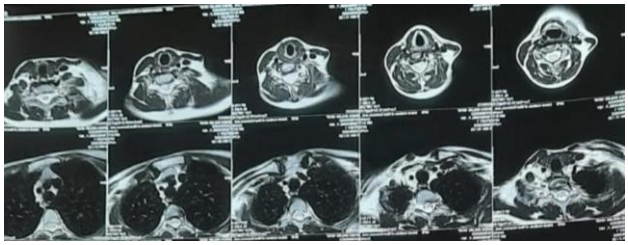

Figure 4: Pre-op Mir scan neck showed a fibrous band replacing the clavicular head of sterno-cleidomastoid muscle on the right side.

Figure 5: Exposure of clavicular head.

Figure 6: Tenotomy of clavicular head.

Figure 7: Exposure of mastoid attachment prior to division.

Figure 8: Z-plasty of sternal head.

Figure 9: Post-op free neck movements.

Evaluation at the paediatric and ophthalmological clinics was undertaken to detect any co-existing abnormality, syndromal or otherwise.

Preoperatively chest X-ray and cervical X-ray were grossly normal showing normal development of cervical vertebrae (Figure 3).

Pre-op MRI scan neck showed a fibrous band replacing the clavicular head of SCM on the right side (Figure 4).

Three step intervention was planned on the SCM: step I, at the clavicular head; step 2, at the mastoid insertion; step 3, at the sternal head.

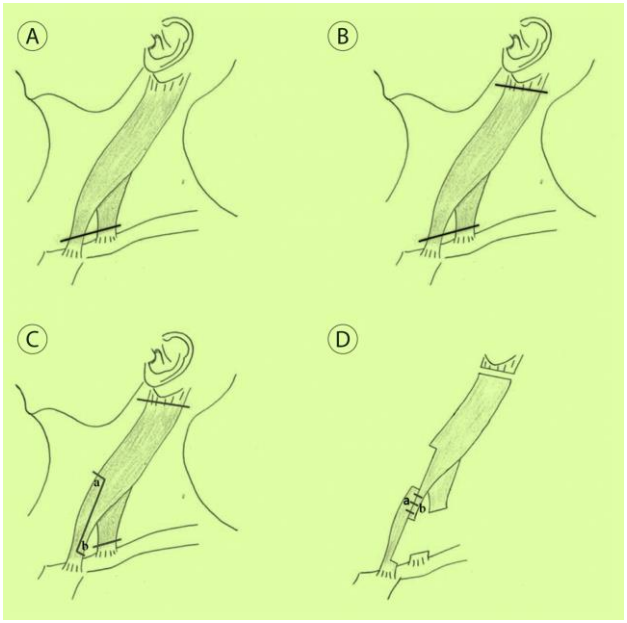

Figure 10: Schematic: three operations for congenital muscular torticollis; A: unipolar release; B: bipolar release; C and D: bipolar release with Z-plasty.

The patient was placed supine with a hyper extension of the neck, on the affected side (right), which was stretched and the head rotated towards opposite side. A 1.5-2 cm transversely placed incision was given above the level of the clavicle. The subplatysmal plane was dissected and thereby the clavicular head of the sternomastoid was delineated (Figure 5). The tendinous clavicular attachment of the muscle was divided, keeping in view and remaining distal to both, the visible bluish appearing internal jugular vein in the carotid sheath and the sternoclavicular joint (Figure 6). The mastoid process was palpated and a transverse 1.5 cm incision was placed over it to expose the attachment of sternocleidomastoid to the postero-lateral surface of the mastoid. This insertion at the mastoid was divided taking care to identify and protect the underlying neurovascular bundle (Figure 7). To free the lower sternal insertion a blunt dissection was carried out and to divide the muscle, a transverse and a longitudinal saggittal incision was given, beginning at the sternal end and ascending upwards. The neck was rotated towards the effected side first and then to the opposite side and palpable taut fibrous bands in the supraclavicular fossa were identified and cut so as to attain a free movement of the neck. The sliding Z-plasty procedure was completed and resected muscle ends were sutured in a released position (Figure 8). Haemostasis was attained. Subcutaneous tissue and skin were closed in 2 layers over an in dwelling drain. The patient was extubated

comfortably in post-op period and a prefabricated/readymade cervical collar was utilized to maintain the neck in normal neutral anatomical position. Drain was removed on the second day. On post op day 4 physiotherapy which included neck strengthening exercises were started (Figure 9).

DISCUSSION

Torticollis term is a Latin word which stands for a twisted neck.[3] A muscular torticollis is consequent to a shortened or excessively contracted thick muscle of the neck, the SCM. Incidence of this deformity ranges between 0.4 and 1.9% with a predominance in the male gender. With an unclear aetiology, global literature abounds with varied theories, suggestive of the likely cause.[4,5] Injury during child birth with a hematoma within the SCM that later heals with fibrosis is the most accepted concept.[3] A prenatal aetiology too is proposed. With evidence of coexisting positioning disorders such like talipes as well as cleft palate.[4] Moreover intrauterine head positioning with a tendency to traumatise the sternomastoid muscle and a resultant compartment syndrome is another theory.

Hereditary, neurogenic, ischaemic and infective aetiologies affecting the neuro-muscular components of the SCM too have been considered.[2,5,6] The diagnosis is usually by physical examination and radiographic imaging 7 congenital muscular torticollis individuals with a twisted neck, exhibit often a compensatory effect in the vertebral column of the neck and the thorax. This usually is a cervical and thoracic scoliotic deformity. Tumours of the posterior cranial fossa, Arnold Chiari malformation and cervical spine tumour effect the muscle innervations and lead to torticollis. These are the common causes of a neurological torticollis, with a rotatory cervical instability.

It is a benign condition which can be treated if diagnosed early and treatment instituted.

An entity known as plagiocephaly is a torticollis sequence which follows persistent contraction of neck muscle. It is also referred to as asymmetric malformation secondary to fusion of one half of the coronal suture.[8] Congenital wry neck associated anomalies are ptosis of eyelids, webbing of the neck and the axilla, micrognathia, cleft palate and scoliosis.[9]

Therapeutic intervention is twofold. The conservative ‘muscle stretch’ exercises and the surgical ‘muscle lengthening’, procedures. In the earlier diagnosed, with minimal deformity and post surgical muscle lengthening the conservative muscle stretching procedures are undertaken, according to a daily session to lengthen the sternomastoid. Surgical intervention depend on the extent of deformity and muscle contracture with limited unipolar release at the sternoclavicular origin or a bipolar open tenotomy, a bipolar release, transection of the middle of the muscle, Z-plasties on the attachments of the sternal muscle, and complete excision of the muscle as the case maybe.[3-5,10] Schematic representation of the three popular operations for congenital muscular torticollis is shown in Figure 10. Usually the overlying and enveloping deep cervical fascia too has contracted and necessitates division to complete the release and achieve full range of neck movement.[5] Contraction of this deep cervical fascia is the likely cause of craniofacial asymmetry rather than positional moulding of open cranial sutures arising from the tilt of the head, or deformation from the pull of the shortened muscle.[11]

In our patient we had undertaken a three step procedure. A bipolar release and a sliding Z-plasty to get a full movement of the neck on table, to be followed by application of a cervical brace during the period of healing for three works, followed by cervical active physiotherapy, to retain the muscle length so achieved.

CONCLUSION

Wry neck necessitates prompt diagnosis and intervention. Triple procedures on the SCM, the bipolar release, Z-plasty and contracted cervical fascia release followed by active cervical physiotherapy result in best outcome.

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