CASE REPORT

Congenital Hypertonia of the Temporalis Leading to Trismus Since Birth

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Abstract Congenital trismus is quite rare especially when its etiology is not the usual. In our case report an 8 year old female patient with no history of forcep delivery, no history of trauma or infection and a non syndromic presents with trismus since birth. After thorough examination we could conclude that the cause is due to hypertonia of the temporalis muscle and its etiology is discussed.

Keywords Trismus · Congenital hypertonia · Congenital hypoplasia · Temporalis muscle · Coronoid hyperplasia

Introduction

The word ‘Trismus’ originated from the Greek word ‘Trismos’, means reduced opening of the jaws. There are various causes for trismus like intra-articular causes such as internal derangement of temporomandibular joint (TMJ), inflammatory arthritis etc.; and extra-articular causes such as tetanus, tetany, oral submucous fibrosis, myositis ossificans, coronoid hyperplasia or syndromic like pseudocamptodactyly.

Case Report

An 8 year old female patient reported to our hospital with a complaint of limited mouth opening. Her parents revealed that their child suffered from limited mouth opening since birth. They also said that the condition increased gradually leading to a limited mouth opening of 7 mm (Fig. 1). They give no history of forcep delivery, no history of trauma or infection. The patient’s medical records showed suction and deglutition difficulty during feeding, failure to thrive and recurrent episodes of choking and aspiration pneumonia. The mother gave a history of being diagnosed of polyhydramnios in her third trimester. On thorough examination the patient had a normal growing mandible, CT imaging revealed normal temporomandibular joint space and normal articular disc (Fig. 2). The only positive features that the patient exhibited were bilateral coronoid hyperplasia and bilateral hypoplasia of the temporalis muscle (Fig. 3). It was also clear that the coronoid process was not causing any mechanical obstruction. Bilateral coronoidectomy was done along with buccal fat pad interposed above the osteotomized site. Immediate post operative mouth opening of 22 mm was achieved (Fig. 1). The patient is on regular follow-up and active physiotherapy.

Discussion

In our literature search we could find only two reported cases of congenital trismus associated with generalized hypotonia of skeletal muscles reported by Puche et al. in 2012. Both their reported cases were male infants with abnormal facial phenotypes in the context of Kabuki syndrome and Penae Shokeir phenotype and X-linked myotubular myopathy. CT facial bone inferred coronoid

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Fig. 1 Pre-operative mouth-opening of 7 mm and post operative mouth-opening of 22 mm

Fig. 2 CT sagittal section of the *left* and *right* TMJ showing normal joint architecture and joint space. *Arrow* pointing to the elongated coronoid process

Fig. 3 3-D CT with soft tissue recon images showing bilateral temporalis muscle hypotonia

hyperplasia with normal temporomandibular joint. Correlating the generalized hypotonia of skeletal muscles in their reported case they hypothesized that the etiology of coronoid overgrowth and trismus due to lack of fetal mandibular movements and deglutition as a result of congenital hypotonia may lead to relative hyperactivity of the

temporalis muscle that is not counterbalanced by the infra and suprahyoid muscles [1].

Gupta and Prasad [2] reported a case of congenital hypoplasia of depressor angularis oris muscle but the etiology was not clear.

This makes our reported case even rarer as we report a female patient with bilateral hypoplasia of temporalis muscle which is hypertonic, not associated with any syndrome causing coronoid hyperplasia and trismus. Analyzing the CT image, bilateral elongated coronoid process is not the cause for trismus as it is not long enough to cause mechanical obstruction.

Although there is another proposed hypothesis by Rowe [3] based on the belief that the disorder tends to develop around puberty however there is no biological support for this. Some authors believe coronoid hyperplasia could be reactive to internal derangement of the TMJ which does not hold well in our case [4–6]. Genetic inheritance and syndromic associations have also been suggested [6, 7].

Compiling and correlating the hypothesis of Abadie et al., Heude et al. and Jubb et al. we propose the following hypothesis in our case. The selective hypoplasia of the temporalis bilaterally can be related to the neuro-embryological hypothesis as poor differentiation of cephalic myogenic mesodermal which are responsive for the future formation of muscles of mastication [8]. Transient dysfunction of the brainstem region from 18th to 24th week of intrauterine life leading to failure of rhythmic fetal reflex of suckling, swallowing and fetal mandibular movements which coincides with the mother's medical history of polyhydramnios [9]. Puche et al. [1] documented a case report where the mother diagnosed with polyhydramnios associated with intrauterine growth retardation of the fetus and abnormal deglutition and fetal mandibular movements. Since the present cardiorespiratory, pharyngeal and laryngeal functions of the child are normal we assumed that the brain stem functions resumed later. Because of the absence of fetal mandibular movements of the fetus the normal equilibrium is lost leading to hypertonia of the temporalis muscle that is not balanced by suprahyoid and infrahyoid muscles. In addition the prolonged absence of mandibular movements reduces the stretch length of the tension in the muscle while at the same time it is stimulated repeatedly through its nerve conduction causing shortening of the muscle fiber length, reduction in weight and bulk which is related to adaptational shortening of the muscle fibers [10]. This is due to reduction in the sarcomeres thus causing a state of prolonged increased hypertonia of the hypoplastic temporalis in our reported case leading to coronoid hyperplasia and trismus since birth.

Treatment implies coronoidectomy and active physiotherapy. An intraoral approach offers enough space for coronoid resection and stripping of the temporalis muscle

insertion avoids external scars and minimizes the risk of facial nerve injury. Extraoral access (through a coronal incision) should be reserved for cases with drastic mouth aperture limitation, concomitant TMJ disease requiring surgery or when the presence of osteophytes on the zygomatic arch would make tendon stripping difficult and hence interfere with complete coronoid process resection [11].

In our patient, we have used buccal fat pad to interposition after the coronoidectomy to prevent fibrosis and fusion. Postoperative physical therapy is absolutely essential. A dynamic physiotherapy program must start as early as possible and continue for 12 months. Different devices (bite blocks, removable activator appliances, spatulas, springs, tweezers, splints, wedges) can be used. In infants and young children, initial sessions may require intravenous analgesia and sedation.

Compliance with Ethical Standards

Conflict of interest The authors declare no conflicts of interest and no funding.

Informed Consent Informed consent was obtained from the participant.

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