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Botulinum-A Toxin Application for the Treatment of Asymmetric Crying Facies

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An infant with a face symmetrical at rest, but one corner of the mouth does not pull downward and outward symmetrically with the other while crying and smiling, is said to have an "asymmetric crying facies" (ACF). The etiology of this asymmetry is congenital absence or hypoplasia of the depressor anguli oris muscle (DAOM) at one corner of the mouth. Its associations with major congenital anomalies, most commonly in the cardiovascular system, have been reported. In this report, a six-year-old boy with asymmetric crying facies and its treatment with botulinum-A toxin (BOTOX) is presented. To our knowledge, the treatment of ACF with BOTOX has not been reported yet.
Key Words: Asymmetric Crying Facies; Botulinum-A Toxin; Child.

Asimetrik Ağlayan Yüz Tedavisinde Botulinum-A Toksin Uygulaması

"Asimetrik ağlayan yüz" istirahat esnasında simetrik olan yüzün, ağlama ya da gülme sırasında ağız köşesinin aşağı ve dışa hareketinin kısıtlı olması ya da hiç olmamasıdır. Asimetrinin etiyolojisinde tek taraflı depresör anguli oris kasının konjenital yokluğu veya hipoplazisi vardır. Major konjenital anomalilerle, en çok da kardiovasküler sistemle birlikteliği bildirilmiştir. Bu yazıda asimetrik ağlayan yüzü olan 6 yaşındaki bir erkek çocuğu ve botulinum-A toksiniyle (BOTOKS) tedavisi sunulmaktadır. Asimetrik ağlayan yüzün BOTOKS'la tedavisi henüz bildirilmemiştir.
Anahtar Sözcükler: Asimetrik Ağlayan Yüz; Botulinum-A Toksini; Çocuk.

Introduction

An infant with a face appearing symmetrical at rest, but one corner of the mouth does not pull downward and outward symmetrically when crying and smiling is said to have an "asymmetric crying facies" (ACF). The cause of this facial asymmetry is the congenital absence or hypoplasia of the depressor anguli oris muscle (DAOM) at one corner of the mouth.[1,2] This minor facial defect may be associated with some major congenital anomalies, most commonly in the cardiovascular system and less frequently involving the genitourinary, musculoskeletal, respiratory, gastrointestinal, endocrine systems, and rarely the central nervous system.[1]

Genetic inheritance is heterogeneous. It may be an autosomal dominant trait, occurring sporadically or can be seen as deletions in chromosome 22q11,2.[3] Facial asymmetry is better noticeable in early ages while the distinction will be little with increasing age because of exerting the compensation mechanisms. In older children, conscious avoidance from the mimics can be hidden by the asymmetry. Parents consult their child to physicians because of anxiety over possible associated neurologic deficits and concern with the lack of clinical improvement in the first few months of life.[2] In history; eye closure, forehead wrinkling, and sucking are normal since birth but, this disorder appears especially crying.

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Case

A 6-year-old boy was consulted with a facial asymmetry on crying or smiling. In history; after an uncomplicated vaginal delivery, this defect had been noted by her parents, only on crying or smiling. Otherwise, he was healthy and no improvement was seen in subsequent months. Any maternal use of medication or antepartum illness were not reported. There were no parental consanguinity and positive family history. On physical examination; the face of child was showing symmetrical at rest (Figure 1).

When he smiled, cried, or grimaced, the right corner of the mouth drew downward and outward, while the left corner did not. Extra-ocular movements, eyelid closure, nasolabial fold depths, and forehead elevation were intact and symmetric (Figure 2). Other system examinations were normal.

Laboratory investigations, such as complete blood count, routine blood chemistry and urinalysis were

normal. Facial electromyography showed an intact facial nerve on the affected side. Lung and spine graphics, abdominal ultrasonographic examination, abdomen CT, brain MRI, ECHO and EEG were all within normal limits.

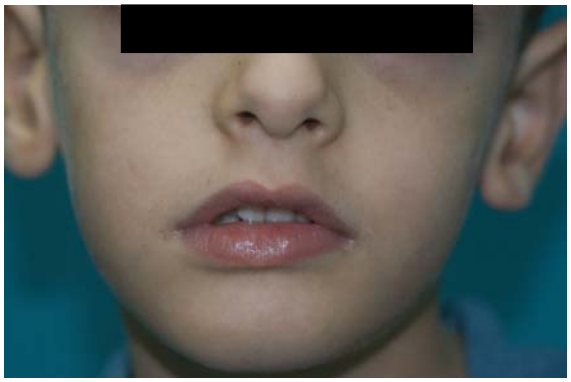

Figure 1. Photograph of the child showing symmetrical face at rest.

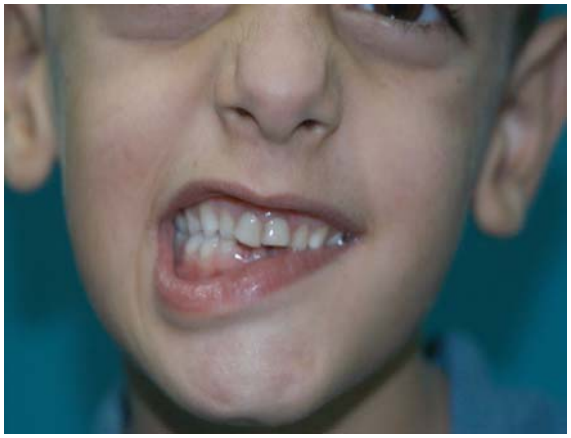

Figure 2. Photograph of child showing asymmetrical face.

Patient and his family had been getting sick and tired off the social and psychological trauma of the environment. Following clinical and laboratory tests, the case was diagnosed as "asymmetric crying facies" with clinical features and normal EMG findings. He had an isolated DAOM hypoplasia or aplasia because of no determination of other system anomalies. To solve this problem in a non-invasive way we planned to use botulinum-A toxin, which is reversible in six months and making the muscle atrophic after three or four applications. We applied the agent into subdermal area as 15 units of botulinum-A toxin on the intact side. This treatment is a little expensive but non-invasive and is a reversible technique, and provided a prominent improvement in the facial asymmetry (Figure 3).

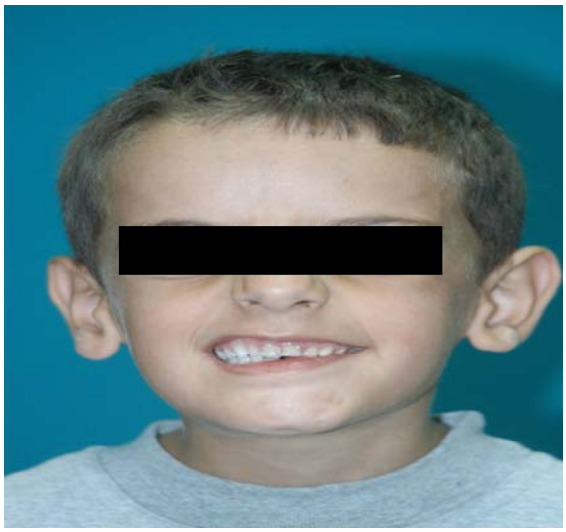

Figure 3. Prominent improvement in the facial asymmetry on the BOTOX application after fifteen days.

Discussion

DAOM hypoplasia or aplasia is solitary in 22% of the cases.[1] Congenital heart defects, musculoskeletal and genitourinary system anomalies appear more than eight times in the cases of ACF.[4] Our case had isolate DAOM aplasia or hypoplasia but no other system anomaly.

The etiology of ACF is still unexplained. In the present patient; history, physical examination, and laboratory investigations gave no clues to the etiology of ACF. The ACF may be familial in some cases and a genetic background may be considered. Autosomal dominant inheritance has been suggested in some families with ACF and, chromosome 22q11 microdeletions and some other rare chromosomal abnormalities have been associated with this facial defect.[5,6] An autosomal dominant trait has been shown in the cardiofacial syndrome.[7] In the patients with ACF, a 22q11,2 deletion has also been detected. In addition, this deletion appeared in the velocardiofacial syndrome, DiGeorge syndrome, and conotruncal anomaly face syndrome.[3,6,8]

Asymmetry of facial movements in a newborn infant suggests seventh nerve palsy due to birth trauma inflicted by forceps blades or pressure against the maternal sacrum, intracranial hemorrhage, congenital maldevelopments of the seventh cranial nerve nucleus, the nerve itself, and surrounding soft tissues.[2] ACF is diagnosed with facial asymmetry appears only on smiling or crying, however nasolabial fold depth, eye closure, and forehead wrinkling are symmetrical and

normal; conduction times and nerve excitability studies are normal.[2]

In these children, it has been revealed that the cosmetic defect lessens with increasing age. It appears as smiling and function of the risorius and other facial muscles come to dominate the child's repertory of facial expression.[2] In later childhood, the imbalance is chiefly seen in crying and grimacing. It is likely that in older children, conscious avoidance of such movements would lead to a very little observable asymmetry.2 Hoefnagel and Penry[9] have considered in these patients the possibility of surgical paralysis of the depressor muscles of the intact side by neurectomy in order to restore symmetry of facial expression. Their cases had no apparent difficulties in eating or speaking during longtime-follow-up. The BOTOX application to intact side in these patients is less invasive and a new treatment tecnique. BOTOX has resulted in significant advances in neurologically based disorders. Clostridium botulinum, an anaerobic bacterium, yields seven antigenically distinct toxin: A, B, C, D, E, F, and G. These are potent neuroparalytic agents, which inhibit the release of acetylcholine at the neuromuscular junction. Synthesis and storage of acetylcholine is not affected by BOTOX and the effects are temporary. The toxin penetrates the endosome membrane into the cytosol where the secretion of acetylcholine is blocked. Type A is thought to have a greater anticholinergic effect at the neuromuscular junction, whereas B is thought to have greater effect at the autonomic junction.[10] The duration of BOTOX's action at the neuromuscular junction appears to be approximately 4 - 6 months. Only type A and most recently B are approved for clinical use. We applied BOTOX subcutaneously into the unaffected side of our patient and provided the facial symmetry with no adverse effects.

The patients with asymmetric facies should be evaluated for ACF and the BOTOX treatment can be applied successfully. BOTOX application offers us an easy, noninvasive, reversible and less psychologically traumatic treatment option. The leading disadvantages of BOTOX are little expensiveness, short period of effect, and reversibility.

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