Asymmetric Crying Facies with a Couple of Primary Mandibular Central Incisor and 22q11 Deletion

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Facial asymmetry present only on crying has been described as a seperate entity and termed asymmetric crying facies. The cause of the facial asymmetry in this disorder is congenital absence or hypoplasia of the depressor anguli oris muscle at the corner of the mouth.
This defect is associated at times with major congenital anomalies, most commonly in the cardiovascular system. Chromosome 22q11 microdeletions in cases with ACF have been reported. We report a newborn infant who had ACF associated with a couple of primary mandibular central incisor teeth and chromosome 22q11 microdeletion. This clinical sign in association with ACF has not been previously described.
Keywords: *asymmetric crying facies; chromosome 22q11 deletion; natal teeth; neonate*
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INTRODUCTION

Hypoplasia or agenesis of the depressor anguli oris muscle on one side of the mouth is a congenital anomaly causing asymmetric crying facies (ACF). This defect is associated at times with major congenital anomalies, most commonly in the cardiovascular system. Beside cardiac anomalies; cervicofacial, urogenital, musculoskeletal, respiratory and endocrine defects have been also described.[1-5] Chromosome 22q11 microdeletions in cases with ACF have been reported.[6,7]

A couple of median mandibular central incisor is a rare dental anomaly, which may be an isolated occurrence or associated with congenital nasal airway abnormalities or DiGeorge syndrome.[8] We report a newborn infant who had ACF associated with a couple of primary mandibular central incisor teeth and chromosome 22q11 microdeletion. This clinical sign in association with ACF has not been previously described.

Case Report

A two-day old female infant with a diagnosis of facial nerve palsy was referred to our neonatal intensive care unit (NICU). The baby was born at term by cesarean section with a birth weight of 3300g. There was no consanguinity between the parents. Physical examination revealed droop of the left corner of the mouth while crying. The mouth of the baby was pulled down and to the left while on crying (Figure 1a). Her face was symmetrical at rest. There were also a couple of lower primary central incisor on her mouth. These teeth were palpable but covered with gingival tissue (Figure 1b). Forehead wrinkling, eye closure and nasolabial fold depth were intact and equal on both sides. The baby was sucking well without drooling from either corner of the mouth. These findings prompted us to differentiate this disorder from facial nerve palsy. On physical examination the baby had no respiratory stress but a systolic ejection murmur was heard at the left sternal border. X-ray examination of the chest revealed normal heart size and cardio-thoracic index and normal thymic shadow. Echocardiography established the diagnosis of atrial septal defect.

Laboratory investigations including hemoglobin, hematocrit, acid-base values, glucose and electrolytes, renal and liver function tests and urine analysis were normal. Fluorescent in situ hybridization testing of the baby provided genetic confirmation of chromosome 22q11 deletion. The baby was consulted to the dental department and teeth extraction has been performed as there was a risk of aspiration during suckling.

DISCUSSION

The overall rate of anomalies including major malformations, mild anomalies and deformations associated with ACF varied ranging from 5%- 70%.[2-5] Infants with ACF have a 3.5 fold higher risk of major malformations, compared with the general population.[3] Associations of this minor facial defect with major congenital anomalies have been reported, most commonly in the cardiovascular system and less frequently

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involving the genitaurinary, musculoskelatal, cervicofacial, respiratory and rarely the central nervous system.[1–7] Therefore Lahat *et al* suggested that ACF can be used as an index of other congenital anomalies.[2] Many mild anomalies including preauricular tag, hypertelorism, dermoid cyst, sternal cleft, anal tag, pilonidal sinus, strawberry hemangioma, hiperpigmented nevus, congenital telangiectasia, accessory nipple, clinodactyly, polydactyly, umblical and inguinal hernia were previously reported within ACF.[2,3,5] In our patient existence of incisor teeth was detected and this defect can be accepted as an additional anomaly in association with ACF.

The incidence of natal teeth is 1:2,000 to 1:3,000 live births. Natal teeth usually occur in pairs and the most commonly affected teeth are the lower primary central incisors.[10] They are present in 2% of infants with unilateral cleft lip and palate and 10% of infants with bilateral cleft lip and palate.[11] Natal teeth have been reported in association with syndromes such as Ellis-van Creveld, Hallermann-Streiff, Sotos, Meckel-Gruber and Pierre Robin.[12,13] Hence, the presence of natal teeth and ACF has not been described.

The prevalence of ACF reported in recent studies was 3-8 *per* 1,000 births.[3,4] Additionally, the incidence of microdeletion in chromosome 22q11 is 1:4,000–5,000.[14] There is no any report in association with ACF, natal teeth and chromosome 22q11 deletion. The exact etiology is not known but same emryologic fault might have occured in these disorders. Developmental field defect of the facial neural crest tissues, involving the third and fourth pharyngeal arches might have been responsible for this co-association.

Left predominance of depressor anguli oris muscle dysfunction was reported within the range of 54%-83%.[3-5] In our patient it is easily recognizable that the left corner of the mouth drew also left and downward while the right moved little or not at all (Figure 1a). The etiology of this interesting left predominance of ACF is not yet justified. Electromyographic examinations generally show diminished activity of the right depressor anguli oris muscle. Unfortunately, this finding is not a sufficient knowledge to mention the etiology of left predominance.

The incidence of chromosome 22q11 microdeletions in individuals with ACF is not precisely known. Early diagnosis is important for these individuals as they are at increased risk of requiring medical interventions for associated other anomalies.[9]

CONCLUSION

A detailed physical examination and consideration of investigations for chromosome 22q11 deletions should be performed in case of an asymmetric crying face is encountered, while considering a search for occult anomalies.

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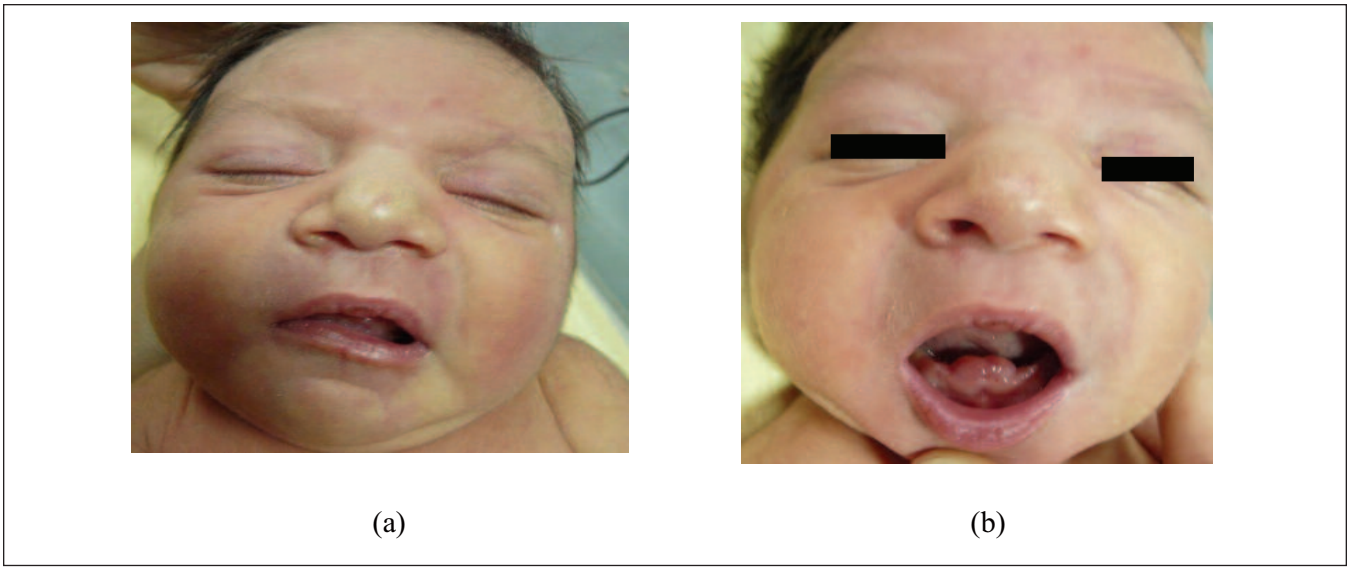

Figure 1. Typical appearence of asymmetric crying facies (a) and a couple of lower central incisor teeth covered with gingival tissue on the mouth (b).

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