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Case Report

CLEFT TONGUE AND PALATE: A RARE CASE PRESENTATION

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ABSTRACT

The tongue develops during the fourth week of intrauterine life, originating from a median swelling, the tuberculum impar on the floor of the pharynx and two lateral lingual swellings joining this central structure. These lateral lingual structures grow rapidly to cover the tuberculum impar to form the anterior two-thirds of the tongue. When this process is disturbed, tip of the tongue is divided longitudinally for a certain distance giving rise to cleft tongue/bifid tongue. Cleft of the palate and congenital anomaly of the tongue is a rare occurrence. In this case report we present a case of a 11 year boy having a rare combination of cleft tongue with palate.

Keywords: Cleft Palate, Cleft Tongue, Developmental Anomaly, Hypertelorism.

INTRODUCTION

Human growth and development is a meticulously planned and precisely executed process. Even a mild disturbance can have clinically significant manifestations later on. Even today, ancient beliefs and practices override/delay patient's aspiration for seeking treatment as seen in the case report presented here. Cleft tongue in association with various other orofacial abnormalities has been reported, many of which have been linked to various syndromes.. Here, we discuss a case of bifid tongue involving the anterior one-third of tongue, reported in a 11-year-old male patient with cleft palate.

CASE REPORT

A 11 year old boy came to the Vedanta hospital smile train cleft surgery centre of Lanjigarh, Kalahandi, Orissa. On taking history his parents revealed the boy cannot speak properly, having problems eating food.

The facial profile of the patient was concave with hypertelorism and depressed nasal bridge (fig 1 and 2).

On examination the boy was having a cleft and trilobed tongue with a complete cleft palate (fig 3 & 4).

We proposed a two stage surgery of tongue followed by the palate.

But the patients parents changed their mind and decided not to opt for surgery of the tongue and cleft palate.

Thus the patient was discharged and we could only document the characteristics of the anomaly and present it.



Figure 1: Facial profile with hypertelorism and depressed nasal bridge



Figure 2: Lateral view showing midfacial hypoplasia



Figure 3: Cleft tongue.



Figure 4: Cleft Palate

DISCUSSION

The combination of cleft tongue with a palate and a neural anomaly is extremely rare.

The development of the tongue starts at the fourth week of intrauterine life in the floor of the primitive cavity from the first three or four brachial arches¹.

Bartholdson *et al.* described a baby boy with a bifid tongue combined with a cleft palate².

Abnormal/partial/non-fusion of these arches may lead to congenital anomalies of tongue, including bifid tongue. In fact, any process affecting mesenchymal fusion and acting toward the end of fourth week of gestation has been suggested to account for this malformation. They may occur as an isolated entity or a part of clinical syndromes³.

Bifid tongue has been reported in syndromic cases like Opitz G BBB syndrome, oral–facial–digital syndrome type I, Klippel–Feil anomaly and Larsen syndrome⁴⁻⁷. Bifid tongue has also been reported as a rare feature associated with infants of diabetic mother syndrome⁸. Literature also gives a reference of bifid tongue as a complication of tongue piercing⁹. Associations have also been postulated with cleft palate, mandibular cleft, midline palatomandibular bony fusion and cervical vertebrae¹⁰.

Aglossia, syndromic microglossia, macroglossia, accessory tongue, long tongue and cleft or bifid tongue are the commonest occurrences listed in the order of frequency.

Double tongue, formed as a result of a developmental anomaly within a lingual tubercle, has previously been described in

association with a cleft palate (Bartholdson *et al.*, 1991); though no salivary gland component was described), and the tongue (Britto *et al.*¹¹).

Syndromic associations of palatal cleft and tongue lesions have been reported in the orofacial digital syndromes 1 and 2 (Martinot *et al.*, 1994), which are inherited in an X-linked and autosomal recessive fashion, respectively.

Double tongue has been reported without palatal clefting in the same syndrome (Bell, 1971)¹².

Bifid tongue and cleft palate have been reported in the Klippel–Feil syndrome (Widgerow, 1990)¹³, and glossoptosis and cleft palate coexist in the Pierre–Robin sequence, possibly as a result of mandibular hypoplasia and failure of the sagittal growth to cause an increase in volume of the oronasal cavity.

The coexistence of mandibular epulis with the cleft tongue/cleft palate combination is probably sporadic.

Cleft tongue/Diglossia are different names given to cleft tongue.

A mutant gene on the X chromosome with a cleft palate and ankyloglossia phenotype has been identified (Stanier *et al.*, 1993)¹⁴, but this does not preclude an obstructive cause of the palatal cleft.

Coexistence of tongue anomalies with cleft palate have been previously reported as isolated findings in nonsyndromic cases and also as coexisting anomalies in syndromic cases. Minami *et al.* (1995)¹⁵ reported a case of ankyloglossia with a cleft palate. Restriction of movement of the tongue, such as in ankyloglossia interferes with the descent and projection of the tongue in fetal head growth, thus preventing palatal fusion.

Neurotransmitters may also play a role (Zimmerman *et al.*, 1981; Lauder and Zimmerman, 1988)¹⁶.

The cellular signals that mediate this process are complex, involving growth factors (Ferguson *et al.*, 1992; Qiu and Ferguson, 1995) that regulate epithelial gene expression (Sharpe and Ferguson, 1988; Ferguson, 1998^{17,18-19}).

The secondary palate forms above the tongue from the seventh to 10th embryonal weeks, after the primary palate has formed, by the midline fusion of the palatal shelves of the maxilla. Initially, the palatal shelves project obliquely and caudally, growing down the oro nasal cavity alongside the tongue.

In a craniofacial growth spurt, which is predominantly sagittally directed, the tongue displaces anteriorly. Mammalian palate elevation occurs at this time and is a function of the turgor of the interstitium, regulated by the hydration of mesenchymal mucopolysaccharides (Ferguson, 1981). The shelves move into a horizontal position above the dorsum of the tongue and fuse in the midline, thereby delineating and separating the oral and nasal cavities (Diewert and Tait, 1979; Britto *et al.*, 1998)¹¹.

Thus this case which is a congenital case of cleft tongue and palate is a rare and interesting case to report.

We were unable to identify any associated etiopathology, genetic predisposition, or history of pre or postnatal trauma.

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