Case Report / Olgu Sunusu

Non Syndromic Hypoglossia With Mandibular and Palatal Abnormalities

Mandibüler ve Palatal Anormallikle Beraber Sendromik Olmayan Hipoglossi

Shishir Ram Shetty[1], Anusha Rangare[2], Priya Sara Thomas[3], Subhas Babu G[4].

[1] Lecturer, Department of Oral Medicine and Radiology A.B Shetty Memorial Institute of Dental Sciences, Nitte University Mangalore-INDIA

[2] Lecturer, Department of Oral Medicine and Radiology, Century international Institute of Dental Sciences and Research Center, Kasargod, Kerala-INDIA

[3] Department of Dentistry, Father Muller's Medical college and Hospital, Mangalore, Karnataka-INDIA

[4] Professor and Head of the Department, Oral Medicine and Radiology, A.B Shetty Memorial Institute of Dental Sciences, Nitte University Mangalore.-INDIA

Corresponding Author:

Dr. Shishir Ram Shetty
Department of Oral Medicine and Radiology
A.B Shetty Memorial Institute of Dental Sciences, Nitte University
Mangalore
INDIA
Mob- +919986221047

Email:

drshishirshettyomr@yahoo.com

Özet

Dil; konuşma, çiğneme , tatma ve yutma gibi birçok fonksiyona sahip, vücudumuzun en hareketli organıdır. Literatürde dilin sayısız konjenital ve kalıtımsal bozuklukları bildirilmiştir. Mikroglossi olarak da bilinen hipoglossi nadir anomalilerden biridir. Çoğunlukla diğer sistemik anormalliklerle ilişkilidir ancak nadiren izole bir antite olarak bulunur. Dilin gelişmesindeki yokluk düşük mandibüler gelişmeye yol açar. Bu yazının amacı nadir bir izole hipoglossi ve sebep olduğu mikrognatiyi sunmaktır. Hastamızda çok çok nadir görülen hipoglossiye eşlik eden palatal ve faucial pillar anormallikler de vurgulanmıştır.

Anahtar kelimeler: Hipoglossi, mikrognati, palatal anormallikler

Abstract

Tongue is the most mobile organ of the body with multiple functions such as speech, mastication, taste and swallowing. Numerous congenital and developmental disorders of the tongue have been reported in literature. Hypoglossia also known as microglossia is one of the rare anomalies. Usually hypoglossia is associated with many other systemic abnormalities but rarely found as an isolated entity. Lack of tongue development leads to decreased mandibular growth. The aim of this article is to report a rare case of isolated hypoglossia and resultant micrognathia. Presence of palatal and faucial pillar abnormalities with hypoglossia which is extremely rare is also highlighted in our report.

Keywords- Hypoglossia, micrognathia, palatal abnormalities.

Introduction

Hypoglossia or microglossia is one of the rare congenital anamolies of the tongue which may occur as a part of syndrome or as an isolated case (1). Occurrence of hypoglossia with abnormalities of soft palate, anterior and posterior pillars is very rare (2). In a study involving 35 cases of hypoglossia, malformations of the hand and feet were the most commonly associated conditions (3). Lack of tongue growth affects development of jaws, malocclusion, difficulty in speech and swallowing (4). Various syndromes such as Goldenhar syndrome, Moebius syndrome, Aglossia-adactylia Syndrome, Hanhart syndrome, Glossopalatine ankylosis syndrome, Limb deficiency-spleno-gonadal fusion syndrome and Charlie M syndrome have hypoglossia or aglossia as one of their clinical feature (5). The report aims to describe an extremely rare case of hypoglossia associated with micrognathia, palatal and faucial pillar abnormalities.

Case Report

A 42 year-old male reported to us with decayed teeth in the upper and lower jaws. The patient had difficulty in speech and swallowing since childhood due to abnormal small tongue. About 30 years ago the patient had visited an otolaryngologist who suggested that the defect could be surgically corrected but the patient's parents refused operation. He had no other relevant medical history. The patient had a convex facial profile (Figure 1).

No other abnormalities were detected during general physical examination. Intraoral examination revealed presence of multiple root stumps in the maxillary and mandibular arches. Palate was high arched with collapsed maxillary arch (Figure 2). Uvula and faucial pillars were ill-developed. Tongue was hypolastic, around 2 cms in long antero-posteriorly and 1 cm in wide (Figure 3).

Tongue surface was devoid of papilla and movements were restricted in all directions. Lingual frenulum was absent, floor of the mouth was pinkish in appearance and leathery. The mandibular arch was v-shaped. The case was provisionally diagnosed as microglossia associated with micrognathia, palatal and faucial pillar abnormality. Lateral cephalogram (figure 4) and orthopantomagram (figure 5) revealed presence of hypoplastic mandible with reduced ramus height. A through medical examination was carried out by medical specialists aided by blood investigations and echocardiography. No systemic abnormalities were reported after the specialists. The patient was explained about the surgical modalities but he refused to undergo the procedure.

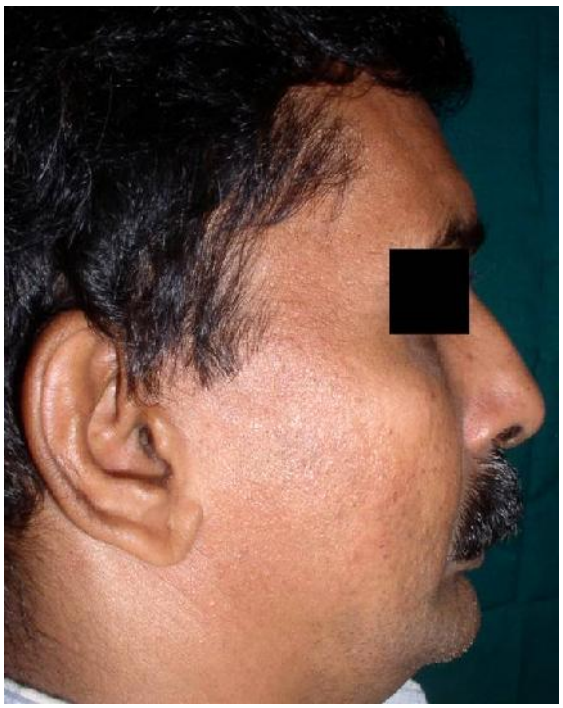

Figure 1: Lateral view of the convex facial profile.

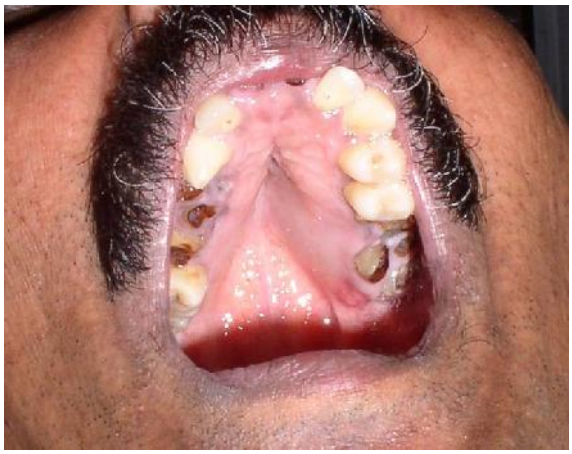

Figure 2: High vault palate with collapsed maxillary arch.

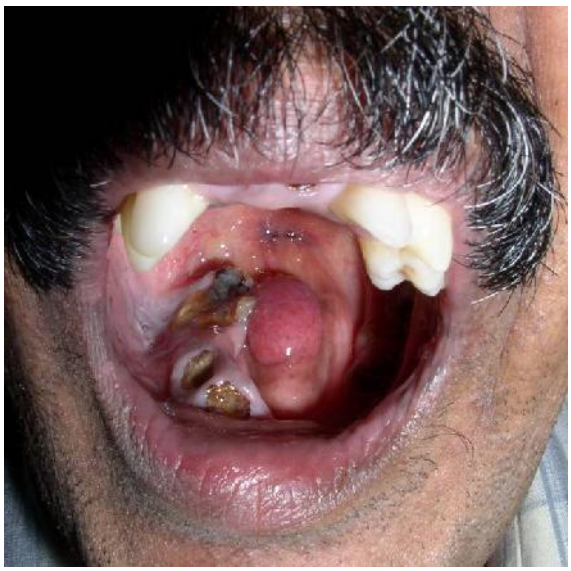

Figure 3: Hypoglossia, pinkish colored floor of the mouth and collapsed mandibular arch.

Discussion

Partial absence of tongue also termed as microglossia is an extremely rare condition, most often associated with limb abnormalities (6). Our case did not feature limb abnormalities, thus can be described as a rarity.

According to Hall's classification, our case can be classified as type IA (7). Various morphological abnormalities like anodontia, 6th and 7th nerve palsy, left hypodactyli and Ventricular septal defects, developmental delay in milestones, hypothyroidism, microcephaly, microsomia, collapse of mandibular arch, congenital absence of mandibular teeth along with persistence of buccopharyngeal membrane (8). Our case did not show any other features mentioned except collapse of mandibular arch. Our patient exhibited collapsed mandibular arch and high arched palate. This could be due to the lack of muscular force of the underdeveloped tongue. Similar features have been reported in rare cases of isolated hypoglossia (9). Our patient was 42 year old and had a history of speech and swallowing difficulties since childhood. Taste sensation was normal but the floor of the mouth was firm and leathery due to abnormal masticatory forces.
Reconstructive surgery is the treatment of choice in these patients along with correction of the malocclusion (2). Our patient did not agree for reconstructive surgery. Speech cannot be improved by reconstruction (8).

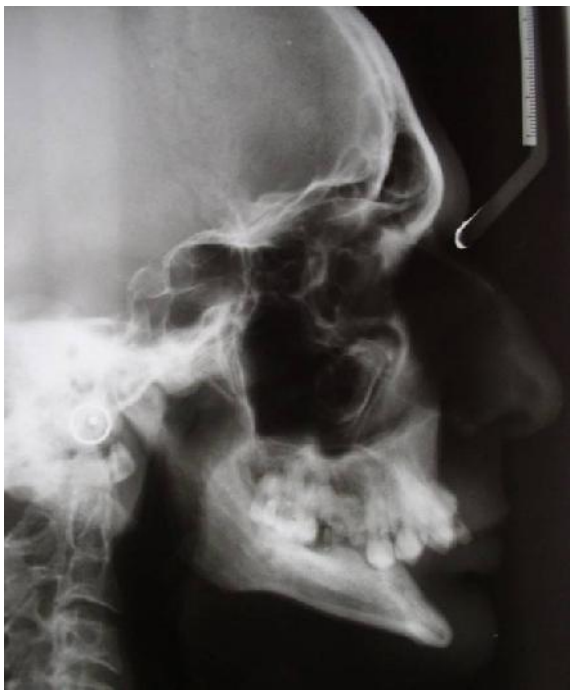

Figure 4: Lateral cephalogram showing prominent chin and short ramus.

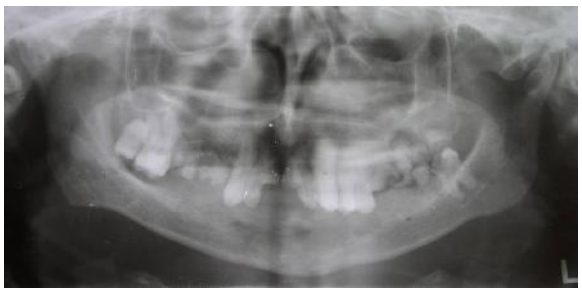

Figure 5: Orthopantomogram showing hypoplasic body and ramus of the mandible with accentuated antegonial notch.

Conclusion

To conclude, case of hypoglossia with underdeveloped mandible is very rare. Most of the cases of hypoglossia have been associated with various craniofacial, limb abnormalities and congenital abnormalities, unlike our case where hypoglossia occurred as an isolated entity.

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