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Review Article

Accessory tongue: Classification and report of a case

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ABSTRACT

Background: Accessory tongue (AT) is a rare congenital anomaly. Due to rarity of AT, various terminologies are applied, including accessory tongue, bifid tongue, double tongue, cleft tongue, and supernumerary tongue. It seems that the anomaly has geographic distribution and most reported cases are from India and Middle East. No comprehensive classification has been introduced yet. So, we present a classification for AT according to review of all papers and documents that we found, and report a 2-month-old male infant with this anomaly.

Contents

1. Introduction

During the fourth week of intrauterine phase, the tongue is originated from the first three or four branchial arches and a median swelling (tuberculum impar) on the floor of pharynx and two lateral lingual swellings joining this central structure [1]. These three swelling are originated from the first pharyngeal arch [2]. Copula or hypobranchial eminence is another median swelling that is formed by mesoderm of the second, third, and part of the fourth branchial arch. It gives rise to posterior part of the tongue [2]. Sometimes, errors in the tongue development may occur which is due to anomalies in lingual tubercle [1,3]. Until yet, 37 cases were reported in literatures. Rarity of this anomaly, results in ill-defined terminology and classification for AT. Developmental abnormality of tongue may be major or minor, single or multiple [4]. So, many different kinds of AT could be happened with various degree of severity and functional disturbance. AT could be isolated (sporadic) or in association with other disorders (syndromic) such as, Opitz G BBB syndrome, Larsen syndrome, short rib syndrome, short rib polydactyly syndrome, oral–facial–digital, median cleft syndrome, Klipple–Feil anomaly, varadi syndrome, oro-maxillary, limb disorder, cleft palate, mandibular cleft, linea alba hernia, hypertelorism, scoliosis, aglossia, microglossia, macroglossia, hemi-glossia, Glossitis Rhombica Mediana, lingual thyroid, long tongue, and ankyloglossia. Sometimes, hamartoma or tumor (teratoma) may happen with AT simultaneously [3–10]. Sign and symptoms are variable in different age groups. Affected neonate may be completely asymptomatic at birth or suffered from breathing difficulties [11], choking [4] or swallowing disturbance [3]. Asymptomatic cases may be missed until childhood or following ages. Trouble in speech [3] may happen later in life in undiagnosed or untreated cases.

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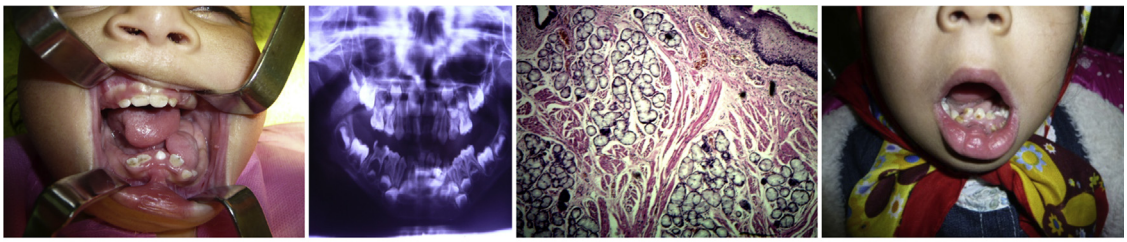

Fig. 1. (A) Complete accessory tongue and cleft mandible. (B) OPG imaging. (C) Accessory tongue which consisted of interlacing bundles of muscular fibers and numerous serous and mucous accessory salivary glands, covered by stratified epithelium (H&E, ×40). (D) Case after 5 years follow-up with good appearance and normal speech.

We report a patient with tongue duplication who was successfully treated with surgical excision and also review the previous cases to reach a novel classification.

2. Report of the case and review article

A three month old female infant, a first child in a family of two, was referred to us due to failure to thrive and tongue and mandible deformity. She had a history of respiratory problem during starting feeding after birth. She hospitalized in another city due to aspiration pneumonia at one and two month old. She was delivered by cesarean section with APGAR score 8–9 and birth weight 3200 g. On admission, her weight, head circumferential, and length were 3500 g, 34 cm, and 57 cm, respectively. Her mother did not have any previous abortion, drug usage during pregnancy or diabetes induced pregnancy. In addition, no family history was found in either maternal or paternal families. In physical examination, an accessory tongue was diagnosed in left lateral side of native tongue which was slightly smaller compared to normal one (Fig. 1A). This caused the mandible to expand and deform. Also, there was evidence for double mandible bone according to OPG imaging (Fig. 1B). Lower lip had some deformity especially in mid line. At first, nasogastric tube and respiratory support were given for patient. With conservative management, breathing got better, and feeding started through nasogastric tube. Patient was discharged with good general condition after one week. Gradually, she tolerated feeding by mouth. Surgical intervention was performed in order to excise the accessory tongue with preservation of native tongue, nerves and salivary glands at age one year. Macroscopic results of accessory tongue demonstrated the resection of 4 cm × 1.8 cm × 1 cm accessory tongue covering by soft mucous membrane. Histopathological examination showed interlacing bundles of muscle fibers and numerous serous and mucous accessory salivary glands scattered throughout the muscle and lamina propria, covered by stratified epithelium and was consistent with normal tongue (Fig. 1C). Postoperative course was uneventful, no complication was observed and patient discharged after 4 days with good condition and ability to feed. We followed the case for 5 years and fortunately, she was growing up normally without any neuro-developmental defect as well as with completely normal speech.

Based on searching keywords (i.e., accessory tongue, cleft mandible, congenital anomaly and pediatric) through various sources like Google scholar, pubmed, and science direct, 34 articles were collected that the oldest and newest are related to 1966 and 2012. Generally, there are 38 cases that listed as following (Table 1).

3. Result

According to this review, we found 37 cases with AT. 62% of cases were female. Associated anomalies were observed in 83% of cases. The most common associated anomaly was cleft palate that was found in 43% of cases. Symptomatic and asymptomatic cases were 37% and 62%, respectively. Speech difficulty was reported in 3 cases with the prevalence of 5% and 2% in dorsal and lateral surfaces, respectively. The most prevalent associated clinical symptom was respiratory difficulties in 18% of cases. Incomplete form of AT was more dominant than complete types (55%). Complete form was mostly asymptomatic and only seen in 15%.

4. Discussion

Despite of lack of genetic pattern in these cases, it seems that the prevalence of AT is more common in female gender. The exact frequency of AT is not clear in the world. Lack of proper data may be due to unavailability of good classification. Accordingly, and during our conducted surveys, we can suggest a suitable classification that helps to distinguish different aspects of this abnormality (Table 2).

AT may be associated with other types of midline clefts that the most common are cleft palate and mandibular cleft. Many reported cases about congenital pathology of tongue are cystic lesions that are not related to AT. It is likely that the cysts of tongue are as a part of duplication of GIT. Given the introduction of AT as tongue duplication in some papers, it makes misunderstanding that AT can also be a subtype of GIT duplication [16–19]. Embryologically, with regard to lack of communication between construction of accessory tongue and GIT, and no reports about the simultaneity of GIT duplication in above cases, it seems that the nature of AT is completely independent and not related to GIT duplication. Because AT is placed in a group of midline defects [8], thus, monitoring the involved cases for these probable anomalies is suggested before surgical excision. The prenatal sonography is popularized now and it is possible to diagnose AT with this modality of fetus evaluation before birth [6,8]. In case of prenatal diagnosis of AT, due to probable respiratory problem just after birth, it is better to be referred mothers to more equipped departments. As polyhydramnios may be coexist with AT, prenatal sonographer must evaluate the fetus for this anomaly in these cases [9]. In postnatal period, evaluation for other midline defects is advised. We checked our case by brain MRI and OPG. No abnormality was seen in brain study despite of conducted survey by Manjila et al. [9] that shows duplication in pituitary glands. It is important to consider diabetic mothers as a predisposing factor in occurrence of AT [2,15,20]. We did not find any history of diabetes in mother of our case. In differential diagnosis, the benign lesions must be considered such as fibroma and haemangioma [1]. Until now, it is not revealed any genetic abnormalities in all the reported cases. Therefore, it seems that geographic distribution may be environmental or caused by other reasons. Rai et al. [2] described that this anomaly is more obvious in the southern coastal belt of India with the prevalence of 0.3%. Moreover, no inheritance pattern or family history has been observed.