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Congenital accessory tongue: a rare case of non syndromic tongue anomaly.

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Abstract:

Background/objectives: Congenital malformations of the tongue are rare and an accessory tongue is extremely uncommon and its incidence is not quoted in literature.

Setting: Department of ENT, Head and Neck Surgery, KVG Medical College, Sullia.

Case report:

Incidental diagnosis of accessory tongue was made in a case of acute tonsillo-pharyngitis while mild symptoms of swallowing discomfort and no speech abnormalities. No other congenital abnormalities were seen in the individual.

Intervention:

simple surgical excision was made and biopsy revealed the lesion to be of an accessory tongue.

Conclusion:

Non syndromic tongue anomalies can be treated with simple surgical correction but syndromic tongue anomalies need a multidisciplinary management.

Introduction:

The tongue develops in the fourth week of intrauterine life, arising from a median swelling, the tuberculum impar on the floor of the pharynx and two lateral swellings joining the central structure.[1] These lateral lingual structures which arise from the proliferation of the mesoderm develop rapidly and gradually cover the tuberculum impar, joining in the midline to form anterior 2/3 of the body of the tongue.[2]

Defects in the development or fusion of these three structures give rise to the embryogenetic malformations.[3] They may occur as an isolated entity or a part of the clinical syndromes.[4] Congenital malformations of the tongue are rare and an accessory tongue is extremely uncommon and its incidence is not quoted in literature.[3] It is seen as a tongue like new growth that can be found in the oral cavity, pharynx, or dorsal portion of the tongue.[3] An accessory tongue is a rare anomaly and only few cases are reported.[5]

Malformations of the tongue, are structural defects, present at birth and occur during embryogenesis.[2]

The most common congenital lingual malformations are Aglossia and microglossia which always occur with other defects and syndromes like Moebius syndrome.[2] Macroglossia is commonly associated with cretinism, Down's syndrome, Hunter's syndrome, Sanfilippo syndrome and other types of mental retardation.[2] Accessory tongue ,long tongue,cleft or bifid tongue are conditions usually associated with orodigitofacial syndrome.[2] Other least common anomalies are glossitis rhombic mediana and lingual thyroid.[2] Double tongue results due to developmental anomaly within a lingual tubercle.[5] We report a patient with an accessory tongue which was excised successfully.

Case report:

A 48 year old middle aged man presented to our OPD for throat pain and mild discomfort while swallowing since 15 days. On examination of the oropharynx a diagnosis of tonsillo- pharyngitis was made. Incidentally a lesion on the dorsum of the tongue was seen at the junction of the anterior 2/3 and the posterior 1/3 just right to the midline. The lesion looked like a small tongue. He did not have dysphagia or speech abnormalities.(fig 1,2).

The anomaly was present since birth but it did not bother him more as it hardly caused difficulty in swallowing or while speaking. On gross examination a 1.5 cm x 1 cm firm polyp like looking lesion was seen on the dorsum of the tongue at the junction of the anterior 2/3 and posterior 1/3. It appeared as a small miniature form of the normal tongue. The mass moved freely with the tongue and free movements were possible in all directions as

the lesion was pedunculated. No free active movements were possible in the lesion.(fig 3)

There was no associated congenital anomalies oro- maxillary or limb disorders. The patient was given oral antibiotics and analgesics for tonsillo-pharyngitis and was reviewed after a week. The acute episode of tonsillo-pharyngitis subsided after a week and the patient was counseled to treating the lesion looking like an accessory tongue. The accessory tongue was excised under local anaesthesia as an OPD procedure. Cautery was not used to reduce the tissue loss for histopathological examination. A single 3-0 vicryl suture was put to stop the small bleeding point.

Histopathological examination confirmed the diagnosis of the accessory tongues composed of fibro-fatty tissues with multi-directional, sometimes striated muscle fibers containing small numerous salivary glands, ecstatic vessels, nerve bundles covered with cuboidal epithelium with fungiform papillae. The patient was followed for 2 months and no recurrence at the site of lesion was seen.

Discussion:

Congenital accessory tongues are rare in occurrence and less than ten case are reported in literature.[3-7] The development of the tongue starts at the fourth week of intrauterine life, from the first three or four brachial arches.[2] Malfusion of these arches may lead to congenital anomaly which may occur as an isolated entity or part of clinical syndromes. [5,6,7]

Tongue anomalies can occur along with cleft palate as a part of syndromic and non syndromic features.[2] Syndromic cases need a multidisciplinary management while non syndromic cases are easier to manage by simple surgical excision as in our case. [2] The syndromic cases associated with other anomalies receive the early attention of parents and physicians, but non-syndromic cases are neglected and usually present late.[5] Non syndromic

tongue anomalies present with dysphagia and speech problems.[8] The earlier can be easily relieved by simple excision and the later by speech therapy.[8] The earlier the treatment better is the results with speech therapy.[8] The important differential diagnosis is benign tumors arising from the dorsal surface of the tongue, like fibroma and haemangiomas. [5]

Other differential diagnosis are oral duplication cysts.[10] A variety of heterotropic mucosa have been reported in association with tongue duplication including gastric, colonic and respiratory mucosa.[11] Oral cavity is a very rare site for duplication. [11] Oral cysts containing heterotropic mucosa has been referred as duplication cyst, choristomatic cyst or heterotropic cyst. [11] Duplication cyst is characterized by a coat of muscle, attachment to some part of GI tract, mucosal lining similar to some part of GI tract.[12] On the contrary, choristomatic cyst or heterotropic cyst need not have a muscle coat.[12] Partial or non fusion of the mesenchymal arches may end up in bifid tongue.[13] Bifid tongue is seen in Optiz G BBB syndrome, oral facial digital syndrome type 1, Klippel Fiel and Larsen syndrome.[14]

Ankyloglossia (tongue tie) ankyloglossia refers to congenital shortness of the lingual frenum or a frenal attachment that extends nearly to the tip of the tongue, binding the tongue to the floor of the mouth and restricting its extention.[15,16] Severe degrees of ankyloglossia often cause a midline mandibular diastema, lingual mandibular periodontal defects, and speech impairment and most of which can be treated by simple frenulotomy.[15,16]

Median rhomboid glossits (central papillary atrophy) is a rounded or roughly lozenge – shaped, raised area that occurs in the midline of the tongue dorsum just anterior to the vallate papillae. [15,16] The affected area is devoid of filiform or other papillae. In recent years, considerable debate has

been centered around the role of chronic candidiasis in median rhomboid glossitis.[17,18,19]

Geographic tongue (benign migratory glossitis) refers to irregularly shaped, reddish areas of de-papillation and thinning of the dorsal tongue epithelium that are usually surrounded by a narrow zone of regenerating papillae.[20] *Fissured (plicated or scrotal tongue) tongue occurs as a normal variant affecting less than 10% of the population and is genetically determined.*[21] *The frequency of fissured tongue is four to five times greater in institutionalized, mentally retarded children and in trisomy 21.*[21]

Conclusion:

Congenital malformations of the tongue are rare and a medical literature search yielded a very little about its incidence. No other syndromic associations were seen in our patient and this unusual condition was treated by simple surgical excision. Non syndromic tongue anomalies can be treated with simple surgical correction but syndromic tongue anomalies need a multidisciplinary management.

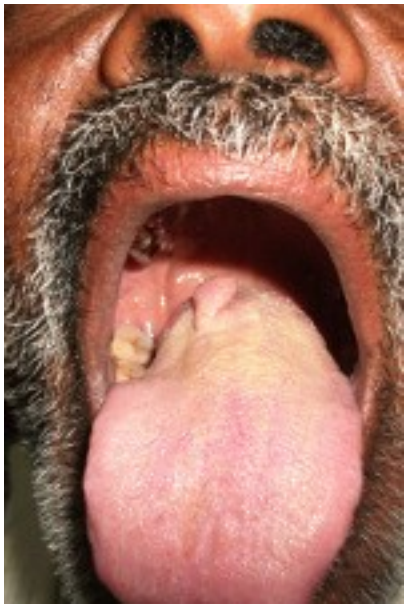

Fig 1 Accessory tongue seen on oral examination

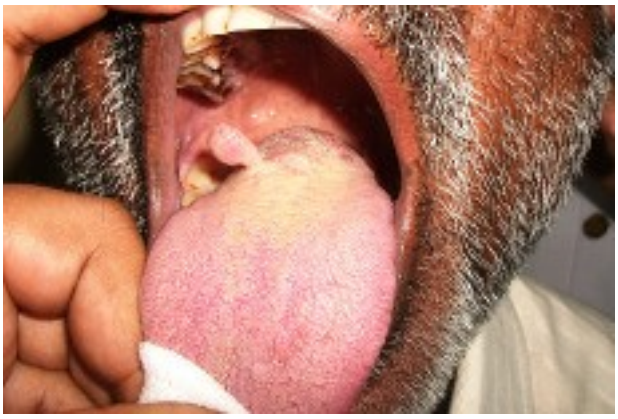

Fig 2 showing accessory tongue on protrusion of tongue

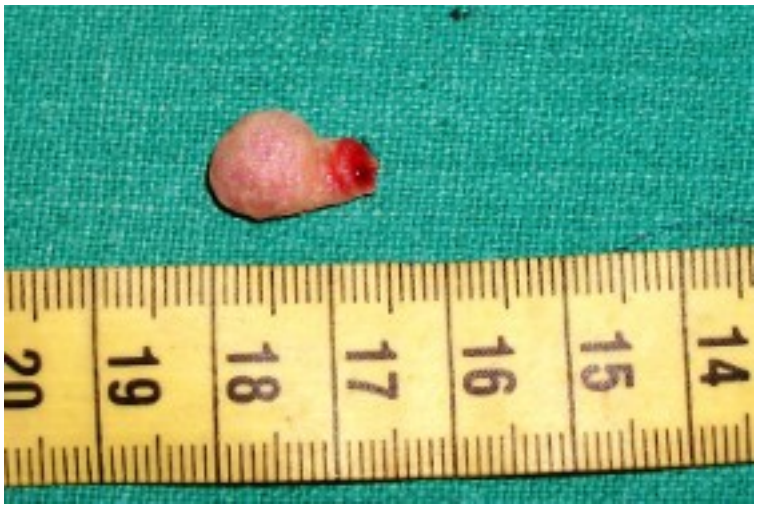

Fig 3 Gross specimen after excision

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