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Case Report

Choristoma of Tongue Presenting as a Congenital Sinus -A Diagnostic Dilemma

*Somnath Saha, **Saibal Misra, ***Padmini Saha ,*Sudipta Pal, *Moushumi Sengupta

ABSTRACT: INTRODUCTION: Choristoma is the presence of ectopic tissue in abnormal locations, where only one germinal layer derivative is present. Choristoma of tongue is a rare entity. Lingual choristoma mostly contains osseous, cartilaginous or gastric tissue derivative in the form of a solid or cystic mass lesion over the dorsum of tongue. MATERIALS AND METHODS: We report a case of a 6 year old boy who presented with a midline tongue sinus which was histologically found to be a choristoma containing colonic tissue. CONCLUSION: A colonic choristoma presenting as a congenital lingual sinus has never been reported in world literature. This is a very rare case which provides hitherto unknown information on tongue choristoma.

Key Words: Lingual Sinus, Choristoma, Chromogranin A.

CASE REPORT: A 6 year old boy presented to our opd with a discharging sinus of the tongue. According to the patient the sinus was present over the tongue since birth. It was generally asymptomatic except occasional bouts of inflammation when there was increased discharge from the opening of the sinus along with pain and surrounding edema. The patient had an uneventful birth history and no similar family history. On examination, opening of a sinus was found over the dorsum of the tongue at the midline. The sinus was situated in the anterior 2/3 of the tongue and was located anterior to the foramen caecum as a separate opening [Fig.-1]. Blunt probing of the tract was done and it seemed to extend deep into the tongue musculature. Systemic examination of the boy was within normal limits. A CT sinography was done to confirm the diagnosis as well as the extent of the lesion. The CT revealed a sinus tract extending through the tongue muscles downwards up to the hyoid bone. The sinus ended at the level of hyoid in a blind sac [Fig. – 2, 3]. No relation to any important structure of neck was visible. Surgical excision of the lesion was done under general anesthesia by a rhomboidal tongue incision and subsequently the tract was delivered through a separate neck incision at the level of hyoid. Body of hyoid was removed along with the tract as it was adherent to the hyoid [Fig. - 4]. The specimen was sent for histopathology, and surprisingly presence of colonic tissue was noted in the specimen in H& E Stains [Fig. - 5]. To confirm the findings, the tissue was again stained with Chromogranin A – a specific stain for colonic glands [Fig.-6]. The Chromogranin stain also came as positive for colonic tissue which confirmed the presence of such unusual cells and helped us to reach the diagnosis of Choristoma Tongue [Fig. - 6]. The patient had an uneventful recovery and he is doing fine after 2 years of follow up. DISCUSSION: Choristoma is defined as presence and/or proliferation of histologically normal tissue at an unusual anatomical site[1]. It is the presence of normal tissues at abnormal locations. Choristoma is closely associated

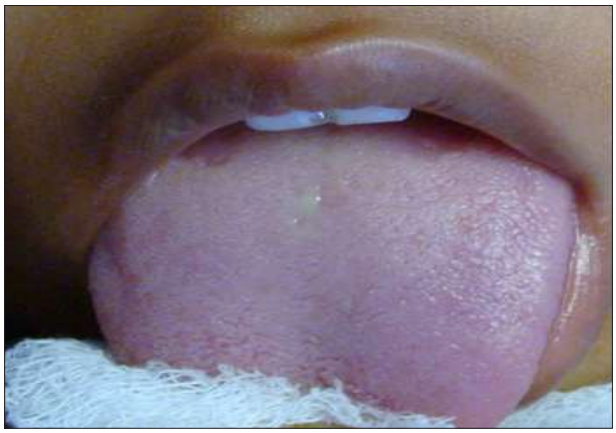

Figure 1: Opening of the sinus in the dorsum of tongue.

Figure 2: CT Sinography showing the sinus tract.

*Department of ENT, Head and Neck Surgery; R G Kar Medical College & Hospital, Kolkata. **Department of ENT, Head and Neck Surgery ;S S K M Hospital, Kolkata. ***Department of Plastic Surgery; R G Kar Medical College & Hospital, Kolkata, India

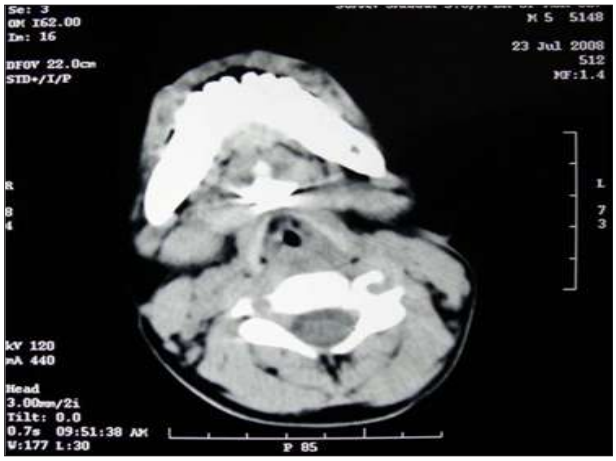

Figure 3: CT Sinography showing the sinus tract ending in front of the hyoid.

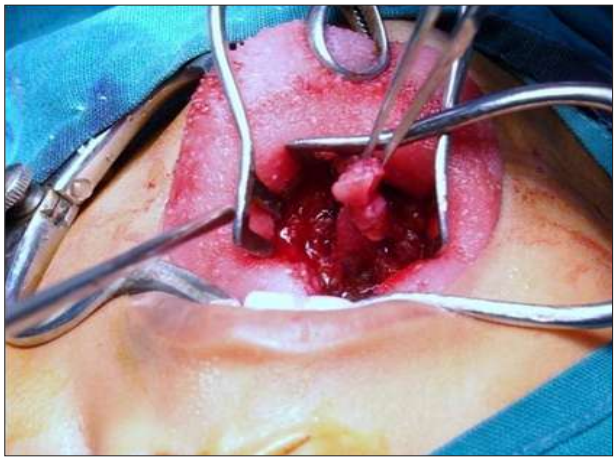

Figure 4: Per operative picture.

with teratoma where three germinal layer derivatives are present histologically, but in choristoma only a single type of tissue is present in the lesion. Choristoma occur mostly in the flat bones of the calvarium or the face and rarely do they affect soft tissues which include that of head, eye, extremities and the tongue[2]. Choristoma of the tongue is a very rare lesion and only a few cases have been reported so far. The first lingual choristoma was reported by Monserrat way back in 1913[3]. Initially it was thought to be a purely osseous lesion but with time choristoma containing other tissues also was reported and the entity was firmly established as a histopathological diagnosis. Batsakis et al classified lingual choristoma as salivary and non salivary types. Nonsalivary choristomas were again sub classified according to their generative tissues as gastroenteric, thyroidal, or sebaceous[4]. However in view of the fact that osseous and cartilaginous choristomas are commoner than the other types hence any classification should include these variants-so a more rational classification would be:

A] Salivary	B] Non Salivary
	i} Osseous
	ii} Cartilaginous
	iii} Gastoenteric – gastric/ colonic
	iv} Thyroidal
	v} Respiratory
	vi } Neural
	vii } Sebaceous

Due to rarity of the cases there is no definite information on the sex predilection of the choristoma patients. Bansal et al in their review of cartilaginous lingual choristoma cases found 15 male and 11 female patients[5]. MJ Ortiz et al reported that patients with gastro intestinal tissue containing choristoma were all males [8 cases] except the one reported by them who was a newborn female[6]. Mir R et al also reported a similar case in a male child[7] whereas VR Naik et al found a female patient having lingual choristoma[2]. Our patient too was a male child going with the observations of the majority of the authors. There is a general consensus that the choristoma is present since birth and the age of presentation depends

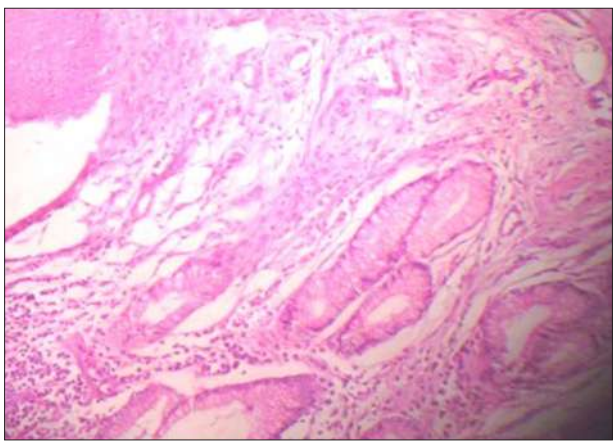

Figure 5: H & E Stain showing colonic glands (200 x).

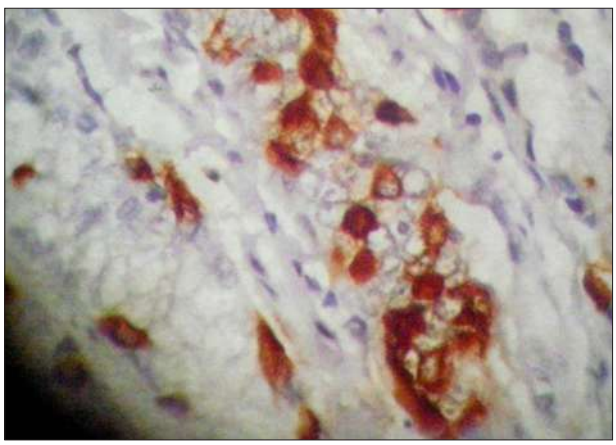

Figure 6: Chromogranin A stain showing cytoplasmic positivity in colonic tissue.

purely on the associated symptoms. The age of presentation varies from birth to 31 years of age in one series[6]. Majority of the authors reported patients who were either newborn or infants presenting very early in life. Generally choristoma is an asymptomatic lesion. However patients may present with a wide range of symptoms; even airway obstruction depending on the size, site and the contained tissues. A review of the related articles revealed that the commonest mode of presentation was as a mass lesion- mostly cystic rarely solid[7]. Apart from our patient only a single paper describes a choristoma presenting as a lingual sinus containing heterotopic gastric mucosa[8]. Osseous or cartilaginous choristoma cases present with a solid lump in tongue which is painless, slowly growing and cause difficulty in speech and swallowing[2,5]. Those having gastric mucosa may come with features of peptic ulceration[8] even with bleeding ulcers[9]. Choristoma of tongue rarely turn malignant except those having origin from thyroid[4]. Sometimes the presenting feature may be due to local inflammation only, like in the present case.

The lesions are hard to diagnose clinically particularly if presenting only as a sinus as in our case. Tc 99 Pertechnate scan has been used by some authors[9] without satisfactory results. In our case we did CT Sinography with surprisingly good visualization of the sinus [Fig.-2, 3]. Treatment is surgical removal in all cases[5, 6, and 10]. The approach is essentially intra-oral but can be combined with external approach depending on the lesion itself. Recurrence is rarely noted.

The present case is unique in view of the fact that here the choristoma presented only as a sinus. So far only a single similar case has been reported[8]. So this is only the second reported case of lingual choristoma presenting as a sinus. Moreover in our patient the generative tissue was from colon, a variant which has never been reported in scientific literature. This being a very unusual finding, the nature of the tissue was reconfirmed with the help of Chromogranin A stain, which came as positive. Chromogranin A stains the cytoplasm of colonic tissue into dark brown colour which is the definitive marker for the presence of colon in the specimen. This case posed with diagnostic dilemma from the very beginning, be it the unusual situation of the sinus, unusual diagnosis of the case as a choristoma and the hitherto unknown presence of colonic tissue in the choristoma. All these factors taken together make this a unique case to be reported for the first time in scientific literature.

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