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Case Report

Heterotopic Oral Gastrointestinal Cyst: Report of Two Cases

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Abstract

Introduction:
Heterotopic gastrointestinal cyst of the oral cavity is an extremely rare lesion with fewer than 40 cases reported in the English literature. It usually involves the soft tissue covering the floor of the mouth and the tongue. Lining of this lesion resembles gastric or intestinal mucosa.

Case Report:
In this report we describe the lesion in two boys and discuss their pathogenesis.

Keywords:
Cyst, Heterotopic gastrointestinal epithelium, Heterotopic oral gastrointestinal cyst

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Introduction:

Heterotopic gastrointestinal cyst of the oral cavity which is also referred to as gastric cystic choristoma, entrocytoma or enteric duplication cyst is a rare lesion which was first introduced by Foderl in 1895 (1,2). To our knowledge fewer than 40 cases have been reported in the English literature so far (3).

In this article we report two cases of heterotopic gastrointestinal cyst of the tongue and the floor of the mouth in two boys and subsequently several theories on their pathogenesis will be discussed.

Case Reports:

A healthy 1-month-old boy was admitted to the pediatric surgery ward of Dr Sheikh hospital due to swelling of the sublingual area that had been present since birth.

Physical examination revealed a 2×3 cm cystic mass lesion in the ventral surface of the tongue, not extending to the floor of the mouth and with no pulsation, hemorrhage or tenderness.

The patient showed no other anomalies on physical examination. Under general anesthesia the cyst was completely excised. On further investigation it was filled with a milky fluid.

Gross examination of the excised tissue showed a monolocular collapsed cyst measuring 9×7mm, with a smooth and hyperemic surface.

The second case was a 9-month- old boy with a 3×4 cm cystic mass lesion in the floor of his mouth which had displaced the tongue upwards. The boy experienced snoring during sleep and minor problems during feeding.

On needle aspiration the cyst was filled with saliva like secretion so it was operated as ranula with an intraoral approach. Interestingly, the cyst's wall was much thicker than ranula but it was completely excised in anatomic plans and two days later the boy was discharged with a generally healthy condition (Fig 1).

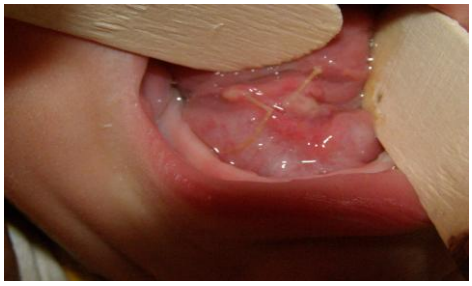

Fig 1: Site of operation

Another gross examination of the excised tissue showed a monolocular collapsed cyst measuring 1×1.5cm with a smooth and hyperemic surface.

Histologically, lining of the two cysts was composed of gastric epithelium with tall columnar mucous cells on the surface and numerous crypts closed short, resembling fundal glands (Fig 2).

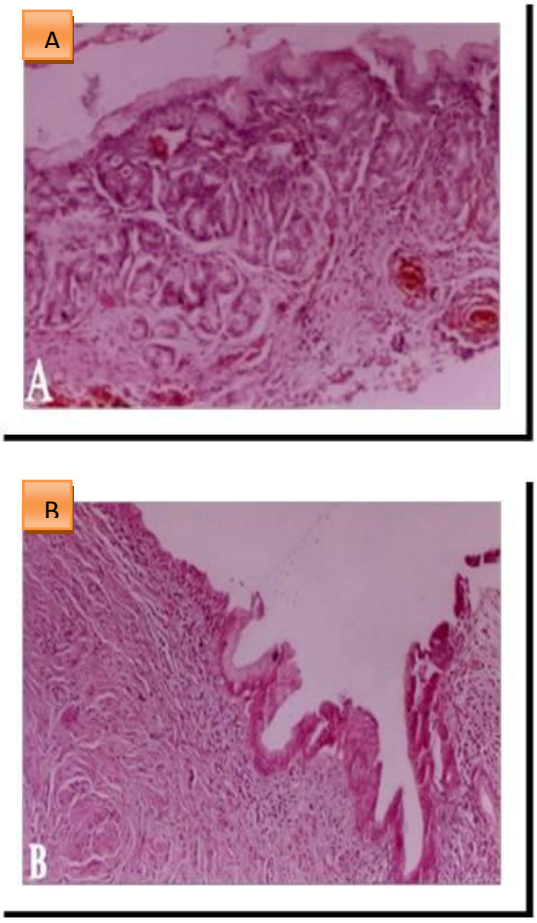

Fig 2: Heterotopic gastrointestinal cyst lined with different types of epithelium:

A) Gastric mucosa-like (H & E- 100).

B) Simple columnar and stratified squamous (H & E- 100)

There was also some smooth muscle in the cysts' walls.

The patients' postoperative courses were uneventful, searching for duplication in other parts of the gastrointestinal tract based on upper gastro-intestinal tract study, follow through and barium enema, were negative and the follow-up to date showed no recurrence.

Discussion:

Heterotopic gastrointestinal epithelium has been more commonly described in the duodenum, gallbladder, common bile duct, jejunum, Meckel's diverticulum, ileum, appendix, colon and rectum (3).

Heterotopic gastrointestinal cyst of the oral cavity is a rare lesion. Since 1895, approximately 40 cases have been reported in the literature (3).

Age of these cases ranged from newborn to 60 years with an average age of 10 years. The majority of cases were infants or young children (2).

The lesions occur more commonly in the ventral surface of the tongue, extending to the floor of the mouth (3). Other reported sites of involvement include the lips, the larynx, submandibular glands, epiglottis and the anterior neck (4).

Regardless of age or location, males are overwhelmingly affected (M/F: 3/2) (3).

Patients are usually asymptomatic at the time of presentation, only 30 percent experiencing increased salivation and difficulty with feeding, swallowing, speech articulation and breathing (5).

Microscopically, lining of these lesions resembles gastric or intestinal mucosa, and stratified squamous, simple columnar or ciliated columnar epithelium may be seen. In most cases, some smooth muscle was present in the cyst wall (2,3).

Simultaneous occurrence with a dermoid cyst in the floor of the mouth has been reported (6). Noorchashm et al reported a mixed heterotopic gastrointestinal and respiratory cyst with involvement of the mandible and extraoral soft tissue (4).

Pathogenesis of this lesion remains uncertain. The most commonly held theory suggests that these cysts may be derived from misplacement of embryonic gastric remnants entrapped in the midline of the tongue, over the tuberculum impar region. This explanation would account for the presence of these lesions in the anterior two thirds of the tongue and floor of the mouth; however, it does not explain lesions occurring in the lateral aspect of the tongue. In addition, this theory fails to explain the presence of intestinal and colonic mucosa in some cysts (7).

It is even more difficult to explain development of dermoid cyst-like areas from embryonic gastric remnants in these lesions.

Another theory proposes that the lesion arises from islands of endoderm that lined the primitive stomodeum and became entrapped during fusion of the embryonic processes. Differentiation into gastrointestinal epithelium may be related to different environmental and inductive influences. Khunamornpong et al reported a case with associated pancreatic tissue and they supported this concept (6).

The lectin staining pattern on mucin from these cysts in Woolgar and Smith's study favors the view that these lesions arise from undifferentiated endoderm subjected to inductive influences (9).

Several other hypothetical pathogenetic mechanisms were proposed including origin from the thyroglossal duct and salivary retention cyst (7). The latter would seem unlikely because this would depend on the dedifferentiation of salivary gland tissue and subsequent differentiation into gastrointestinal tissue.

Presence of smooth muscle in the cyst's wall in our cases suggests that it may be a true duplication cyst. So it is necessary to rule out simultaneous occurrence of

duplication at any level of gastrointestinal tract by imaging studies. According to complete gastrointestinal studies (including: UGIS, follow through and barium enema) we could not find any other site of duplication in these cases.

Irrespective of the pathogenesis of heterotopic cyst, surgical excision is the treatment of choice. Symptoms may be temporarily alleviated by aspiration. However, this approach should be used only as an emergency procedure or during surgical removal because of the possibility of subsequent infection (2).

Recurrence is uncommon but can occur many years later, probably because of incomplete excision (7).

As such cystic lesions with an intraosseous component have been occasionally reported, it is advisable to determine the extent of the lesion by computed tomographic and/or magnetic resonance imaging studies, before surgery.

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