Case Report

Report of a rare cyst at a rare site: Heterotopic gastrointestinal cyst partially lined with dermoid cyst epithelium

ABSTRACT

Heterotopic gastrointestinal cysts (HGCs) of the oral cavity are rare benign lesions mimicking benign or malignant pathologies. These cysts are usually discovered during infancy but may not appear until well into adulthood. The epithelial lining of these cysts shows variable presentation with the presence of an enteric lining as an essential component. The histogenesis is related to entrapment of undifferentiated, noncommitted endoderm within the oral cavity during the 3rd–4th week of fetal life. We report a rare case of oral HGC located at a very rare site, i.e., infratemporal fossa with histopathologic features that included portions resembling dermoid cyst also. Histopathology and etiopathogenesis of the lesion are discussed. Origin from misplaced embryonic remnants, i.e., undifferentiated endodermal cells is suggested.

Keywords: Dermoid cyst, gastric mucosa, infratemporal fossa

INTRODUCTION

Cysts are not very common pathologies found in the oral cavity. They are defined as collections of fluid within a cavity lined by epithelium. Cysts can be developmental or congenital in origin. Most common congenital cysts are epidermoid and dermoid which are encountered throughout the body, with 7% occurring in the head and neck area and 1.6% within oral cavity. Etiopathogenetical theories suggest that congenital cysts are dysembryogenetic lesions that arise from ectodermal elements entrapped during the midline fusion of first and second branchial arches between 3rd and 4th weeks of intrauterine life.

The presence of normal alimentary tissue in an abnormal location is referred to as a heterotopic gastrointestinal cyst (HGC) or duplication cyst. These are also thought to be congenital and rarely occur in the neck. They are formed due to the entrapment of undifferentiated, noncommitted endoderm within the oral cavity during the 3rd–4th week of fetal life.

HGC of the oral cavity was first introduced by Foderl in 1845. Its synonyms include gastric cystic choristoma, enterocystoma, and enteric duplication cyst. It can be found all the way through the gastrointestinal tract including the esophagus, small intestine, pancreas, gall bladder, and Meckel’s diverticulum.

In this case report, the presence of HGC with portions of dermoid tissue is evident at a very unusual site, i.e., infratemporal space, and no single case has been reported in literature at this site till today. Only forty cases of oral HGC (OHGC) have been reported in literature with

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predominant occurrence in the tongue and floor of the mouth.

CASE REPORT

A healthy 17-year-old girl was admitted to the Department of Oral and Maxillofacial Surgery because of swelling on the left side of face since birth. Decreased mouth opening was reported. The patient showed no other anomalies on physical examination. Swelling was tender, firm, afebrile, nonfluctuant, noncompressible, nonpulsatile, and bony hard in consistency. It was extending from the upper border of zygomatic arch superiorly and medially up to body of zygoma [Figure 1]. Lateral and inferior borders of the swelling were not distinct. Patient also gave a history of surgery at the same site 16 years back when she was 1 year old. She had lost all the records pertaining to that surgery. She also informed that after her first surgery, the surgeon had asked her to come after 6 months for another surgery at the same site. She did not report back as she considered herself as well as treated and was totally symptomless. Now, again the reason for reporting to the doctor was the patient’s concern of reduced mouth opening which she wanted to get corrected and also got obvious facial swelling which was making her esthetically compromised. Facial swelling had become obvious only within 6 months.

Axial computed tomography (CT) scan sections revealed a well-delineated variable density soft tissue lesion in left infratemporal and buccal space. The lesion was heterogeneously enhancing on contrast with areas of hypodensity and peripheral enhancement. The lesion was abutting the posterolateral wall of left maxillary sinus and medial aspect of left zygoma causing anterior and lateral displacement of the zygomatic bone. Posteriorly, the lesion was extending up to left ramus of mandible and between the pterygoid plates. CT scan features were consistent with a diagnosis of dermoid cyst [Figure 2].

The cyst was entirely removed under general anesthesia. The excised specimen was submitted to the Department of Oral Pathology with the provisional diagnosis of dermoid cyst or epidermoid cyst. Gross examination of the lesion showed a dumbbell-shaped cystic mass with thick fibrous extension anteriorly. Cystic component measured 3.5 cm × 4.5 cm × 2.3 cm with the fibrous extension measuring 6.5 cm × 2.4 cm × 1.2 cm. On cut section of the cystic lumen, the yellow-colored granular material was seen and stained for cystic content [Figure 3]. The material showed the presence of cholesterol crystals and no keratin. Different bits from different parts of cystic wall as well as fibrous extension were kept for routine processing in the

Department of Oral Pathology. Microscopically, a cystic lining which is thin and thick at places showed uniform fibrous connective tissue stroma supported a lining that contained foci of different types of epithelium: stratified...
squamous epithelium, intestinal type of epithelium, and single layer of tall columnar epithelium. In some parts, the cyst was surrounded by an external layer of smooth muscle [Figures 4 and 5].

The patient is kept on regular follow-up after every 3 months.

DISCUSSION

About forty cases of gastrointestinal cysts or heterotopic masses in the oral cavity have been reported during the past 100 years in the literature. HGCs are abnormally placed rests of gastrointestinal mucosa outside the stomach. Although these lesions are found along the entire digestive tract, oral involvement is extremely rare while it is most commonly found in the esophagus. In the oral cavity, the ventral surface of tongue extending to the floor of mouth is the most common location for OHGC occurring in 60% of patients. The floor of the mouth is the second most frequently affected region. The larynx, anterior neck, submandibular gland, and lips are the other possible rare localizations of OHGC in head and neck region.

Although cases have been reported in oral cavity, this present case of cyst occurring at a very unusual site, i.e., infratemporal fossa has never been reported in literature till date.

A review of literature done in 1992 by Christopher et al. shows that the age of patients at the time of diagnosis of OHGC ranges from the newborn to 34 years. Out of 22 cases reported till 1992, 14 lesions (63%) were present in males and 8 in females. Our case reports this cyst in a female of 17 years. In all cases, the lesions were treated by conservative surgical excision. Recurrence was described in only one case where incomplete removal originally occurred. On the basis of the history of surgery at the same site given by the patient, it was assumed that some parts of the cyst were partially removed 16 years back and now it has recurred.

The clinical presentation of OHGC is usually an asymptomatic swelling ranging from 1 to 3 cm in diameter. The present case also showed an asymptomatic swelling, but due to cosmetic reasons, the patient wanted the cyst removal.

The pathogenesis of this lesion remains uncertain. Several theories have been postulated; the most commonly held suggests that these cysts may be derived from misplacement of embryonal rests. Early in embryonic development, the undifferentiated primitive stomach lies in the mid-neck region close to the primordium of tongue, and it is assumed that the endodermal gastric mucosa becomes entrapped in the midline of tongue by fusion of lateral lingual swellings over tuberculum impar. This explanation would account for the presence of these lesions in the anterior two-third of tongue and floor of mouth; however, it does not explain lesions occurring in the lateral aspect of tongue. In addition, this theory fails to explain the finding of intestinal and colonic mucosa in some cysts. Other theories propose that these cysts arise from the thyroglossal duct or from salivary retention cysts. The latter would seem unlikely because it would depend on the differentiation of salivary gland tissue and subsequent differentiation into gastrointestinal tissue.

Reports from the literature reveal that lining of the cyst was composed of gastric epithelium in some cases; in others, there were foci of stratified squamous, respiratory, or intestinal epithelium. In most cases, some smooth muscle was found in the cyst wall. In the present case, the cyst had developed into dumbbell shape in the infratemporal space with one end toward the buccal cavity and the other end toward the infratemporal space. Histopathological sections from the end toward the oral cavity showed thickened stratified squamous epithelium. There were abundant areas in the specimen where this thickened cystic lining had thinned down to single layer of tall columnar cells and was also broken down into crypts. The other end of the cyst which was toward the temporal bone or infratemporal space; the cystic lining was pseudostratified with tall columnar cells and
HGC is an extremely rare clinical entity in the head and neck region. It is a rare cyst in the oral cavity with most common location on the tongue followed by floor of the mouth. The larynx, anterior neck, submandibular gland, and lips are the other possible rare localizations of OHGC in the head and neck region.[5,13] Although OHGC is mostly seen in infants, it can be encountered in adults. Clinically, masses of OHGC presented usually in cystic form and rarely in the solid form.[13] Sometimes, these masses approach huge dimensions because of which various clinical complications arise such as excessive salivation, difficulty with feeding, swallowing, speech, or breathing in isolation or combination. Regardless of the localization of OHGC, the treatment of choice is complete surgical excision and long-term follow-up is recommended. However, CO₂ laser has also been used for the excision. Recurrence is uncommon but has been reported in the literature as in our case, probably because of incomplete excision.[5,13]

In addition to routine hematoxylin and eosin staining, other confirmatory investigations include the use of special stains such as periodic acid-Schiff (PAS), Alcian-PAS, and high iron diamine staining which show positivity for simple columnar, gastrointestinal, and ciliated epithelium. Immunohistochemically, cytokeratin 7, 8, and 19 and Pan-cytokeratin demonstrate positivity for simple columnar, gastrointestinal, and ciliated epithelium. Lectin histochemistry pattern of OHGC suggests the presence of galactose, N-acetylgalactosamine, and galactose-N-acetylgalactosamine compounds that also support the origin of the lesion from undifferentiated endoderm.[7,14]

CONCLUSION

This article emphasizes the importance of rare oral pathologies which impose not only serious esthetic problems but also functional insufficiencies; hence, OHGC in differential diagnosis must be considered. Complete surgical excision should be the treatment of choice as incomplete excision can cause recurrence as reported in this case.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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