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Heterotopic gastrointestinal cyst in the oral cavity

Quiste gastrointestinal heterotópico en la cavidad oral

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Keywords:
Choristoma, heterotopic gastrointestinal cyst, enterocystoma, duplication cyst.

Palabras clave:
Coristoma, quiste gastrointestinal heterotópico, enterocistoma, quiste de duplicación.

ABSTRACT

Heterotopic gastrointestinal cyst (HGC) is a rare, unilocular and asymptomatic variable size lesion that when reaching a larger diameter, it can compromise the patient's airway, feeding and speech. It is mainly located on the tongue and floor of mouth on the anterior dorsum and ventral surface. This case report is intended to increase the statistics of the few reported cases worldwide and to foster anatomopathological studies of different surgical specimens in order to ensure a diagnosis with clinical and radiographic support. All this for the patients' benefit and their timely recovery. The surgical treatment was successful.

RESUMEN

El quiste gastrointestinal heterotópico (CGH) es una lesión rara, unilocular y asintomática de tamaño variable que, cuando alcanza un diámetro mayor, puede comprometer la vía aérea, la alimentación y el habla del paciente. Se localiza principalmente en la lengua y en el piso de la boca, en el dorso anterior y en la superficie ventral. Este informe de caso pretende aumentar las estadísticas de los pocos casos reportados en todo el mundo y fomentar los estudios anatomopatológicos de diferentes especímenes quirúrgicos con el fin de asegurar un diagnóstico con apoyo clínico y radiográfico. Todo ello en beneficio de los pacientes y de su oportuna recuperación. El tratamiento quirúrgico fue un éxito.

Introduction

The term cyst is defined as a cavity lined with epithelium while the terms heterotopia or choristoma are used to describe an organ or tissue displaced to an abnormal location.[1] Therefore, heterotopic gastrointestinal cyst of the oral cavity is a rare, benign pathology of approximately 100 documented cases in the literature and was first explained by Foderl in 1895.[2,3] Although the affected patients are usually infants, this condition has also been described in adults up to 70 years of age. Its etiology is still poorly understood and the most accepted theory hypothesizes that around the 4th week of embryonic development, some islands of endodermal cells undergo a process of sequestration of the stomodeum.[4]

This lesion may be located on the tongue and floor of mouth on the anterior dorsum and ventral surface, on the midline or laterally. It is of unilocular appearance and on palpation is fluctuant and with greater consistency.[4,5]

Patients may have no symptoms, but when expanding and reaching huge dimensions, it makes proper feeding difficult. Regarding newborns, sucking becomes a difficult task therefore, it prevents them from breastfeeding and causes excessive salivation and partially airway compromise; moreover, adults may have speaking difficulty.[6]

The diagnosis is based on a clinical examination, besides a recommended imaging test, mainly a magnetic resonance imaging to determine the full extent and proximity to important anatomical structures in order to obtain an optimal and minimally invasive surgical planning. Additionally, a computed tomography scan and an ultrasound scan can be performed, the latter are very useful to verify its internal composition and cyst confirmation. On the other hand, a needle aspiration may be chosen.[7,8] The histopathological examination along with haematoxylin and eosin staining are the base diagnosis that allow for tall cylindrical mucosal cells with pale cytoplasm observation, in addition

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to goblet cells, all forming a pseudostratified epithelium that is consistent with intestinal epithelium. keratinized or non-keratinized stratified squamous epithelium, and even respiratory epithelium can also be found. According to the previous information, no evidence of atypia or malignancy has been reported and for certain cases immunohistochemistry or special stains would be useful.[2,4,5]

The suggested treatment for this pathology is complete conservative surgical excision in order to protect nearby structures and keep the patient under control. Recurrence is uncommon, therefore a good prognosis.[3,9] This case report shows a new clinical case with clinical and histological features of choristoma, also referred as enteric duplicative enteric cyst or heterotopic gastrointestinal cyst.

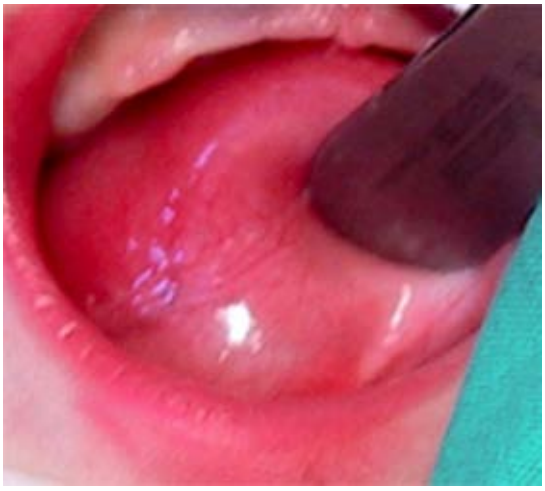

Figure 1: *Large non-compressible mass in the body of the tongue.*

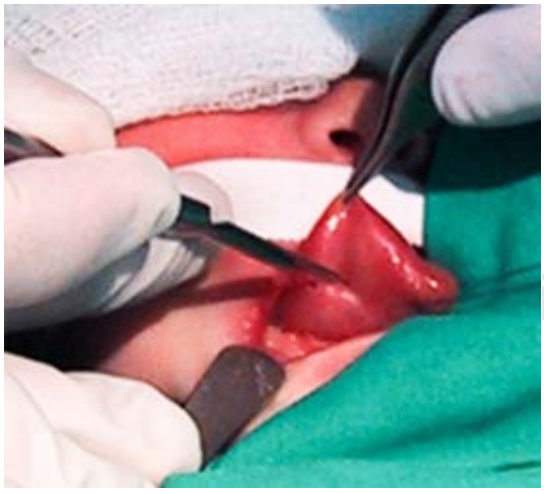

Figure 2: *Oval incision.*

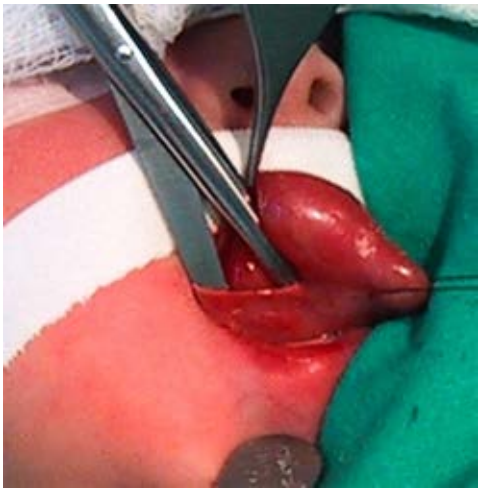

Figure 3: *Scissors´s mass separation maneuver.*

Case report

A 3-day-old male newborn referred by his pediatrician to the Stomatology and Oral Surgery Service of the *Hospital Infantil Universitario de Manizales*. At the time of consultation, the mother referred «the child has something growing on his tongue».

Intraoral examination revealed an enlarged tongue on the ventral surface extending right to the floor of mouth due to a soft, non-pulsatile and non-compressible mass in the body of the tongue *(Figure 1)*. A needle aspiration and extraction of contents of the pathology is performed, obtaining a viscous, milky-colored material that forms threads on contact with the metal instruments, which is presumed as saliva Previous characteristics suggest a presumptive diagnosis of ranula thus, the patient is scheduled for surgery under general anesthesia in order for marsupialization in compliance with the management guidelines.

During the procedure, an encapsulated lesion is evidenced, which allows for the complete excision treatment *(Figures 2 and 3)*. During surgery, because of this pathology's cystic conditions, an extravasation shows a milky and viscous consistency cyst *(Figure 4)*. The entire specimen is sent to the pathology laboratory for processing and analysis *(Figure 5)*.

Macroscopic examination reported a 1 × 1 × 0.7 cm nodule with partially open clefts containing translucent mucoid material. Under microscope observation with basic hematoxylin and eosin staining, clefts with epithelial coverings of various types are seen: respiratory, intestinal, keratinized squamous and non-keratinized. No atypia or changes indicative of malignancy *(Figures 6 to 9)*.

By correlating the clinical and histopathological features, a diagnosis of certainty is obtained: Heterotopic

Gastrointestinal Cyst in the oral cavity or choristoma. This was the best choice treatment. The patient attended the postoperative control for stitches removal, and an optimal healing process was observed according to the postoperative course of time. The mother reported proper sucking function and food intake.

Discussion

The origin of the heterotopic gastrointestinal cyst is during embryonic development and can be prenatally identified or discovered after birth. Although its etiology is still unknown, some theories have stated that between the 4th and 5th week of embryonic development, the cells of the gastrointestinal endoderm become entrapped in the primitive stomodeum, that is, some of these endodermal cells would be sequestered during the fusion of primordial tissues during the fusion of the lateral lingual protuberance (distal bud of the tongue) and the odd tubercle (medial bud of the tongue). By embryonic induction effect, these cells can be differentiated into various types of epithelium (gastric, colonic type, intestinal, respiratory, squamous, and glandular epithelium).[4,7,10] Another theory states that it could be a pathology that develops from retention cysts where dedifferentiation into other types of epithelia.[3]

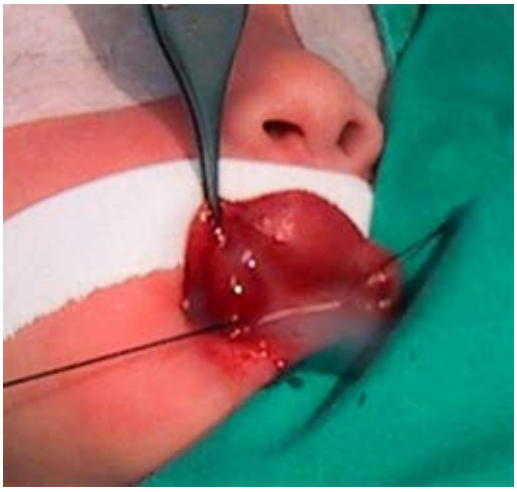

Figure 4: *Removal of the mass.*

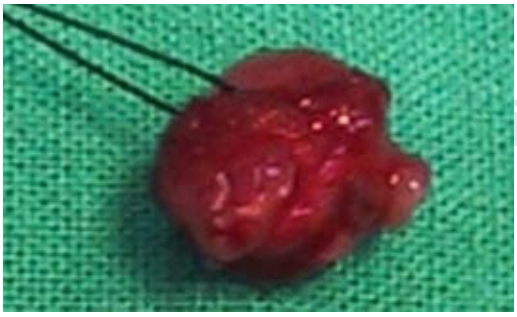

Figure 5: *Final mass: 1 × 1 × 0.7 cm.*

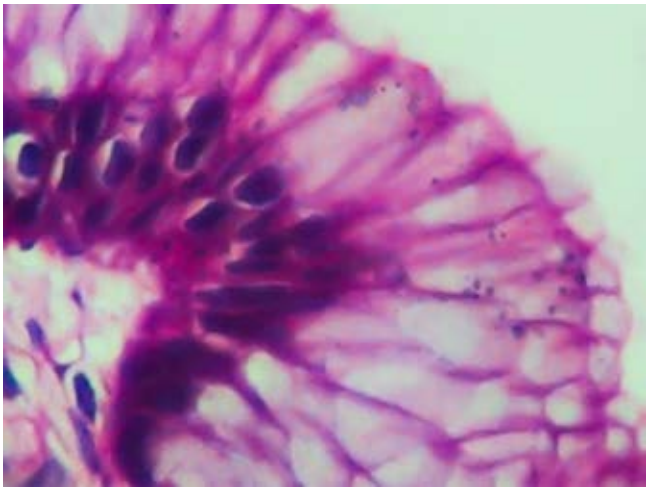

Figure 6: *Intestinal epithelium, mucosecretory.*

The preoperative evaluation of lingual cystic mass in newborns may involve: palpation, ultrasound, computed tomography and magnetic resonance imaging; however, the latter are a risk for the child because of sedation, but when the lesion occupies a large area, magnetic resonance imaging is an effective tool to determine its extension. On the other hand, high-resolution ultrasound is an excellent test to verify the non-vascular nature of the lesion and allows for its size identification. Needle aspiration has also been suggested for its diagnosis, however, it disposes the patient to infection, the need for sedation, and an increase in the number of appointments for treatment which conditions a negative attitude.[8,11]

The most common pathologies in the floor of mouth are dermoid cysts with 21.5%, duplication cysts and ranula with 13.6%, teratoma with 12.5%, and in the case of heterotopic gastrointestinal cyst with only 3.4%. The other percentages correspond to other lesions, so it can be identified how rare it is to have a case like the one presented.[8]

The first cyst, with intestinal and gastric mucosa, was reported by Foderl in 1895. These lesions are rarely seen in the mouth, in the ventral surface of the tongue. These cysts are usually reported along with the intestinal tract from the esophagus to the colon and liver, pancreas, lungs, larynx and urinary system. The first case published as a duplication cyst was by Duncan and Daniel in 1942.[12]

Heterotopic gastrointestinal cyst of the oral cavity is more common in boys than in girls. These masses involve

the anterior part of the tongue in 60% of the cases. Some are symptomatic and others cause difficulties in breathing, feeding, manifest unexpected hemorrhages or a brownish discharge of intralingual material. Most of these cysts are solitary, but in some cases more than one cyst has appeared. This lesion has been given several names including: choristoma, congenital enteric duplicated enteric cyst, enterocystoma, and duplication cyst. This different terminology has led to confusion among practitioners in describing its nature. Some of these misunderstanding were resolved by Rickham et al. who established criteria for duplication cysts. These criteria include: presence of some type of digestive tract mucosa and location adjacent to some part of the digestive tract.[10]

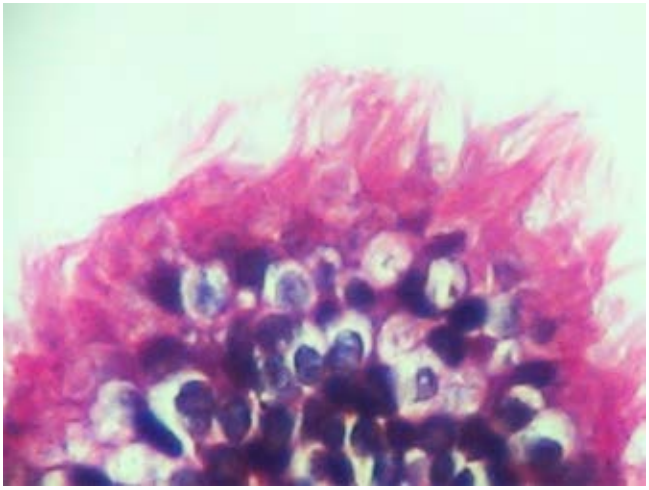

Figure 7: *Ciliated, cylindrical and respiratory epithelium.*

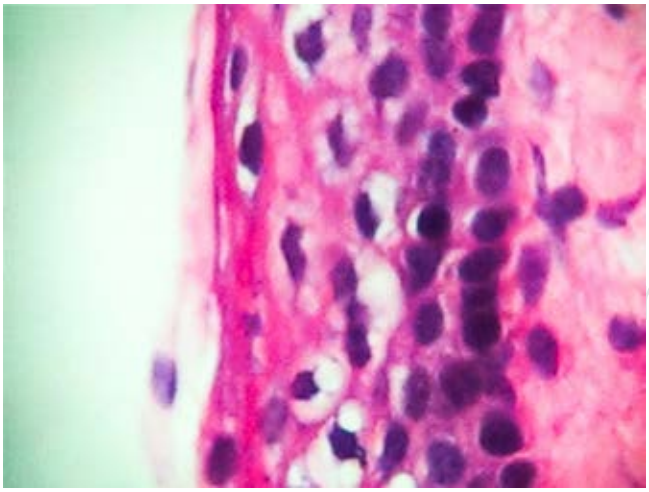

Figure 8: *Squamous epithelium.*

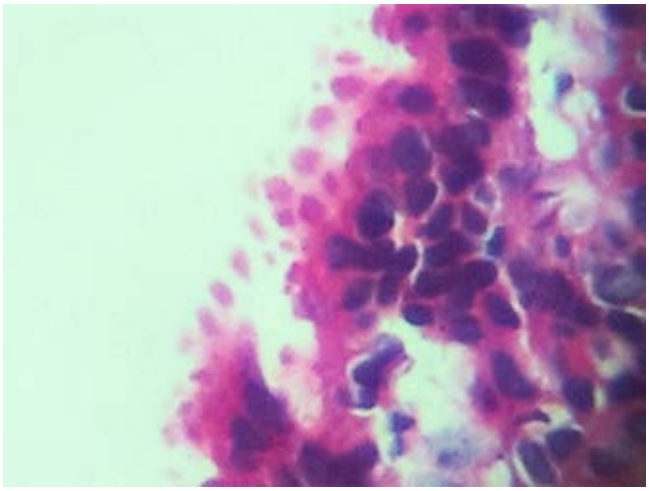

Figure 9: *Epithelium with decapitation secretion.*

A lingual cystic mass in a newborn may be an interesting finding for the head and neck professional, especially because of their differential diagnoses which clinically resemble. Among them are congenital ranula, teratoma, dermoid and epidermoid cyst, duplication cyst of the anterior intestine, hemangioma, lymphangioma and thyroglossal duct cyst.[4,8]

There is still no consensus on the appropriate age for treatment. It has been proposed that for safe surgery it is better to wait until the first year, although this is controversial because if the lesion causes impairment of vital functions, then it should be performed as soon as possible.[13] The excision of an intralingual cyst is complex, as removing the adherent walls around the muscle is a tedious exercise, especially in patients with a huge cyst, so imaging is essential to avoid complications such as injury to Wharton's duct or lingual muscles. Conventional techniques, including blunt or sharp dissection, can cause bleeding, limit observation of nearby vessels and nerves, and substantial postoperative edema. Incision on the ventral aspect of the tongue has been found to be the most conservative approach, as it produces less scarring and allows the surgeon to preserve normal tongue function postoperatively.

Laser therapy provides good hemostasis and better identification of the anatomy around the lesion, resulting in minimal tissue damage, little postoperative inflammation and less scarring. Some authors prefer to perform the procedure with bipolar cauterization because it is a safer and less expensive alternative.[13-15]

Conclusions

The location of heterotopic gastrointestinal cysts in the oral cavity are rare and when they occur, being more frequently

observed on the tongue and floor of mouth, thus the health practitioner must perform the anatomopathological study to determine its nature. This pathology may be asymptomatic and cause feeding or breathing difficulties. Prompt surgical treatment should be performed, however, parents should be reassured that the prognosis is good.

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