



Case report

Respiratory distress associated with heterotopic gastrointestinal cysts of the oral cavity: A case report



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H I G H L I G H T S

- There are less than a hundred cases reported in the medical literature.
- It is vital for surgeons and physicians to include this pathology in their differential diagnosis.
- Imaging studies are essential for obtaining clearer diagnostic possibilities.
- Complete surgical resection is needed to prevent recurrence.

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A B S T R A C T

Heterotopic gastrointestinal cysts of the oral cavity are benign lesions usually discovered during infancy. Their pathogenesis is not very clear. They are rare congenital anomalies that result from remnants of foregut-derived epithelium in the head, neck, thorax or abdomen during embryonic development. The majority of these lesions occur in the anterior ventral surface of the tongue and extend to the floor of the mouth. They are confused clinically by surgeons in cases of head and neck masses in children as ranulas, dermoid and thyroglossal cysts, and lymphangioma. We report the case of a 28-day newborn with a 3.6 cm oval mass on the floor of the mouth causing difficulty eating and cyanosis during crying. Complete surgical excision was performed by an oral approach under general anesthesia. Microscopic examination revealed gastric epithelium with tall columnar mucous cells on the surface and numerous short closed crypts, resembling fundal glands and mature gastric epithelium.

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1. Introduction

A gastrointestinal heterotopic cyst is a rare benign lesion diagnosed mainly in children. Heterotopic gastric mucosa can appear almost anywhere in the gastrointestinal tract, but mostly in the mediastinum, retroperitoneum, and oral cavity. Congenital oral masses may interfere with vital functions such as respiration and deglutition in neonates and cause failure to thrive [1,2]. Toyama [3], in 1927, was the first to describe oral cavity cysts lined by gastrointestinal mucosa. Foregut cysts develop from embryonic remnants

of foregut epithelium, and are usually observed in the abdomen and thorax. They are rare but may occur on the tongue, floor of the mouth and pharynx and occasionally cause airway obstruction or feeding difficulty. These cysts, also called choristomas, contain heterotopic islands of gastrointestinal mucosa that is usually gastric but occasionally intestinal, colonic or pancreatic. The duplication of this mucosa is based on a triad of (1) a cyst covered by a smooth muscle coat, (2) the presence of epithelium derived from the foregut, and (3) attachment to a portion of the foregut, which gives this entity its name [4–8].

This is a report of a patient with a congenital heterotopic gastrointestinal cyst, located in the anterior ventral surface of the tongue with extension to the floor of the mouth causing difficulty feeding and airway obstruction.

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2. Presentation of case

The patient is a 28-day-old boy from northern Mexico, product of a fourth gestation, with a history of prenatal supplementation of folic acid and iron, and born by caesarean section. The child was a full-term normal newborn with normal APGAR scores, weighing 3.87 kg, and 51.5 cm in length. He was referred because of a mass located in the anterior ventral surface of the tongue with extension to the floor of the mouth (Fig. 1). The mass was present from birth and gradually increased in size causing posterior displacement of the tongue, difficulty during feeding and cyanosis while crying. Laboratory tests on admission reported hemoglobin of 16.2, a white blood cell count of 12,500 with a differential count of 62.9% lymphocytes, and 17.8% neutrophils. Total bilirubin was 4.4 mg/dL, with a direct bilirubin of 0.4 mg/dL, and an indirect bilirubin of 4.0 mg/dL; phosphorus was 6.0 mg/dL, and coagulation tests showed a prothrombin time of 10.3 seconds, and a partial thromboplastin time of 38.3 seconds.

Computed tomography (CT) was performed and reported a cystic tumor $3.6 \times 3 \times 2.2$ cm located in the midline of the floor of the mouth and the ventral portion of the tongue (Fig. 2A), following the path of the sublingual gland with an approximate volume of 12 cc. In a sagittal section, the cyst was seen located in the ventral portion of the tongue extending above the mylohyoid muscle (Fig. 2B). The initial report suggested a ranula of the sublingual gland.

Tumor resection was achieved under balanced general anesthesia. Nasal intubation was performed after repeated orotracheal attempts that failed due to the large size of the cyst, which also added more space for a comfortable surgical procedure.

A reference probe was placed on the dorsal tongue and transoral excision of the lesion was carried out by an Otolaryngologist specialist. During surgical resection, the mass was identified on the ventral edge of the tongue with extension to the depth of the mylohyoid muscle. The lesion was covered by a firm, whitish



Fig. 1. Clinical appearance of Heterotopic gastrointestinal cyst of the oral cavity presenting as a mass located in anterior ventral surface of the tongue with extension to the floor of mouth.

surface. It did not adhere to adjacent tissues, revealing an amber liquid content. The cyst was fully resected and the raw area was covered by mucosal flaps, which were approximated with absorbable sutures.

Macroscopically, the cyst was well circumscribed measuring 3×2.5 cm with an oval shape and covered by light brown smooth, fibrous, soft, whitish and shiny connective tissue and containing a mucinous transparent liquid. The histopathology study reported the presence of gastric epithelium in the lining of the cyst with tall columnar mucous cells on the surface and numerous closed short crypts, resembling fundal glands. The mature gastric epithelium was intermingled with goblet cells, along with pseudostratified ciliated columnar epithelium and nonkeratinized stratified squamous epithelium. A lymphocytic infiltrate was also present (Fig. 3). The final histopathologic diagnosis was a heterotopic intestinal cyst. The liquid content of the cyst sample was positive for mucin using the Mayer Mucicarmin staining and no inflammatory cells were found. Bacterial cultures of the fluid were negative. No recurrence was observed 6 months after surgical treatment.

3. Discussion

Heterotopic gastrointestinal cysts of the oral cavity are very rare benign lesions that are usually discovered during infancy. Although around 90 cases have been reported in the literature, it is difficult to determine the incidence of these lesions because of varied nomenclature [9]. Heterotopic gastrointestinal cysts are also referred to as choristomas, which are defined as a mass of tissue that is histologically typical of an organ in an abnormal location. They are located in the tongue in 60% of patients and the floor of the mouth is involved in almost 40% of cases. These lesions involve the ventral surface of the base of the tongue extending to the floor of the mouth, which is the second most common location. It is more frequent in males in 97% of cases and can cause respiratory obstruction in neonates. This type of cyst is most common in the gastrointestinal tract and is commonly described in the duodenum, gallbladder, jejunum, Meckel's diverticulum, ileum, appendix, colon and rectum, but it can be found in the lung, larynx, pancreas and bladder, usually presenting as an asymptomatic mass [10–15].

Tucker et al. [15] reported that heterotopic gastrointestinal cysts consist of gastric-type epithelium, although there have been reports of squamous, small intestinal, and colonic-type epithelium. More than 1 type of mucosa can be present within the same cyst. It may contain only mucosal lining or have a full-thickness duplication consisting of mucosa, submucosa, and muscularis propria. Skeletal muscle may be seen at the cyst's periphery, demonstrating an association with the tongue. The etiology of these cysts is unknown. The widely accepted theory is that the lesion originates from islands of endoderm from the primordial stomach in the 3 to 4 mm (4-week) embryo. The growth rate of the heterotopic gastrointestinal cysts is not stated precisely in the literature. However, most cases are reported from newborns to childhood, suggesting a slow growth [15].

In our case, the patient had symptoms of airway obstruction and difficulty swallowing in an intermittent pattern due to the size of the mass and emergency surgical treatment was not required. The histological report in our patient consisted of more than 1 type of mucosa (gastric and intestinal) and it was composed of stratified squamous non-keratinized epithelium, and non-keratinized ciliated columnar epithelium.

The differential diagnoses of pediatric cystic head and neck masses are described as follows. Ranulas are mainly paramedian having a characteristic clinical appearance with fluctuant consistency. Dermoid cysts have a firm-elastic consistency, mobility in the adjacent area, leaves pressure marks and characteristically a grey-

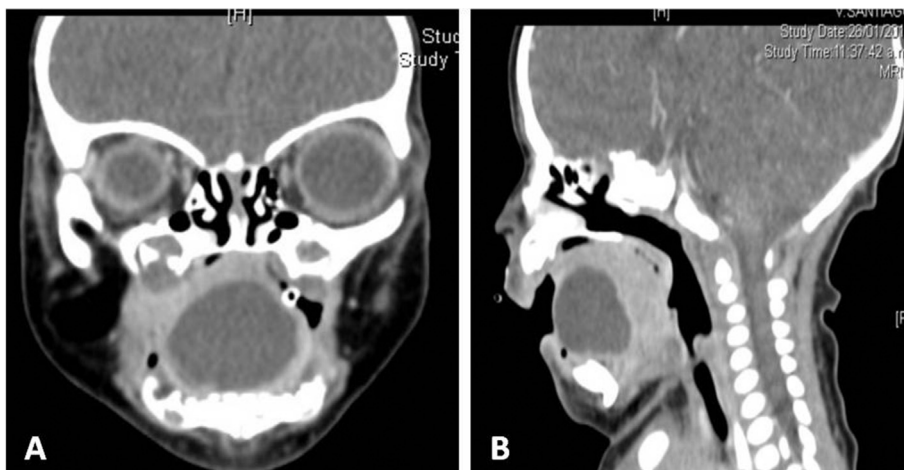


Fig. 2. A. CT coronal section showing a heterotopic gastrointestinal cyst in the midline of the floor of the mouth and the ventral portion of the tongue. B. Sagittal section showing a relatively well-defined cystic structure in the ventral portion of the tongue extending above the mylohyoid.

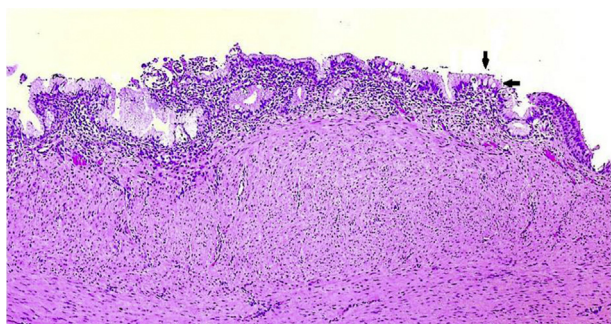


Fig. 3. Histology of the cyst. A thick capsule composed of dense fibrous tissue and smooth muscle is lined by columnar cells similar to gastric epithelium, occasional goblets cells can be seen (arrows). H&E 10x.

yellowish viscous content. Lymphangiomas are present at birth or infancy but frequently with a polycystic appearance with a sero-hemorrhagic liquid content. Thyroglossal cysts on the other hand are present in a paramedian plane and can be distinguished by its mobilization when swallowing and protrusion of the tongue. The definitive treatment to prevent recurrence of these masses is a complete surgical resection [16].

As part of the preoperative evaluation, studies such as CT and MRI are used because they provide an outline of the extent and character of larger lesions. MRI provides superior soft tissue resolution compared with CT. Prenatal ultrasonography is a possible tool for diagnosis depending on the size of the cyst, equipment and skill of the radiologist [17–19].

We emphasize the importance of CT or MRI depending on availability, in the evaluation of head and neck masses, to obtain clearer diagnostic possibilities and to determine the extent of the lesion and assist in the surgical approach [19].

4. Conclusion

In conclusion, most heterotopic gastrointestinal cysts in the oral cavity are rare lesions characterized by the presence of ectopic gastrointestinal mucosa. They are usually asymptomatic and, in the head and neck region, this mandates vigilance with respect to the airway. Some can cause respiratory distress and swallowing problems. The differential diagnosis of masses in the oral cavity is difficult. Imaging studies can help to determine the nature and

growth of the lesion and the participation of a pathologist in the definitive diagnosis is very important. Complete resection assures non-recurrence with an excellent prognosis.

This work has been reported in line with the SCARE criteria [20].

Ethical approval

Ethical Approval number OT16-00002 by Comité de Ética en Investigación del Hospital Universitario “Dr. José Eleuterio González”.

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Author contribution

Marco Antonio Mendez Saenz MD: medical procedure application.

Marco de Jesús Villegas González MD PhD: project supervision.

Marco A. Ponce Camacho MD PhD: sample analysis.

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Barbara Sáenz Ibarra MD: simple analysis.

Blanca I Esquivel Gacía MD: medical records management.

José Luis Treviño González MD PhD: project planning, manuscript development, and analysis.

Conflict of interest

The authors declare that they have no conflicts of interest.

Trial registry number – ISRCTN

This does not apply to case reports.

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