CONGENITAL CHORISTOMAS OF THE ORAL CAVITY IN CHILDREN

Raymond L. Chai, MD; John A. Ozolek, MD; Barton F. Branstetter, MD; Deepak K. Mehta, MD; Jeffrey P. Simons, MD

OBJECTIVES/HYPOTHESIS: To review our institutional experience with oral cavity choristomas in children.

STUDY DESIGN: Retrospective case series and medical record review.

METHODS: Medical records including clinic notes, operative reports, radiologic studies, and pathology specimens were reviewed. All imaging studies and pathology material were reviewed by a head and neck radiologist and pediatric pathologist, respectively.

RESULTS: Sixteen patients (10 males and 6 females) with congenital oral cavity choristomas were identified. Mean age at diagnosis was 1.8 months. Location of the lesions included tongue (n = 9) and floor of mouth (n = 7). Preoperative imaging included magnetic resonance imaging (n = 6), computed tomography (n = 5), plain radiography (n = 1), and no imaging (n = 4). Radiographically, the lesions were consistently lobular with well-defined margins, but other imaging features often mimicked other masses that can arise in the tongue and floor of mouth. Symptoms were present in five of 16 patients and included difficulty feeding, swelling with upper respiratory infection, and partial airway obstruction. Complete surgical excision was performed in 15 of 16 patients; mean age at the time of surgery was 12.7 months. One patient underwent marsupialization. No complications were noted perioperatively. No recurrences of choristoma were seen. On histologic examination, the predominant component was cystic with cyst linings of respiratory epithelium (n = 5), gastric (foveolar) epithelium (n = 1), or both (n = 10).

CONCLUSIONS: This study supports surgical excision as an effective intervention for children with oral cavity choristomas. Because the etiology of these cysts is unknown and diagnostic terminology is widely variable, we propose a more descriptive diagnosis based on the histology (i.e., lingual choristoma or lingual developmental cyst with respiratory epithelium or lingual choristoma with gastric epithelium).

KEY WORDS: Choristoma, foregut duplication cyst, lingual cyst, pediatric.

LEVEL OF EVIDENCE: 4.

LARYNGOSCOPE, 121:2100–2106, 2011

INTRODUCTION

In the pediatric population, the head and neck region is a common location for benign abnormal tissue proliferations that include ectopias, heterotopias, choristomas, and hamartomas. Ectopic, heterotopic, and choristomatous tissues all represent histologically normal tissue where it should not be, and hamartomas are abnormal proliferations of tissue that would normally be present in a particular location. In the pathologic sense, choristomas are defined as histologically normal tissue that is present in an abnormal location and may differ from ectopias and heterotopias by having tissue that would normally be present outside of the head and neck region (i.e., gastric).

Choristomas of the head and neck can occur in the tongue (lingual choristoma), floor of mouth, pharynx, and hypopharynx and are clinically important lesions because they can be a cause of neonatal airway obstruction and significant feeding difficulties. Oral cavity choristomas are typically found in either the anterior two thirds of the tongue or the floor of the mouth. Variable terminology has been used for their pathologic diagnoses, including anterior median lingual cyst, lingual bronchogenic cyst, lingual cyst of foregut origin, enterogenous cyst, gastric heterotopias, and others. The pathogenesis of these lesions is unknown, but implicit within the definition of choristoma is the aberrant developmental nature of the lesion. The aim of this study is to review the clinical, radiologic, and pathologic findings of a series of lingual choristomas from a single large tertiary-care children’s hospital.

MATERIALS AND METHODS

Sixteen cases of congenital choristomas of the oral cavity were identified from pathology records at the Children’s Hospital of Pittsburgh of UPMC.
Hospital of Pittsburgh during the period from January 1991 to May 2010. Pathology records were searched by using a natural language search function within CoPath Plus (Cerner Corporation, Kansas City, MO). Search terms included “floor of mouth cyst,” “choristoma,” “foregut duplication,” “gastric tongue,” “teratoid cyst,” and “tongue cyst.”

Clinical records were then identified for candidate patients and reviewed by using either Cerner Powerchart Electronic Medical Record System (Cerner Corporation, Kansas City, MO) or paper charts. Radiology images were reviewed using the iSite Picture Archive and Communications System (Philips Healthcare, Andover, MA) by a head and neck radiologist (B.F.B.). The clinical information obtained via chart review included demographic data, medical history, physical examination, operative approach, complications, and follow-up information. Pathology slides were obtained from the slide archives of the Division of Pediatric Pathology, reviewed, and histologic findings documented by a pediatric pathologist (J.A.O.). As a retrospective review, this study was granted exemption from informed consent by our institutional review board under the University of Pittsburgh Institutional Review Board guidelines for exempt review (IRB #PRO09060008).

### RESULTS

#### Clinical

Table I summarizes the clinical and demographic information for this cohort of patients with tongue/floor of mouth choristomas. The mean age at the time of diagnosis was 1.8 months (range, birth to 2 years). The mean age at time of surgery was 12.7 months (range, 6 days to 6 years). Males outnumbered females by approximately a 2:1 ratio (10 males and 6 females). Lesions were evenly divided in location between the tongue (n = 9) and floor of mouth (n = 7). Symptoms were present in five of 16 patients (31%) and included difficulty feeding (4 of 16, 25%), swelling with upper respiratory infection (1 of 16, 6%), and partial airway obstruction (2 of 16, 12%). One patient with airway obstruction required urgent tracheotomy followed by excision of the lesion under the same anesthetic.

#### Radiology

Seven patients (44%) underwent preoperative contrast-enhanced magnetic resonance imaging (MRI), five patients (31%) underwent contrast-enhanced computed tomography (CT), one patient (6%) had a lateral neck x-ray, and four patients (25%) had no imaging. One patient underwent both MRI and CT. One patient had a preoperative CT that could not be reviewed because it predated our Picture Archiving and Communication System.

Parameters for CT imaging varied with changes in scanner technology; axial images varied from 3- to 1-mm thickness, and multiplanar reformatted images were available only for thin-section images from multidetector scanners. The MRIs consisted of varying pulse sequences, but T2-weighted, T1-weighted, and post-contrast T1-weighted images in multiple imaging planes were consistently included. Fat suppression was applied to at least some of the postcontrast sequences in every case.

The lesions were consistently well-defined on imaging, with lobulated margins. The mean and standard
deviation of the greatest lesional diameter were 29 ± 10
mm (range, 17–47 mm). CT imaging revealed all of the
four masses (100%). On CT, three of the four lesions
(75%) demonstrated homogenous fluid density, and the
fourth lesion demonstrated an internal fluid-debris level.
No surrounding enhancement was seen on any of the
four scans.

On MRI, five of six lesions (83%) were nonenhanc-
ing; the sixth lesion had been previously treated with
sclerotherapy and demonstrated surrounding enhance-
ment, presumably from scar tissue. T2-weighted images
were homogeneously isointense to cerebrospinal fluid
(CSF) on the top and isointense to muscle on the bottom.
T1-weighted images showed variable signal: hyperin-
tense to muscle for two of six patients (33%), isointense
to muscle for one of six patients (17%), and hypointense
for one of six patients (17%). Three of six patients (50%)showed signal heterogeneity on T1-weighted images; two
patients had debris levels with iso- and hypointense
components, and one patient had two small antidepen-
tant nodules of T1 hyperintensity that suppressed with
fat saturation. MRI revealed trans-spatial spread of the
lesion in two of six cases (33%).

Operative Management

Complete surgical excision of the lesion was per-
formed in 15 of 16 patients (94%), including one with
anterior glossectomy (6%) and one excision via YAG
laser (6%). One patient underwent marsupialization of
the lesion rather than excision (1 of 16, 6%). No compli-
cations occurred at the time of surgery or immediately
postoperatively. No recurrences of choristoma were pres-
ent in those patients that had follow-up (10 of 16, 63%,
range 0.25–12 months).

Pathology

Gross pathology revealed intact cysts in nine of 16
specimens (56%) and disrupted (collapsed or fragmented)
cysts in seven of 16 specimens (44%). Fourteen of the 16
lesions (88%) had a unilocular cyst, and two had mul-
ple cysts, each with two cystic cavities. The mean size
of the greatest dimension of the submitted tissue was 24 ±
12 mm (range, 6–45 mm). Eight of 16 (50%) specimens
had either hemorrhagic, turbid, mucoid, or clear fluid or
cheesy material associated with the cyst wall whether or
not the cyst was received intact or fragmented. The most
common epithelium represented in these cysts was
ciliated, pseudostratified epithelium akin to sinonasal/res-
piratory type epithelium, present in 15 of 16 specimens
(94%). This type was followed by squamous epithelium
(12 of 16, 75%) and gastric epithelium (10 of 16, 62%).
Seven of 16 specimens (44%) contained all three epithe-
lium types; two specimens (12%) had only ciliated-
respiratory type, and one specimen (6%) had only gastric
epithelium. Other elements present included smooth
muscle (5 of 16, 31%), adnexal structures (2 of 16, 12%),
and rarely neural (motor unit) elements (1 of 16, 6%).

Minor salivary glands and striated muscle were also
present in and around some lesions but may have repre-
sented tissue normal for location. Final pathologic
diagnoses included lingual choristoma (n = 6), benign
cyst (descriptive) (n = 4), foregut duplication cyst (n =
3), enteric cyst (n = 1), congenital teratoid cyst (n = 1),
and gastric heterotopia (n = 1).

Three individual cases are presented that illustrate
pertinent clinical, radiographic, and pathologic findings:

Case 1

This patient is a 6-month-old female with a ventral
tongue cyst noticed at birth that resulted in feeding dif-
culties. On physical examination, a cystic tongue mass
was observed located in the midline with extension later-
ally (Fig. 1A). Preoperative MRI demonstrated a
nonenhancing, well-defined mass in the anterior aspect
of the tongue that was hypointense to muscle on T1-
weighted imaging and isointense to CSF on T2-weighted
imaging (Fig. 1B and 1C). The mass had a lobulated bor-
der and was homogenous in signal intensity on all
sequences, measuring 17 mm in diameter at its greatest
extent. Intraoperatively, after nasotracheal intubation,
the capsule was carefully dissected away from the
tongue (Fig. 1D). The resulting defect in the anterior
ventral tongue was closed with bilateral local advance-
ment flaps. The patient was successfully extubated on
postoperative day 1. Pathology revealed fibromuscular
tissue surrounding a cyst that had a respiratory-type
epithelial lining consistent with a lingual choristoma
(Fig. 1E). Focal areas of salivary glands and salivary
duct profiles were also seen (not shown). Follow-up 6
weeks postoperatively demonstrated a well-healed
tongue with good mobility. The patient's dysphagia had
resolved and there were no dyspnea or airway issues.
There was no evidence of recurrence of the lesion.

Case 2

This patient is a 2-month-old female with a cystic
tongue mass diagnosed in utero at 20 weeks of gestation.
The lesion did not appear to be compromising the air-
way, and the patient was successfully delivered vagi-
nally. The patient was asymptomatic, and physical
examination revealed a cystic mass extending from the
floor of mouth to the base of tongue. A CT scan with con-
trast revealed a 37-mm nonenhancing, multiloculated
cystic structure that displaced the tongue posteriorly
and superiorly. The lesion was trans-spatial and
extended across the mylohyoid muscle into the sublin-
gual space, adjacent to the anterior belly of the digastic
muscle. Cyst contents were homogeneous and of fluid
density (Fig. 2A and 2B). The trans-spatial nature of the
mass suggested a radiologic diagnosis of lymphatic
malformation.

After nasotracheal intubation, the cyst was com-
pletely excised via careful dissection around the cyst
wall. Pathology revealed a cystic cavity lined by squa-
mous, respiratory-type, and gastric epithelium (Fig. 2C).
The patient was successfully extubated on postoperative
day 2. At 2-week and 4-month follow-up, she was feeding well with normal tongue movement and no evidence of recurrence.

**Case 3**

This patient is a 13-month-old male with a recurrent cyst on the left side of the oral tongue. The cyst was first noticed at birth and was aspirated monthly for approximately 6 to 7 months by an otolaryngologist at an outlying hospital. The patient subsequently underwent three courses of sclerotherapy at Children’s Hospital of Pittsburgh by interventional radiology for a presumed lymphatic malformation. The final sclerotherapy treatment resulted in lingual edema necessitating intubation and stay in the intensive care unit. However, the cyst recurred, and examination revealed a large cystic mass palpated on the left floor of mouth and ventral tongue, displacing the tongue superiorly. MRI revealed a 47-mm, heterogeneous, lobulated cyst that was isointense to muscle on T1 sequence and had both iso- and hypointense components on T2 sequence. Two small nodules in the anterior aspect of the cyst were hyperintense to muscle on T1 imaging and were antidependent in their position. These nodules were not visualized with fat-suppressed, T1-weighted sequences, suggesting that they contained fat. Thus, the preferred radiologic diagnosis was dermoid cyst. Peripheral enhancement was seen, presumably attributable to scarring from the previous interventions (Fig. 3A and 3B).

Prior to this patient’s operation, the patient was orotracheally intubated with difficulty due to the location and size of the mass. Although he had a grade I view, laryngoscopy was attempted four times by the anesthesia team without success. Eventually, the patient was intubated with a 2.0 uncuffed tube using the

---

**Fig. 1.** A 6-month-old female with an anterior cystic tongue mass. (A) Intraoral examination reveals a cystic mass located in the ventral midline of the tongue. (B) Midline sagittal T2-weighted magnetic resonance image reveals a well-defined homogeneous mass (arrow) that is isointense to cerebrospinal fluid. (C) Gadolinium-enhanced axial T1-weighted image demonstrates a nonenhancing cystic mass (arrows) that is hypointense to muscle. (D) Intraoperative view after partial excision of the cyst. Mucosa of the ventral tongue, residual cyst lining, and edge of tongue musculature are shown. (E) Portion of cyst wall showing epithelial lining of pseudostratified ciliated respiratory-type epithelium (arrow). The cyst wall contains skeletal muscle fibers from the tongue (hematoxylin and eosin, ×100).

**Fig. 2.** A 2-month-old female with a multilocular tongue mass. Axial (A) and sagittally reformatted (B) postcontrast computed tomography demonstrates a nonenhancing, well-defined multilocular mass (arrows) extending into the sublingual space (arrowhead). The multilocular trans-spatial nature of the lesion suggests lymphatic malformation. (C) Cyst wall shows hybrid epithelium transitioning from ciliated respiratory-type (upper left) to gastric foveolar (lower right) (hematoxylin and eosin, ×200).
Seldinger technique over a rigid telescope. Intraoperatively, the cyst was removed by careful dissection. However, the wall was ruptured during the procedure, and the contents of the cyst were suctioned. The pathology showed ciliated-respiratory-type, squamous, and gastric epithelium (Fig. 3C). This patient was extubated on postoperative day 2. At 1-month follow-up, no recurrence was seen and the patient was feeding normally with normal tongue movement.

DISCUSSION

The differential diagnosis of pediatric oral cavity cysts is broad and includes mucocele, lymphatic malformation, venous malformation, and dermoid cyst. Inclusion of oral cavity choristomas in the differential diagnosis of pediatric oral cavity cysts is important because of potential complications with misdiagnosis. Case 3 illustrates this point; the patient underwent multiple courses of aspiration and sclerotherapy for a presumed lymphatic malformation that eventually resulted in a brief stay in the intensive care unit for airway compromise. In this review, we describe a series of 16 patients from one children’s tertiary care center with oral cavity choristomas in the past 19 years.

The majority of our patients were asymptomatic at presentation. However, patients can present with a large enough mass to cause feeding and airway problems. Choristomas of the oral cavity (and oropharynx and hypopharynx) should always be considered in the differential diagnosis of lesions causing neonatal airway obstruction. Other lesions that can cause neonatal airway obstruction include venolymphatic malformations, congenital granular cell tumor (epulis), teratoma, salivary gland anlage tumor, melanotic neuroectodermal tumor of infancy, congenital mucoceles or ranulas, and others.9

As part of the preoperative evaluation, physical examination can usually sufficiently localize the lesion and determine its cystic nature. Supplemental studies including radiographic imaging can be helpful to delineate the nature of the lesion (if in doubt) and the location of the lesion in relation to other anatomic structures, thus providing guidance for operative intervention. Also, imaging can further delineate the extent of lesions, particularly if trans-spatial extension is present. MRI and CT scans are the most common modalities used. Although CT is more readily obtained, MRI provides superior delineation of the extent and character of the lesion. As demonstrated in this case series, oral cavity choristomas appear as cystic masses that do not enhance with intravenous contrast.

One patient in our series demonstrated peripheral enhancement on MRI, which we attribute to prior sclerotherapy. These cysts have high signal, at least in part, on T2-weighted sequences. Patients with debris within their cysts will demonstrate a fluid-debris level in the center of the cyst. T1-weighted images show unpredictable intensity within the cyst, presumably related to variable cyst contents that include proteinaceous material from prior infection, hemorrhage, or mucoid fluid produced by the cyst wall epithelium or ectopic salivary gland present in some lesions. Thirty percent of patients in our series with imaging had heterogeneous cystic interiors, and 20% of patients had cyst with a trans-spatial component, findings more characteristic of lymphatic malformations. One patient had two small antidependent nodules in the anterior aspect of the cyst that were hyperintense to muscle on T1-weighted imaging but were hypointense after fat suppression. This finding could easily be mistaken for the fat lobules typically present in dermoid cysts.

The radiologic differential diagnosis for cystic lesions of the oral cavity includes dermoid cyst, lymphatic malformation, and choristoma. Although each of these lesions traditionally has their own specific radiographic characteristics, they frequently overlap and can be indistinguishable on imaging.

For preoperative assessment of oral cavity lesions in children and infants, we advocate MRI rather than CT when possible. MRI provides superior soft-tissue resolution compared to CT, provides accurate anatomic assessment of extent of the lesion, does not use ionizing radiation, and has a better chance of determining a
specific diagnosis. We note, however, that for logistical reasons including ease of scheduling and the possibility of avoiding general anesthesia, it may be more practical to obtain a CT scan.

Almost all oral cavity choristomas are diagnosed in childhood, but these lesions will occasionally be encountered in adults.10 In this scenario, the prognosis and treatment options are the same, as is the radiologic evaluation.

The treatment of choice for oral cavity choristomas is complete excision with removal of the mucosal lining. Aspiration alone results in recurrence, as functional mucosa continues to secrete fluid. Malignant transformation has been reported in one case of a long-standing lingual foregut duplication cyst in a 61-year-old man.11 Acid-producing gastric mucosa also carries the risk of ulceration and bleeding.12 Finally, untreated cysts can lead to fistula formation with chronic mucus secretion.13 Although our institutional experience also includes treatment with marsupialization and YAG laser excision, these patients were unfortunately lost to follow-up. The long-term prognosis of these lesions is excellent with proper treatment, with no recurrences seen in our series.

Most of the specimens reviewed in this series demonstrated at least 2 types of epithelium, usually respiratory-type with either squamous or gastric epithelium. Only one cyst was lined solely by gastric epithelium. This finding contrasts somewhat with the experience of Manor et al., who identified a total of 53 cysts in the literature from 1942 to 1997.6 Their findings revealed a combination of respiratory and either gastric or intestinal epithelium in roughly 31% of cases and gastric or intestinal epithelium alone in 43% of cases. Meyer in 1955 proposed a classification of sublingual cysts based on the lining epithelium into epidermoids (epithelial lining, no adnexa), dermoids (with adnexa), and teratoid (dermoids with connective tissue elements and/or respiratory or gastrointestinal epithelium).14 Using this classification, all lesions in this series would be considered teratoid. From the pathology perspective, the more common differential diagnoses in this age group and location include ranula, mucus extravasation phenomenon, thyroglossal duct cyst, vascular lesions, and hamartomatous lesions.9

The exact pathogenesis of these lesions is debatable. One theory suggests fusion of pluripotent embryonal rests with developing tongue musculature that when exposed to different environments can differentiate into respiratory or gastrointestinal epithelium.15 Another less accepted theory is the split notochord theory, where gut endoderm abnormally adheres to neural tube-derived ectoderm, which then herniates through a split notochord, leading to a neuromeric fistula.16

Two case series of choristomas in the head and neck have been published in the last 10 years, one in 2001 by Eaton et al. from Children’s Medical Center of Dallas and the other in 2010 by Kieran et al. from Children’s Hospital Boston.13,16 Our series had a similar male predominance (62%) and mean age at time of surgery (12.7 months). However, in contrast to these prior reports, our study discusses specific radiographic findings such as heterogeneous cystic interiors and trans-spatial components that may mislead the clinician in diagnosis. Furthermore, our series reports a significant (44%) proportion of specimens that contain a combination of respiratory type, gastric, and squamous epithelium. Only a minority of cysts presented with single-type epithelium (3 of 16, 19%). Similarly to these other two series, we report no cyst recurrences.

Our institutional experience also includes alternative methods of surgical excision, including anterior glossectomy and the use of a YAG laser. One patient in our series had a lingual choristoma addressed by marsupialization, although this was an older case in our series in which the pathology was not known or suspected at the time of the surgical procedure. We advocate complete surgical excision, not marsupialization, as the surgical treatment of choice for oral cavity choristomas.

The primary weakness in this study is a lack of follow-up longer than 1 year for any of the patients in our series. Six patients did not have any reported follow-up, including two patients with alternative methods of excision (YAG laser and marsupialization). It is, however, likely that any patients with evidence of recurrence would have returned to our clinic for evaluation, particularly because we are the only tertiary-care children’s hospital in the region. Because there are no reports of recurrence in the literature with complete surgical excision, long-term follow-up greater than 1 year may not be necessary. In fact, in our recent experience, we have advised patients to return to clinic only as needed after a 1-month follow-up if there were no complications.

Because of confusion in diagnostic terminology along with the unknown pathogenesis of these lesions, it may be more useful to differentiate these cysts by using descriptive and specific histologic terms. This nomenclature would provide a more objective aid to the clinician for subsequently identifying literature on the same cysts that are now classified by using the current myriad of different terms. Thus, oral cavity choristomas could be differentiated as lingual/floor of mouth choristoma with respiratory epithelium, lingual/floor of mouth choristoma with gastric epithelium, or lingual/floor of mouth choristoma with respiratory and gastric epithelium.

CONCLUSION

Oral cavity choristomas are rare lesions that typically present in the neonate or infant and can be associated with significant feeding problems or airway compromise. Deep oral cavity lesions should be imaged preoperatively, preferably with MRI, to assess their anatomic extent. Oral cavity choristomas are cystic radiographically but may be impossible to distinguish from the more common diagnoses of lymphatic malformation and dermoid cyst. Pathologically, lingual/floor of mouth choristomas are composed of usually two or more distinctive types of epithelium of ectodermal or endodermal derivation. To simplify diagnostic terminology, we suggest using specific descriptive terms for these lesions. Complete surgical excision is usually curative.
BIBLIOGRAPHY