

Enteric duplication cyst of the tongue in a newborn: A case report and literature review

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Abstract

Enteric duplication cysts (EDCs) are congenital anomalies. We report the case of a 5-day-old girl with a 2.0 cm congenital oral mass in the tongue, an extremely uncommon site of EDC. The tongue mass was resected without complication; microscopic findings were characteristic of an EDC, and the patient is doing very well. The English literature was researched for the cases of single congenital oral cavity masses diagnosed prenatally or at birth as EDC or EDC-like lesions. Cystic lesions of the oral cavity partially lined by gastrointestinal epithelium, without teratoma features, have received several names. Similarities between EDC and EDC-like lesions favor the idea of one lesion with several morphologies manifest along a spectrum and that the necessity for EDC diagnosis of the smooth muscle coat criteria could be re-evaluated to improve the categorization of these lesions and better understand the pathogenic mechanism.

Keywords: Case report 4, congenital tongue tumors 2, oral cyst 3, tongue neoplasms 1

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INTRODUCTION

Enteric duplication cysts (EDCs) are rare, congenital anomalies that usually present in the perinatal period and can be found anywhere along the gastrointestinal tract. There is a slight predominance in males and an overall incidence in the whole gastrointestinal tract of 1:4500 in autopsy case series.^[1] Enteric duplications are cysts or tubular structures that often share the muscular gastrointestinal wall, laying alongside the intestinal tube, but they can also lie outside the gastrointestinal tract.^[2] The tongue is an extremely unusual site for EDCs, representing 0.3% of all EDCs^[3] with a few published cases in the English literature. Here, we present the case report of a congenital tongue lesion, partially lined by gastric, respiratory, and squamous type epithelia, partially

covered by smooth muscle coat, and diagnosed as an EDC.

CASE REPORT

A 5-day-old girl was brought from a rural area to the maxillofacial surgery department, due to a congenital oral mass that was hindering the feeding process. Her mother, a previous healthy 16-year-old woman who did not attend regular prenatal clinical visits. Toxoplasma (IgM-IgG), Hepatitis B virus, HIV, and remaining routine laboratory tests ruled out prenatal infection, and thyroid stimulating hormone was within normal range. The patient was born at 38 weeks gestation through spontaneous vaginal delivery, with an appearance, Pulse, Grimace, Activity, and

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Respiration score of 9¹/10⁵, a birth weight of 2550 g, a size of 44 cm, and a cephalic perimeter of 32 cm. Physical examination was unremarkable except for a 2.0 cm cystic, nonpulsatile, renitent, translucent lesion in the tip of the tongue [Figure 1], with an implantation base of equal size, that was hindering mouth occlusion, suction development, and the breastfeeding process. The tongue mass was resected without complications, allowing the initiation of normal breastfeeding. The patient was discharged without any further interventions and is doing very well in the follow-up evaluations regarding weight gain and neurodevelopment.

The resected mass was sent to the pathology laboratory for analysis. Upon macroscopic examination, the 2.0 cm cystic lesion was translucent, unilocular, filled with a clear, watery fluid, possessed no papilla on the external or internal surfaces and had an average wall thickness of 0.2 cm. The microscopic analysis shows a benign cystic lesion, whose internal surface was covered by epithelial cells, most of them squamous admixed with respiratory and antral type epithelia. The wall of the cyst was composed of dense connective tissue with thin strands of striated and smooth muscle, and congestive small vessels. No teratoma elements were identified, and the diagnosis of an EDC was rendered.

DISCUSSION

The endoderm is an epithelial layer of the embryonic trilaminar germ disc that lines the innermost surface of the embryo, from the buccopharyngeal membrane to the cloacal membrane. Its relationship with the development of many organs and accessory glands in the head and

neck region, and the respiratory and gastrointestinal tract, highlights the plasticity of endodermic epithelial cells. One expression of this plasticity is the diversity of endodermally derived epithelium such as squamous, gastric, respiratory and intestinal, which can be found alone or mixed, lining benign cystic lesions.^[4] Several mechanisms have been proposed as theories for EDC or EDC-like lesion pathogenesis, relying on the presence or persistence of endodermally-derived components in unusual locations. Those mechanisms include the misplacement of embryonal rest,^[5] the local differentiation or metaplasia of pluripotent epithelial cells,^[6] the incomplete coalescence of the lacuna between epithelial cells in the elongating, developing gut,^[7] and the presence of buds of intestinal epithelium or endoderm within the submucosal tissue,^[8] among others. The most widely accepted theory for EDC development was elucidated by Veeneklaas, whose theory is based on the observation of an association between clefts and rib anomalies and EDC.^[9] As a result of these derangements in notochord development, Veeneklaas proposed that EDC can arise from entrapped endoderm tissue during notochord plate infolding. To reinforce Veeneklaas' theory, Qi *et al.*, induced EDC via Adriamycin injection in pregnant Sprague-Dawley female rats, finding that notochord anomalies such as esophageal atresia/tracheoesophageal fistula were related to EDC.^[10] In this experimental survey, the muscle coat in EDC was not observed in young embryos, suggesting that the smooth muscle coat may be a part of the maturation process of the lesion and that a real EDC may or may not be covered by the muscle coat. Oral EDCs are not commonly associated with malformations and Veeneklaas' EDC pathogenesis theory cannot explain mouth EDCs. Hence, all theories proposed so far have failed to describe a mechanism common to the diverse anatomic locations in which EDCs can occur and thus there may be distinctive pathogenic mechanisms related to specific anatomic locations of EDCs.

Cystic lesions of the oral cavity partially lined by gastrointestinal epithelium, without teratoma features, have received several names such as “EDC, s” “foregut duplication cysts,” “gastric duplication cysts,” “heterotopic gastrointestinal cysts” and “lingual choristoma.” All these lesions are very similar from a histological and clinical point of view are cystic lesions that infrequently involve the mouth cavity or the tongue and can be lined not just by gastrointestinal epithelium but also by a mix of gastrointestinal and respiratory epithelia. The term EDC was coined by Fitz in 1884 in his book of persistent omphalomesenteric remains,^[11] then in 1927 Schultz and Toyama published several descriptions of an intra-oral cyst lined by respiratory and gastric epithelia.^[12] Later, Gorlin

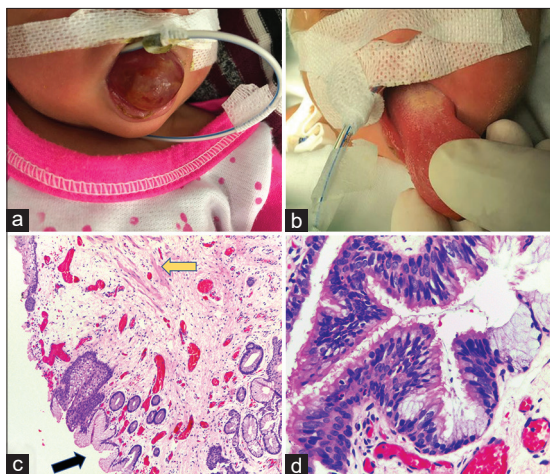


Figure 1: (a and b) Preoperative clinical images of the congenital tongue mass. (c and d) Microscopic findings of the cystic lesion (H & E, x100), lined by a mixture of squamous, respiratory and antral type of epithelium (black arrow) with a discontinuous smooth muscle coat (yellow arrow) on the wall

and Jirasek,^[13] Brown and Lister,^[14,15] published work on similar clinical and histological lesions but with different denominations, limiting the grouping of the cases. In the semantic chaos, Brown^[15] proposed that an EDC must fulfil three criteria: (1) it must have a coat of smooth muscle, (2) it must be attached to some part of the alimentary tract and (3) it must have a mucosal lining similar to that of some part of the alimentary tract. These three criteria were proposed to differentiate EDCs from other cystic lesions lined by epithelial components of the foregut. In the three criteria proposed by brown, the hallmark of an EDC is the presence of smooth muscle tissue in the cyst wall. It is possible that EDCs with the smooth muscle coat or EDC-like lesions without it are, broadly speaking, the same lesion with slightly different histological characteristics

associated with the tumorigenic mechanism or time of evolution, although ultimately a benign cystic lesion lined by endodermally derived epithelium.

We reviewed the English literature looking for single congenital oral cavity masses diagnosed prenatally or at birth, that were partially covered by gastrointestinal, respiratory, squamous, or a mixture of these epithelia, with or without muscle in the cyst wall, and absent teratoma features. We were able to retrieve 42 cases including the current case that are summarized in Table 1. From the 42 cases, the male/female ratio was 2:1, the average size of the masses was 2.8 cm, the tongue and the mouth floor were almost equally involved, and there were no differences regarding sex and mass location. Regarding the epithelial

Table 1: Resume of published cases of congenital mouth masses covered by endodermally-derived epithelium

Author (year)	Sex	Birth weight (g)	Location	Mucosa	Size	Diagnosis	Reference
Lister and Zachary (1968)	Female		Tongue	G		Cystic duplication	[14]
Ortiz et al.(1982)	Male		FOM	G		Heterotopic mucosa	[18]
Burton et al.(1992)	Male	ND	Tongue	G	0.5	Gastric choristoma	[6]
Surana et al.(1993)	Male	ND	Tongue	G	1.5	Heterotopic GC	[19]
LaBagnara et al.(1993)	Male	3090	FOM	G-I	5	ED	[2]
Morgan et al.(1996)	Female	ND	FOM	G-I	3	Heterotopic GC	[12]
Chen et al.(1997)	Female	ND	FOM	E	5	EDC	[20]
	Female	ND	Tongue	E	2.5	EDC	
Corić et al.(2000)	Male	ND	FOM	I-R	2.5	Oral GI cyst	[8]
Eaton et al.(2001)	Male	ND	FOM	G	1.8	FDC	[3]
	Male	ND	FOM	G	2.4	FDC	
	Male	ND	Tongue	ND	2	FDC	
	Male	ND	FOM	G	2.4	FDC	
Mandell et al.(2002)	Female	ND	Tongue	G-R	3	Lingual choristoma	[21]
El-Bitar et al.(2003)	Male	ND	Tongue	G	2.7	FDC	[22]
Kadkade et al.(2004)	ND	ND	FOM	I	3	Heterotopic GC	[23]
Rousseau et al.(2004)	Male	3050	Tongue	E	4	EDC	[24]
Kong et al.(2004)	ND	3480	FOM	E	ND	FDC	[7]
Puvanewary and Cassey (2005)	ND	ND	FOM	E	2.6	FDC	[25]
Hall et al.(2005)	Male	5160	FOM	G-R	3	Tongue DC	[26]
Satish Kumar et al.(2006)	Male	2500	Tongue	G-I	5	Lingual DC	[27]
Aviram et al.(2009)	Female	3450	FOM	G	2	FDC	[16]
Davis et al.(2010)	Male	ND	Tongue	R	ND	FDC	[17]
Hambarde et al.(2011)	Male	3000	Tongue	G	4	FDC	[28]
Houshmand et al.(2011)	Female	3158	Tongue	G-I	4.5	FDC	[4]
	Female	ND	Tongue	G-I	3.3	FDC	
Blanchard et al.(2012)	Male	ND	Tongue	I-R	2.4	FDC	[29]
	Male	ND	FOM	I-R	ND	FDC	
	Male	ND	Tongue	I	ND	FDC	
Madan et al.(2012)	Male	3000	FOM	G	7	Lingual DC	[30]
Joshi et al.(2013)	Male	ND	Tongue	G-R	1.7	Lingual cyst	[31]
Garcia-Rosa et al.(2013)	Male	ND	FOM	R	ND	FDC	[32]
Oginni et al.(2014)	Male	3400	FOM	G	4	Epidermoid cyst	[5]
Gantwerker et al.(2014)	Female	4160	Tongue	G-R	3.5	FDC	[1]
Luo et al.(2015)	Female	ND	FOM	G-R	2	Lingual DC	[9]
Schrotenboer et al.(2016)	Male	ND	FOM	G-I-R	1	FDC	[33]
Méndez Sáenz et al.(2016)	Male	3870	FOM	G-I	3.6	Heterotopic GC	[34]
Knowles et al.(2017)	Male	ND	Tongue	G-R	0.5	FDC	[35]
Ginat et al.(2019)	Female	ND	FOM	G-I	3.7	Heterotopic GC	[36]
Lee et al.(2020)	Female	ND	FOM	G-I	2.5	Lingual GC	[37]
Shabani et al.(2020)	Male	2500	Tongue	G-I	5	FDC	[38]
Arteta (2021)*	Female	2550	Tongue	G-R	2.5		

*Actual case, FOM: Floor of the mouth, GC: Gastrointestinal/gastric cyst, ED: Enteric duplication, EDC: Enteric duplication cyst, GI: Gastrointestinal, FDC: Foregut duplication cyst, DC: Duplication cyst, G: Gastrointestinal, I: Intestinal, R: Respiratory, E: Enteric, ND: No data

lining, the gastric epithelium was the most common at 29%, followed by gastric and respiratory covering at 23%. There were no statistically significant differences between epithelial lining, location, and sex. There was just one case associated with a gross malformation, a right sclerocorneal microphthalmia,^[16] unrelated to the notochord plate, suggesting that congenital EDC or EDC-like lesions do not have a strong association with malformations. However, there is a case in which two brothers, 112 years old and the other a newborn, were both affected with oral EDC.^[17] This phenomenon was not reported in the remaining patients' mothers, fathers, or siblings, and thus there is no clear epidemiological support for EDC as an inherited trait. The clinical, macroscopic, and microscopic similarities between EDC and EDC-like lesions favor the idea of one lesion with several morphologies manifest along a spectrum, and that the smooth muscle coat criteria proposed by Brown could be re-evaluated to improve the categorization of these lesions and better understand the pathogenic mechanism.

Statement of ethics

The Institutional Human Ethics Committee approved this case report. Written, informed consent was given by the patient's mother.

Author contribution statement

AAA-ACP: Slide review and interpretation of pathology specimens. EAM: Clinical data acquisition and interpretation. All the authors were involved in drafting and revision of the manuscript, and approval of the final version.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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