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CASE REPORTS

Concomitant Congenital Intraoral Dermoid Cyst and Heterotopic Gastrointestinal Cyst

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Abstract
Sublingual dermoid cysts and oral heterotopic gastrointestinal cysts are well-documented causes of sublingual swellings in infants. However, the simultaneous occurrence of both cysts is an extremely rare phenomenon, with only two previous cases reported in literature. We present a case of 1-month-old neonate with a cystic ventral tongue mass that on histologic examination showed a dermoid cyst with co-existing heterotopic gastric-type epithelium in the wall. Additional histochemical stains revealed focal areas of colonic metaplasia within the heterotopic gastric epithelium. CDX2 immunohistochemical staining confirmed derivation from colonic epithelium.

Keywords Congenital sublingual cyst · Sublingual dermoid cyst · Oral heterotopic gastrointestinal cyst

Introduction

Sublingual dermoid cysts and oral heterotopic gastrointestinal cysts are well-documented causes of sublingual swellings in infants. However, the simultaneous occurrence of both cysts is an extremely rare phenomenon, with only two previous cases reported in literature [1, 2]. This case report highlights such an occurrence in a one-month-old male neonatal patient with a history of respiratory stridor since birth.

Case Report

A 1-month-old male neonatal patient, with a normal gestational age, presented with his mother to the Department of Otolaryngology at Steve Biko Academic Hospital, Pretoria, with a history of respiratory stridor since birth.

Physical examination was normal and there were no syndromic stigmata or signs of physical disease. On intraoral examination the patient had a concurrent cleft palate with laryngomalacia. A large ventral tongue mass was noted, appearing cystic in nature. This mass aspirated a whitish, keratin-like material suggestive of an epithelial inclusion cyst. Advanced radiographic imaging of the head and neck, in the form of computed tomography (CT), showed a well-defined thin-walled cyst measuring 18.9 × 29.9 × 21.8 mm at the base of the tongue (Fig. 1).

When faced with the abovementioned clinical presentation, a number of clinical differential diagnoses should be considered. The most common causes of a swelling in the floor of the mouth include the following:

- Ranula (mucous retention cyst);
- Thyroglossal cyst;
- Congenital vascular malformation;
- Ludwig's angina;
- Dermoid cysts;
- Cystic teratoma;
- Salivary calculi obstructing submandibular duct;
- Sialadenitis and;
- Oral alimentary/choristomatic tract cyst/oral heterotopic gastrointestinal cyst [1].

A clinical differential diagnosis of an epidermoid cyst or cystic teratoma was suspected in this case and the mass was subsequently excised and submitted for histological assessment.

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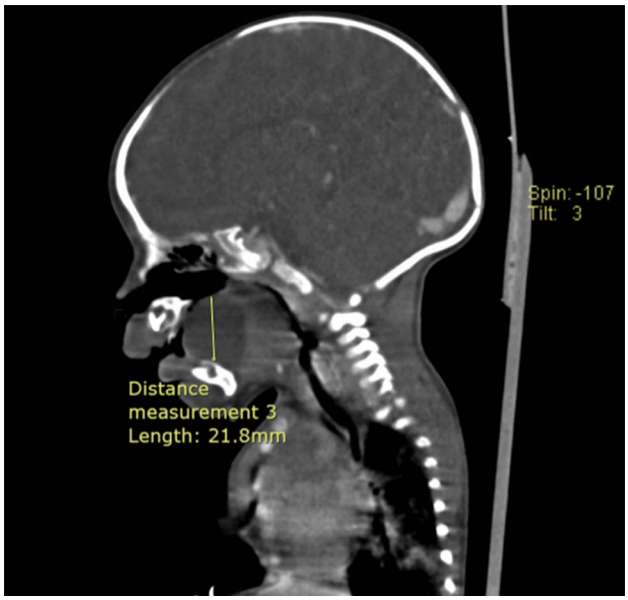

Fig. 1 CT scan showing the cystic lesion involving the base of the tongue

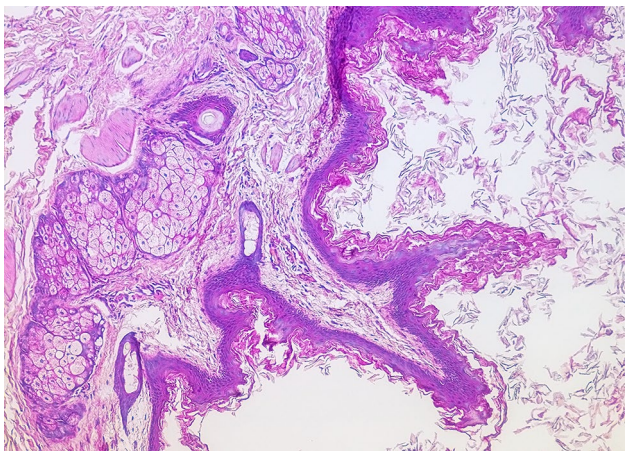

Fig. 2 Hematoxylin and eosin (H&E)-stained section showing the dermoid cyst component including skin adnexal structures (original magnification × 10)

Pathologic Findings

Gross examination of the specimen showed a ruptured cystic lesion measuring 2.5 × 1.2 × 0.6 cm, with a wall thickness of 0.1 cm.

Histological evaluation showed the presence of a true cystic lesion with adjacent residual glossal skeletal muscle. The cyst lining was characterized by keratinized stratified squamous epithelium with numerous skin adnexal structures, including hair follicles and sebaceous glands

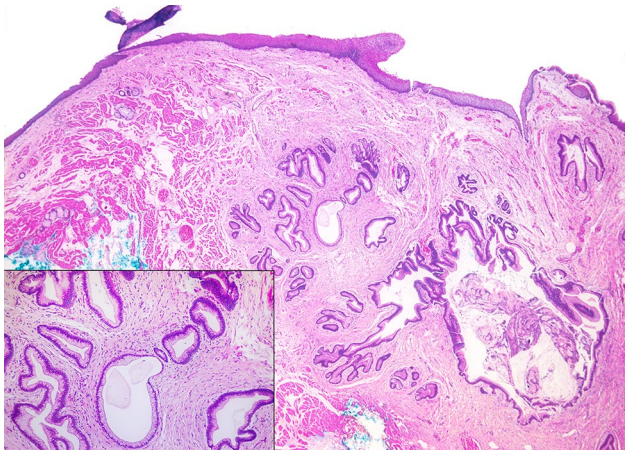

Fig. 3 A low-power H&E-stained section showing co-existing cystic structure (original magnification × 2). Insert: A high-power H&E-stained section showing gastric-type epithelial lining of the cyst (original magnification × 10)

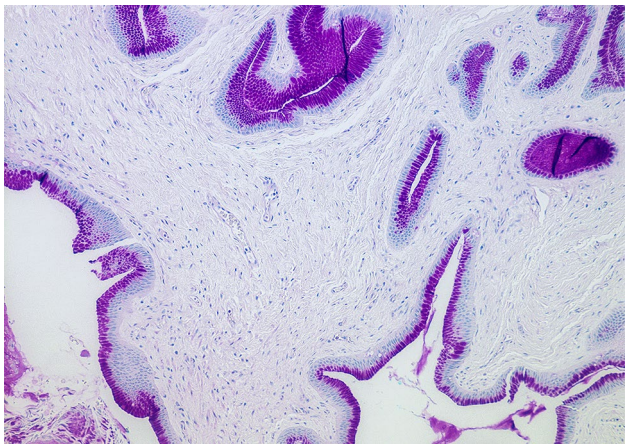

Fig. 4 A Periodic acid-Schiff (PAS)-stained section highlighting the neutral mucin within the gastric epithelium (original magnification × 10)

(Fig. 2). A co-existing finding, within the wall of the cyst, included the presence of heterotopic gastric-type epithelium (Fig. 3).

The presence of gastric epithelium containing neutral mucin was confirmed with the assistance of Periodic acid–Schiff (PAS) histochemical staining (Fig. 4). Additionally, focal areas of colonic metaplasia were also noted within the heterotopic gastric epithelium with the assistance of Alcian blue histochemical staining method (pH 2.5) followed by the conventional PAS technique (Fig. 5). CDX2 immunohistochemical staining showed positive nuclear staining in this area, confirming derivation from colonic epithelium (Fig. 6).

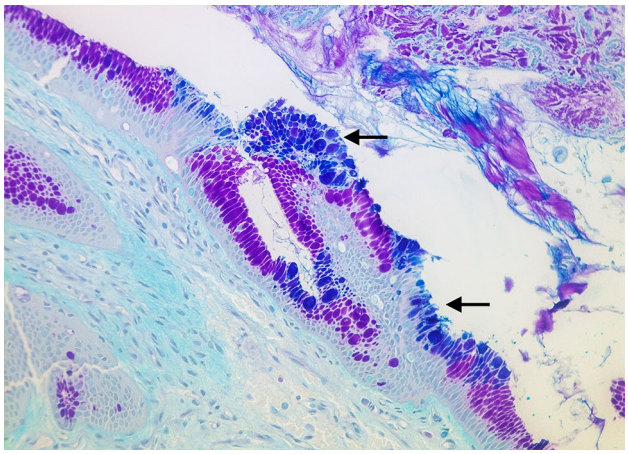

Fig. 5 An Alcian blue/PAS-stained section highlighting areas of colonic metaplasia within the gastric epithelium (black arrows) (original magnification × 20)

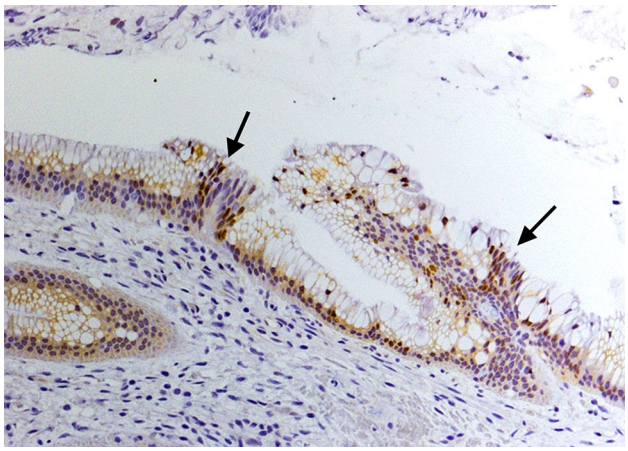

Fig. 6 A CDX2 immunohistochemical stain showing nuclear positivity in the areas of colonic metaplasia (black arrows) (original magnification × 40)

A final histologic diagnosis of a congenital dermoid cyst with an adjacent heterotopic gastrointestinal cyst was made.

Discussion

The most common cause of cystic masses affecting the head and neck region in children are developmental cysts. Of these, the thyroglossal cyst is the most common, followed by the branchial cleft cyst [3]. Most other cystic lesions are rarely encountered.

Dermoid cysts (DCs) are germ-cell-derived lesions that usually occur in relationship to the testes or ovaries, where they are referred to as mature cystic teratomas [1]. A review of 1,495 cases of dermoid cysts seen at the Mayo Clinic over a 25-year span revealed that only 7% of these cysts arose in the head and neck region, with 25% of these occurring in the floor of the mouth [1, 2, 4]. These cysts arise during the formation of the face and neck when a fragment of ectodermal tissue is trapped in the midline, eventually forming a sublingual dermoid cyst [1, 2]. The mechanism by which this ectodermal tissue becomes entrapped is not completely understood [3]. Shafer postulated that this entrapped tissue is totipotent and hence derivatives of all three germ layers may be present in the cyst [2].

Gibson and Fenton separated congenital intraoral dermoid cysts into three principle types: (1) epidermoid cyst—lined by stratified squamous epithelium without skin adnexal structures, (2) dermoid cyst—lined by stratified squamous epithelium with skin adnexal structures, and (3) teratoid cyst—lined by epithelium ranging from keratinizing squamous to respiratory epithelium with dermal appendages along with derivatives of all three germ cell layers (ectoderm, mesoderm and endoderm) [1, 4–6].

DCs typically present as an asymptomatic, slow-growing, soft mass without lymphadenopathy [7]. Their lumens usually contain abundant keratinous, caseous, sebaceous material and in some instances hair, fat and cholesterol [7]. The cyst can enlarge to a point that the patient presents with difficulty in articulation, mastication and deglutition, and airway compromise [2]. The principle histologic component in these cysts is keratinized, stratified squamous epithelium with adjacent dermal appendages. In most instances the diagnosis of a dermoid cyst is not difficult, however cases with concurrent infection may pose added complexity [8]. Complete surgical removal of the cyst is curative [4].

Teratoid cysts contain tridermic components within their walls originating from totipotential cells [9]. This type of tumour commonly occurs in the ovaries and testes as a benign cystic teratoma, but is exceedingly rare in the oral cavity [8, 9].

In the current case report, the cyst demonstrated a keratinized stratified squamous epithelial lining with numerous dermal skin adnexal structures. Additionally, within the wall of the cyst, was the presence of heterotopic gastric-type epithelium. The surrounding muscle fibers were interpreted as resident glossal skeletal muscle rather than a true mesodermal component, hence excluding the diagnosis of a true teratoid cyst.

Heterotopic gastric mucosa has been described in many areas of the gastrointestinal tract including the duodenum, gallbladder, common bile duct, jejunum, Meckel's diverticulum, ileum, appendix, colon and rectum [10, 11]. An oral heterotopic gastrointestinal cyst (HGC), which is also referred to as a gastric cystic choristoma, entrocytoma or enteric duplication cyst, represents an extremely rare lesion with a male–female ratio of 3:2 [1, 12, 13]. These cysts usually occur in neonates and infants, with about forty cases of

HGCs in the oral cavity being reported in literature [7, 12, 14]. Although rare, when occurring in the oral cavity, the usual location is the anterior two-thirds of the tongue [10]. The floor of the mouth is the second most frequently affected region [14]. This cystic lesion is characteristically lined by gastric (42%), intestinal (19%) and mixed gastric and intestinal epithelium (10%) [1, 4, 7]. Gastric epithelium may be similar to that of cardiac, fundus or pyloric regions of the stomach [10]. Histochemical studies highlight the presence of parietal, chief and mucous cells within the lining of these cysts [11]. Smooth muscle is usually identified surrounding the cyst [15]. Authors have also reported cases in which the mucin composition of the cyst was not in accord with that of the normal gastrointestinal tract [4].

The histogenesis of this lesion is unclear, but it is thought to arise from the entrapment of undifferentiated endoderm within the developing oral cavity, normally during the 3rd–4th week of fetal life [1]. The primitive stomach, in the early embryo, lies in the midneck region close to the anlage of the tongue [10]. This explains the presence of these lesions in the anterior two-thirds of the tongue. This theory however fails to explain the presence of intestinal and colonic mucosa in some of these cysts [12]. A mucin histochemical study of these cysts by Woolgar and Smith [16] in 1988 highlighted the presence of well differentiated columnar and goblet cells. This supports the theory that these cysts arise from primitive endodermal gastric mucosa under inductive influences [13, 15].

Clinically, these cysts are often discovered during infancy, with only 30% experiencing difficulty with feeding, swallowing, speech and breathing [12].

Conservative surgical excision is the treatment of choice, with reports of CO_2 lasers also being used for the excision [10, 11, 14]. Recurrence is uncommon, but has been reported in literature, most likely due to incomplete excision [14].

The simultaneous occurrence of dermoid and heterotopic gastrointestinal cysts in the oral cavity has been confined to two reported cases in literature [1, 4]. These cystic lesions, although rare, demand a prompt response from the clinical team to prevent any airway compromise. Interesting, in both reported cases the infants were born otherwise healthy. Several theories have hypothesized the possible histogenesis of these rare oral cystic lesions. Currently, the most widely accepted theory proposes origin from displaced germ cells during embryologic migration [4].

Literature emphasizes that the simultaneous occurrence of dermoid and heterotopic gastrointestinal cysts within the oral cavity should be considered as separate bilobular cystic masses [4]. This is supported by a failure to explain the development of dermoid cyst-like areas from embryonic gastric remnants [12]. The gastric/intestinal type epithelium was hence not considered as part of the spectrum of a dermoid cyst [4]. Another hypothesis extrapolates that dermoid cysts usually contain gastrointestinal epithelium, and, the adjacent heterotopic gastrointestinal cyst simply represents an invagination of the already present gastrointestinal epithelium [1, 4]. Other less accepted mechanisms include origin in the thyroglossal duct and salivary retention cysts [17].

Prompt surgical excision is the treatment of choice, with long-term follow-up recommended.

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