Co-existence of Congenital Epidermoid Cyst and Ranula in a Newborn. Report of a Unique Case

Erofili Papadopoulou[1], Efstathios Pettas[1], Lampros Gkoutzanis[2], Konstantinos Katoumas[2], Maria Georgaki[1], Emmanouil Vardas[1], Evangelia Piperi[1], Nikolaos G. Nikitakis[1]

[1]Department of Oral Medicine & Pathology and Hospital Dentistry, School of Dentistry, National and Kapodistrian University of Athens (NKUA), Athens, Greece.
[2]Department of Oral and Maxillofacial Surgery, School of Dentistry, National and Kapodistrian University of Athens (NKUA), Athens, Greece.

Corresponding Author:
Erofili Papadopoulou
Department of Oral Medicine & Pathology and Hospital Dentistry
School of Dentistry, University of Athens
2 Thivon St., Goudi 11527, Athens
Greece
E-mail: erofilipa@gmail.com

ABSTRACT

Background: Congenital cystic swellings involving the floor of the mouth include various lesions such as developmental cysts (e.g., dermoid and epidermoid cysts), ranulas, vascular malformations etc. However, coexistence of such conditions, possibly with a cause-and-effect- relationship, is rare. The purpose of this case report is to present a rare case of a congenital epidermoid cyst associated with a mucous retention cyst in a newborn.
Methods: A 6-month-old female infant was referred to an Oral Medicine Clinic in Athens, Greece on October 2019 for evaluation of a swelling at the floor of the mouth, first noticed by her paediatrician just after birth. Clinically, a yellowish "pearly" nodule in close association with the orifice of the left submandibular duct, posteriorly transitioning to a diffuse bluish cystic swelling of the left floor of the mouth was observed. With a provisional diagnosis of a dermoid cyst and/or ranula, a surgical excision was performed under general anaesthesia.
Results: Histopathologically, a well-defined, keratin-filled, cystic cavity lined by orthokeratinized stratified squamous epithelium was observed in the anterior aspect while posteriorly and in close proximity, a dilated salivary duct lined by cylindrical, cuboidal or pseudostratified epithelium was noted. A final diagnosis of an epidermoid cyst intimately associated with a mucus retention cyst (ranula) of the submandibular duct was rendered.
Conclusions: The coexistence of two cystic lesions in the floor of the mouth with features of epidermoid and mucous retention cyst, respectively, is rare and its pathogenesis intriguing, especially in a newborn.

Keywords: epidermoid cyst; mucocele; newborn; nonodontogenic cysts; ranula.

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INTRODUCTION

Congenital cystic lesions involving the floor of the mouth are rare entities encompassing a broad spectrum of pathoses, including developmental cysts (e.g. dermoid/epidermoid cysts), ranulas, vascular malformations etc. In a recent systematic review by Lucas et al. [1], out of 65 studies involving 85 patients with congenital lesions of the floor of the mouth, the most common lesion was dermoid cyst, followed by foregut duplication cyst, ranula, epidermoid cyst, and heterotopic gastrointestinal cyst.

Dermoid/epidermoid cysts are usually diagnosed in children or young adults, either incidentally or due to associated signs and symptoms, while presentation in infants is a more infrequent phenomenon [2,3]. Rarely, such cystic lesions may coincide with or facilitate the development of cyst-like dilation of salivary ducts or other reactive lesions such as mucoceles/ranulas, possessing diagnostic pitfalls.

The purpose of this case report is to present a rare case of a congenital epidermoid cyst associated with a mucous retention cyst in a newborn.

CASE DESCRIPTION AND RESULTS

A 6 month-old Caucasian, full term female infant in good general health was referred to an Oral Medicine Clinic in Athens, Greece, on October 2019 for evaluation of a 'pearly' yellowish swelling located in the midline of the floor of the mouth. The lesion was first noticed by her paediatrician just after birth, slowly enlarging since then according to her mother. Clinical examination revealed a soft, fluctuating, non-pedunculated, yellowish nodule covered by normal mucosa with a prominent vasculature (Figure 1A), in close association with the orifice of the left submandibular duct (Figure 1B), posteriorly transitioning to a diffuse bluish cystic swelling (Figure 1C). The rest of the oral cavity was within normal limits and no lymphadenopathy or other extraoral findings were detected. Ultrasound evaluation revealed an irregularly shaped cystic lesion with a maximum diameter of 5 mm and maximum craniocaudal lenght of 13 mm (Figure 2A, 2B). With a provisional diagnosis of a dermoid cyst and/or mucous cyst, surgical excision was performed

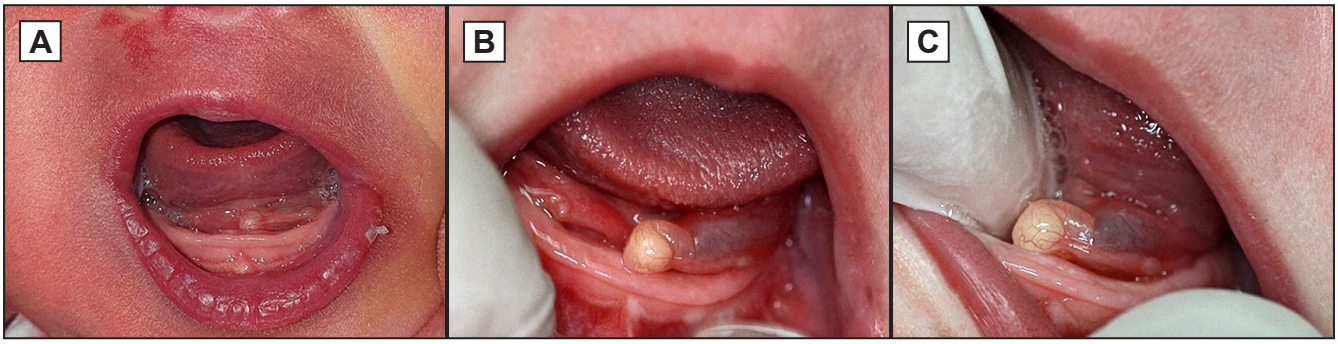

Figure 1. Clinical examination.
A = yellowish nodule covered by normal mucosa located in the midline of the floor of mouth; B = close association of the lesion with the orifice of the left submandibular duct; C = the lesion shows transition to a diffuse bluish cystic swelling posteriorly.

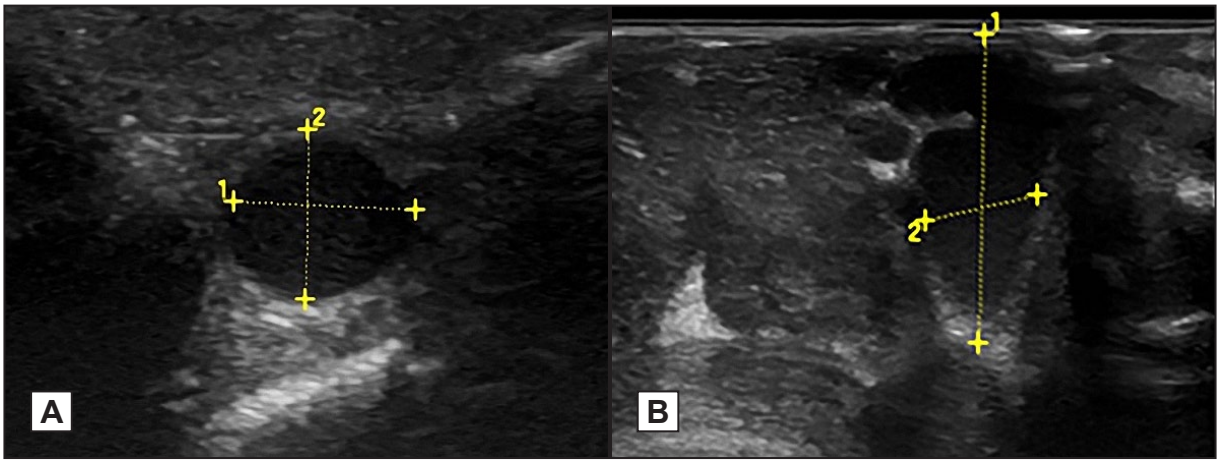

Figure 2. Ultrasonographic imaging.
A = an irregularly shaped cystic lesion of 5 mm maximum diameter; B = 13 mm craniocaudal length.

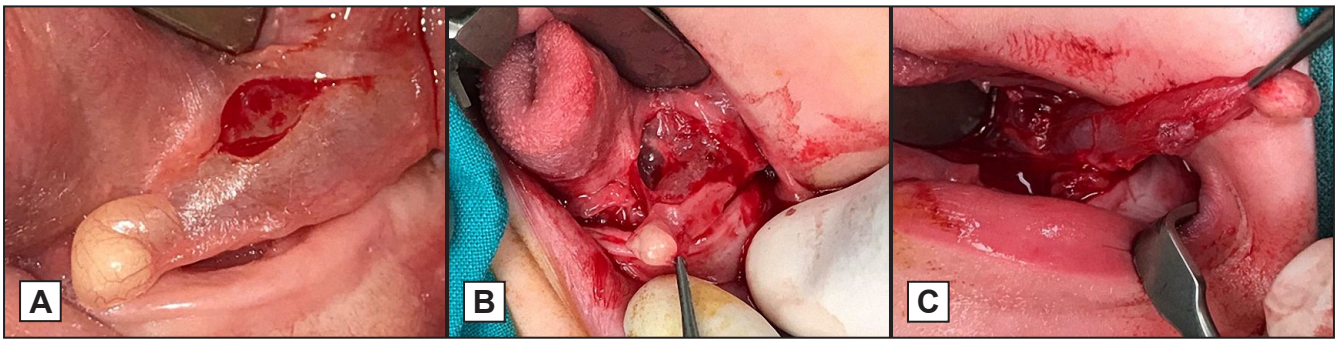

Figure 3. Intraoperative pictures: (A) anterior round lesion in continuity with a more diffuse cystic lesion (B), exhibiting significant posterior extension towards the submandibular gland (C).

under general anaesthesia. Intraoperatively, a round cystic lesion was identified anteriorly and in continuity with a second more diffuse cystic lesion. The latter exhibited a significant posterior extension towards the submandibular gland as an elongated cylindrical dilatation (Figure 3).

In summary, an oval shaped cystic nodule containing whitish creamy material was observed in the anterior aspect of the surgical specimen, transitioning posteriorly to another cystic lesion (Figure 4). Histopathologic examination revealed two cystic spaces in close proximity, both showing epithelial lining (Figure 5A). The anterior lesion was a large, well-delineated, keratin-filled, cystic cavity lined

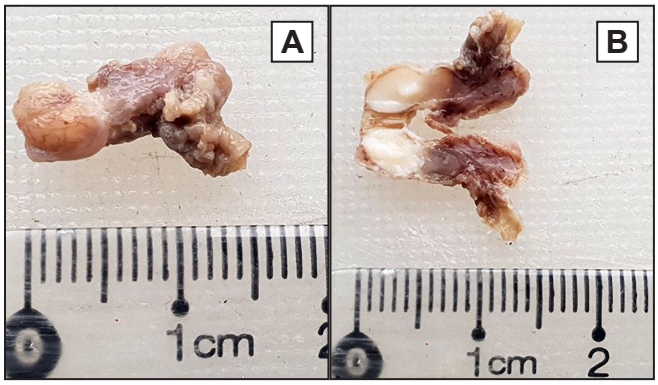

Figure 4. Gross examination: (A) oval-shaped cystic nodule containing whitish creamy material (anteriorly), transitioning to a brownish cystic process posteriorly (B).

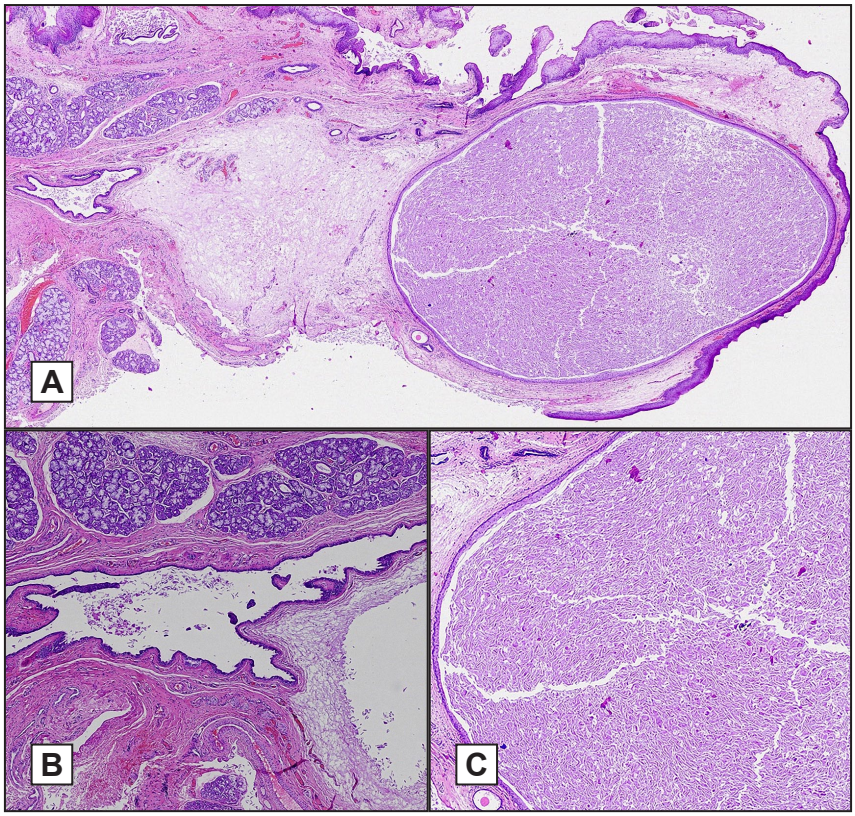

Figure 5. Histopathologic examination (haematoxylin and eosin stain, original magnification x20).
A = two large cystic spaces with epithelial lining; B = anteriorly, a keratin-filled cystic cavity with squamous epithelial lining; C = posteriorly, a cystic cavity with columnar or cuboidal cells in the epithelial lining.

by orthokeratinized stratified squamous epithelium with absence of dermal appendages in the fibrous cystic wall (Figure 5B), while posteriorly, a dilated salivary duct lined by single columnar, cuboidal or pseudostratified epithelium, partially containing amorphous eosinophilic material, was observed (Figure 5C). Multiple lobules of seromucinous minor salivary glands were also noted in the surrounding area. Taking into consideration the clinical and histopathological findings, a final diagnosis of an epidermoid cyst intimately associated with a mucous retention cyst of the submandibular duct was rendered. The postoperative course was uneventful with complete healing and 2 years postoperatively the patient remains free of any symptoms, while there has been no evidence of recurrence.

DISCUSSION

Dermoid/epidermoid cysts are rare developmental lesions considered as cystic forms of teratoma, with about 6.5% of them involving the head and neck region and 1.6% found in the oral cavity, accounting for less than 0.01% of all intraoral cystic processes [2,4]. They predominantly present at the midline of the floor of the mouth in children and young adults, while approximately 15% of them are congenital, although even then, diagnosis is usually rendered during the second and third decades of life, when cysts are large enough to become clinically evident [4,5]. These lesions are extremely rare in neonates and only about 30 cases of congenital epidermoid cysts of the floor of the mouth have been reported in infants so far [2,4-6].

Depending on whether these cysts are located above or inferiorly to the geniohyoid muscle, they may appear either as painless, yellowish, intraoral nodules with a rubbery texture or as submental swellings respectively. They usually show an indolent clinical course but when intraoral and of a considerable size, they may lead to serious complications such as tongue displacement with associated dysphagia, dysphonia, and even obstruction of the airway [4]. Both types of cysts are lined by an epidermis-like epithelium, with the dermoid type also showing dermal appendages (e.g. sebaceous and/or sweat glands, hair follicles) in the cystic wall [5]. Treatment of choice is surgical excision with a good prognosis and a low relapse rate [7,8].

Ranulas are most commonly acquired mucus cysts appearing either as asymptomatic bluish swellings off-midline in the floor of the mouth (simple ranula) or as plunging masses in the submandibular or submental area [9]. They are caused either by trauma resulting in a mucus extravasation cyst or by obstruction of a salivary duct (mucus retention cyst), the latter presenting histopathologically as a cystic cavity lined by 1 to 2 layers of cuboidal or columnar epithelium. Congenital ranulas are observed in about 0.74% of newborn infants and they are thought to result from congenital atresia of the salivary duct orifices of Bartholin's or Warthon's duct. Rarely and when large enough, ranulas may cause tongue displacement, feeding difficulties or even airway obstruction [9,10].

In paediatric population, intraoral dermoid/epidermoid cysts may clinically masquerade as mucous cysts [11]. However, multi-focality or coexistence with other congenital or reactive cystic lesions is extremely rare, with only sparse cases of epidermoid cysts co-existing with other intraoral developmental cysts reported in the literature so far such as with heterotopic gastrointestinal cyst [12], oral alimentary tract cyst [13], thyroglossal duct cyst [14], and in one case with two dermoid cysts. [6] To the best of our knowledge, the concomitant presence of an epidermoid and a mucous retention cyst has never been described before, with the exception of a concurrent extravasation mucocele and an epidermoid cyst of the lower lip reported in a 13-year-old boy [15].

The pathogenetic mechanism resulting in the concurrence of these two cystic lesions in the floor of the mouth is intriguing, especially in a newborn. Theoretically, the coexistence of these two lesions could be coincidental. Nevertheless, although the ranula in our case could also be congenital, there has been no recurrence that could suggest congenital atresia of the Wharton's duct, suggesting that its nature was rather reactive. A more reasonable explanation would be that a slowly growing congenital epidermoid cyst, incidentally developing adjacent to the submandibular duct orifice, could cause obstruction of the duct tract and a secondary mucous retention phenomenon.

CONCLUSIONS

The development of congenital cystic lesions in the floor of the mouth is uncommon, but the coexistence of more than one of these entities, possibly with a cause and effect relationship, is extremely rare. Increased awareness of these lesions by obstetricians and neonatologists is mandatory, since prenatal diagnosis of these lesions enables proper management and avoidance of complications such as respiratory compromise during delivery.

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The authors report no conflict of interest related to this study

Authors declared taking informed written consent for the publication of clinical photographs, from the legal guardian of the patient with an understanding that every effort will be made to conceal the identity of the patient, however it cannot be guaranteed.

Authors declared to fulfil authorship criteria as devised by ICMJE and approved the final version. Authorship declaration form indicating individual contribution, submitted by the authors is available with the editorial office.

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