

Research Article

## Concurrent Presence of Congenital Epidermoid Cyst and Ranula in a Newborn

Erofilia Papadopoulou<sup>1\*</sup>, Efstathios Pettas<sup>1</sup>, Lampros Gkoutzani<sup>2</sup>

<sup>1</sup>Department of Oral Medicine & Pathology and Hospital Dentistry, School of Dentistry, National and Kapodistrian University of Athens (NKUA), Athens, Greece.

<sup>2</sup>Department of Oral and Maxillofacial Surgery, School of Dentistry, National and Kapodistrian University of Athens (NKUA), Athens, Greece.

\***Correspondence:** Erofilia 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](mailto:erofilipa@gmail.com)

**Received:** February 08, 2023; **Accepted:** March 10, 2023; **Published:** March 18, 2023

**Citation:** Papadopoulou E, Pettas E, Gkoutzani L. Concurrent Presence of Congenital Epidermoid Cyst and Ranula in a Newborn. J Dent Oral Maxillofac Res. 2023;1(1):01-04.

**Copyright:** © 2023 Papadopoulou E. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

### 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 retentioncyst, respectively, is rare and its pathogenesis intriguing, especially in a newborn.

**Keywords:** Epidermoid cyst; Mucocele; Newborn; Nonodontogenic cysts; Ranula.

## 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.

## METHODOLOGY

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 length of 13 mm (Figure 2A, 2B). With a provisional diagnosis of a dermoid cyst and/or mucous cyst, surgical excision was performed 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).

## RESULTS

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.

The anterior lesion was a large, well-delineated, keratin-filled, cystic cavity lined by orthokeratinized stratified squamous epithelium with absence of dermal appendages in the fibrous cystic wall, while posteriorly, a dilated salivary duct lined by single columnar, cuboidal or pseudostratified epithelium, partially containing amorphous eosinophilic material, was observed. 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



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.

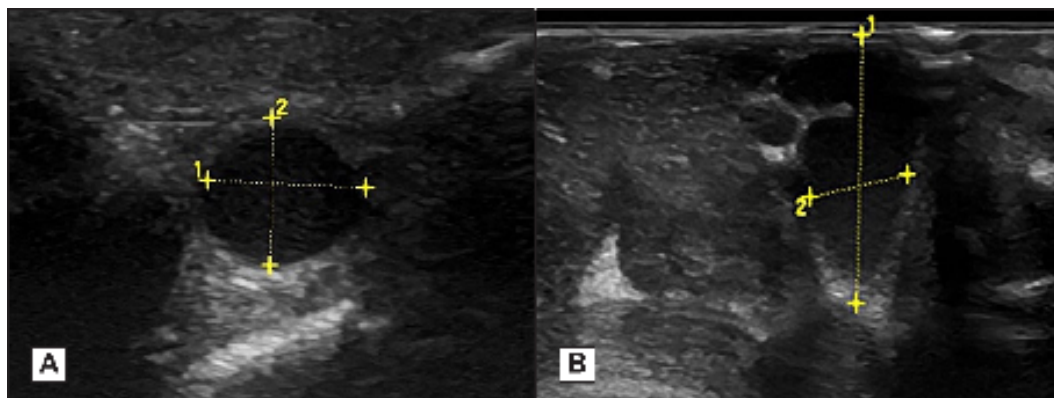


Figure 2: Ultrasonographic imaging.

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).

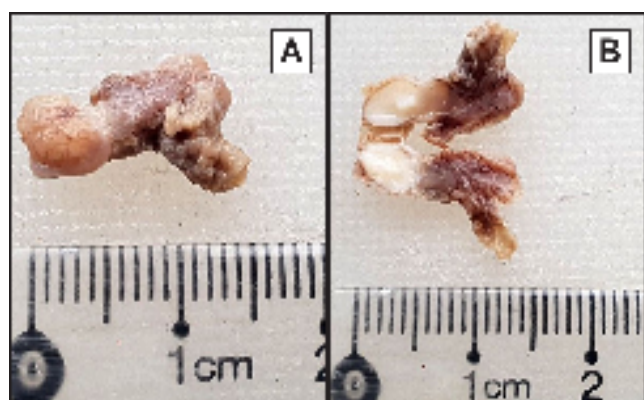


Figure 4: Gross examination: (A) oval-shaped cystic nodule containing whitish creamy material (anteriorly), transitioning to a brownish cystic process posteriorly (B).

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.

## CONCLUSION

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.

## REFERENCES

- Lucas JP, Allen M, Siegel B, Gonik N. Diagnosis and management of congenital floor of mouth masses: A systematic review. *Int J Pediatr Otorhinolaryngol.* 2021 Jan;140:110541.
- Mohta A, Sharma M. Congenital oral cysts in neonates: report of two cases. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.* 2006 Nov;102(5):e36-8.
- Misch E, Kashiwazaki R, Lovell MA, Herrmann BW. Pediatric sublingual dermoid and epidermoid cysts: A 20-year institutional review. *Int J Pediatr Otorhinolaryngol.* 2020 Nov;138:110265.
- Park SW, Lee JJ, Chae SA, Yoo BH, Kim GJ, Lee SY. Congenital epidermoid cyst of the oral cavity: prenatal diagnosis by sonography. *Clin Exp Otorhinolaryngol.* 2013 Sep;6(3):191-3.
- Oginni FO, Oladejo T, Braimoh RO, Adenekan AT. Sublingual epidermoid cyst in a neonate. *Ann Maxillofac Surg.* 2014 Jan;4(1):96-8.
- Voss JO, Buehling S, Thieme N, Doll C, Hauptmann K, Heiland M, Adolphs N, Raguse JD. Sublingual cysts of different entities in an infant - A case report and literature review. *Int J Pediatr Otorhinolaryngol.* 2018 Oct;113:260-265.
- Ho MW, Crean SJ. Simultaneous occurrence of sublingual dermoid cyst and oral alimentary tract cyst in an infant: a case report and review of the literature. *Int J Paediatr Dent.* 2003 Nov;13(6):441-6.
- Bitar MA, Kumar S. Plunging congenital epidermoid cyst of the oral cavity. *Eur Arch Otorhinolaryngol.* 2003 Apr; 260(4):223-5.
- Pontes FSC, de Souza LL, Pedrinha VF, Pontes HAR. Congenital Ranula: A Case Report and Literature Review. *J Clin Pediatr Dent.* 2018;42(6):454-457.
- Soni A, Suyal P, Suyal A. Congenital ranula in a newborn: a rare presentation. *Indian J Otolaryngol Head Neck Surg.* 2012 Sep;64(3):295-7.
- Reddy A, Kreicher KL, Patel NA, Schantz S, Shinhar S. Pediatric epidermoid cysts masquerading as ranulas: A case series. *Int J Pediatr Otorhinolaryngol.* 2016 Feb;81:26-8.
- Şimşek-Kaya G, Özbudak İH, Kader D. Coexisting sublingual dermoid cyst and heterotopic gastrointestinal cyst: Case report. *J Clin Exp Dent.* 2018 Feb 1;10(2):e196-e199.
- Eppley BL, Bell MJ, Sclaroff A. Simultaneous occurrence of dermoid and heterotopic intestinal cysts in the floor of the mouth of a newborn. *J Oral Maxillofac Surg.* 1985 Nov;43(11):880-3.
- Drucker C, Gerson CR. Sublingual contiguous thyroglossal and dermoid cysts in a neonate. *Int J Pediatr Otorhinolaryngol.* 1992 Mar;23(2):181-6.
- Wang WC, Lin LM, Shen YH, Lin YJ, Chen YK. Concurrent extravasation mucocele and epidermoid cyst of the lower lip: a case report. *Kaohsiung J Med Sci.* 2005 Oct;21(10):475-9.