Congenital Ranula: A Case Report and Literature Review

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Ranula consists of a pathological process induced by ductal disruption of the minor salivary glands followed by extravasation of mucous material surrounding adjacent structures. A swelling causing breathing and feeding problems associated with tongue displacement is frequently observed. It is a disease that generally involves the younger age group. In newborns congenital ranula may occur, an uncommon variance that differs from common ranula by not relate to post-traumatic reactions. There are reports that indicate a salivary gland duct atresia as the main cause of this pathology. The aim of this study is to analyze the clinicopathological characteristics of congenital ranula by reporting a new case report of this salivary cyst and reviewing the case reports previously published in the English literature.

Key words: *Salivary gland; ranula; congenital.*

INTRODUCTION

Ranula is a mucocele in the floor of the mouth. It is a mucus extravasation pseudocyst arising from the sublingual glands[1]. It is most commonly observed as a bluish cyst located below the tongue[2]. Ranulas in paediatric patients are infrequent, corresponding to 0.2 cases *per* 1000[3].

The incidence of congenital ranula (CR) is estimated to be about 0.7% in newborn infants[1,3-6]. Prenatally diagnosed ranula is an uncommon pathological finding [1]. An extremely small proportion of ranulas can become large enough to cause airway compromise, feeding difficulties and facial defects[3]. They may spontaneously resolve over a period of time or require surgery to solve the problem[7].

The aim of this study was to analyse the clinicopathological characteristics of CR by reporting one new case of this salivary gland lesion and reviewing all the previously published case reports in the English literature.

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CASE REPORT

A 4-month-old male patient was referred to the Oral Pathology and Surgery Centre of the João de Barros Barreto University Hospital, Belém, Pará, Brazil. His parents complained that he had been born with a swelling in the left side of the floor of the mouth which had increased over time. The baby was delivered via Caesarean section and no significant diagnosis was made during the antenatal period. The parents reported no significant event/condition. No trauma history of the affected site was observed.

Clinical examination revealed good general health of the patient. No enlarged lymph nodes were palpable. Intraorally, a smooth and fluctuating swelling on the left side of the mouth's floor, with no buccal or palatal expansion, was revealed. The overlying mucosa presented intact, translucent and of normal color. In addition, the swelling caused tongue elevation but no difficulty in feeding, pain or airway obstruction was reported by the child's parents (Fig. 1A).

Based on the diagnosis of CR, under general anesthesia the patient underwent a puncture of the lesion, in which a mucous material consistent with being a salivary component was found (Fig. 1B-C). At the same section, a surgical excision with tumor-free margin was performed (Fig. 2A). Histopathological examination showed areas with an accumulation of mucoid material with granulation tissue reaction as well as mononuclear inflammatory infiltrate (Fig. 2B). The post-operative course was uneventful. The patient remains under regular follow-up for 51 months with no sign of recurrence (Fig 2C).

Figure 1. Initial clinical aspect, showing left side swelling on the mouth's floor, with posterior displacement of the tongue (A). Lesion's puncture (B). The mucus liquid obtained under puncture (C).

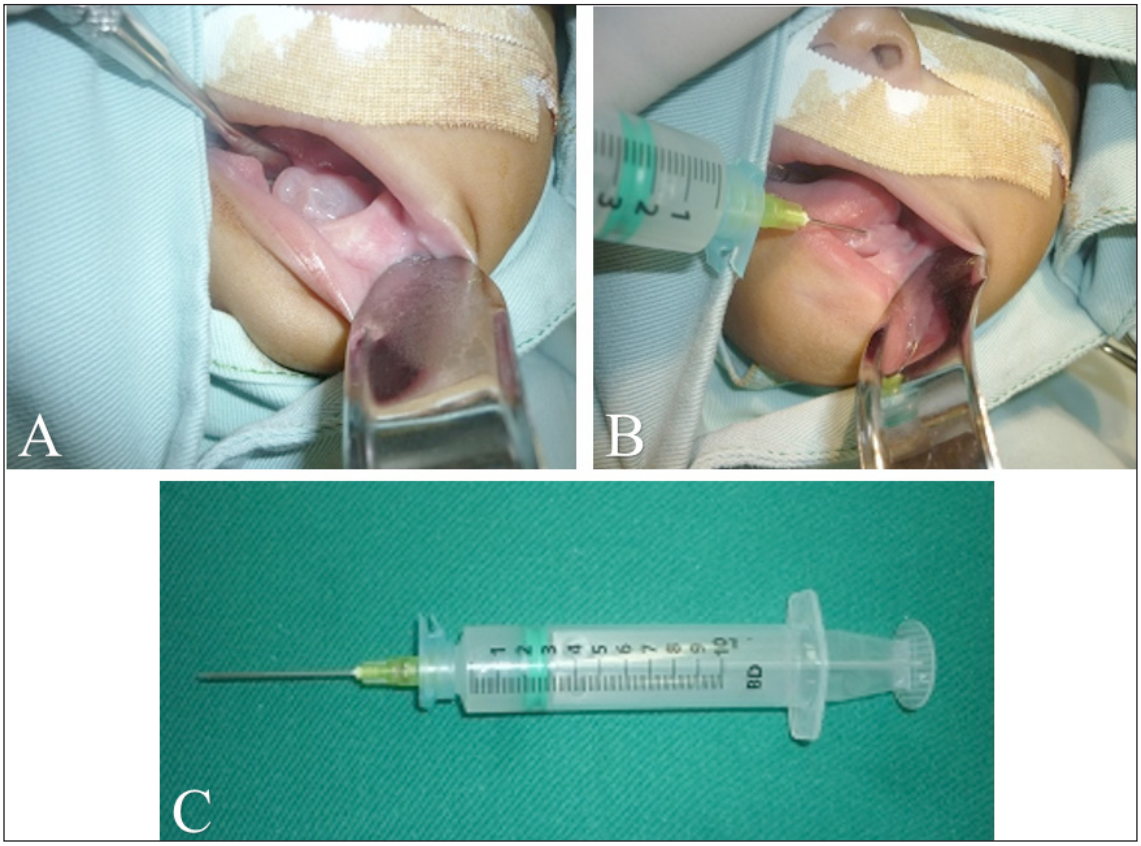
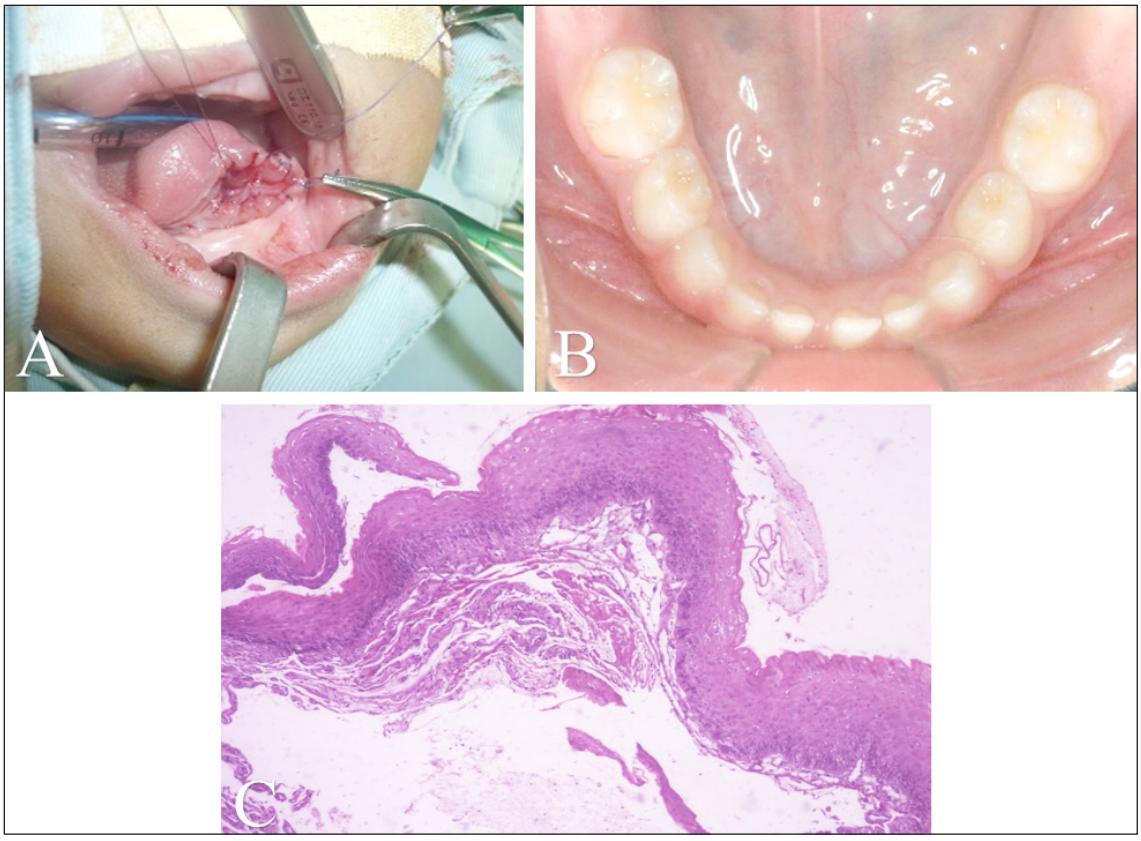


Figure 2. Excisional biopsy of the lesion and the gland (A). Clinical aspect after 51 months of follow-up (B). Microscopic examination revealed an area of extravasation of mucous with granulation tissue-type reaction (C).



DISCUSSION

Ranula is derived from the Latin word 'rana' (meaning 'frog') and is clinically described as a translucent swelling on the floor of the mouth, presenting a frog's belly characteristic[1]. Clinically, there are two subtypes of ranula: the simple and the plunging. The first type presents mucous retention cyst characteristics[4,7]. It is usually located in the floor of the mouth, is mostly asymptomatic, although it can lead to airway obstruction[6,8,9]. The plunging type is rare, and occurs due to an accumulation of mucus in the submandibular and submental spaces, with or without associated intraoral lesions[1,9]. The protein content of the secretion triggers an intense inflammatory reaction and results in pseudocyst formation[1,4,6].

CR pathogenesis is not well established. However, Mun *et al* [10] in a prospective study verified its pathogenesis by an analysis of the anatomical variation of the sublingual gland. In general, numerous ductules from the posterior sublingual gland open onto the summit of the sublingual fold; however, several of the ductules can also join to form a common duct (Bartholin's duct) that empties into Wharton's duct. Trough meticulous dissection, they found that 88.9% of simple ranulas contained Bartholin's duct in comparison to 42.9% of plunging ranulas.

Ranula can be acquired or congenital. Acquired ranula occurs as result of a post-traumatic obstruction of the sublingual gland ducts and is frequently seen in young patients[9,11]. According to Gul *et al* [1], congenital ranula occurs principally following atresia or failure of the canalization of the salivary gland ducts and the incidence has been estimated to be 0.74% in newborn infants[2,6,7,9]. Our case report shows this characteristic, since our patient presented the CR in a postnatal period. However, this variance can be identified during the antenatal period[1,3,4,6,8,9,11] under intrauterine diagnosis, and during the postnatal period, after the baby's birth[2,5,7,12,13,14], and both genders may be affected with no predilection. Swelling with tongue displacement, breathing and feeding difficulty are the most common symptoms found in the literature review[1,5,6,8,14]. In the case presented here, only a small swelling was observed.

It is important that CR is diagnosed during intrauterine life through the use of imaging techniques[1,3,4,5,6,8,9,11]. Ultrasound scan, computed tomography (CT) and magnetic resonance (MR) imaging are able to reveal and monitor the region affected by this disease. Ultrasound scan can detect the lesion in prenatal evaluations and facilitates the evaluation of any increase in the size of the lesion. Under ultrasound scan, the image consists of a well-circumscribed cystic appearance[11-14]. After a baby's birth, CT and MR are the examinations of choice and are currently employed to assess the CR's extension. The cystic fluid's color provides some information on the inflammation level and the lesion's duration[11,12]. A clear liquid is seen when inflammation is minimal and the lesion is relatively new[5]. In the present case, a clear liquid was observed during surgical excision. Histological sections reveal fragments of glandular cyst characterized by the presence of tubules and ducts of minor salivary glands. In focal areas, the presence of amorphous material interpreted as mucin permeated by foamy macrophages is observed. Surrounding the mucin is granulation tissue coated by blood vessels and mononuclear inflammatory infiltrate[3,7,9,14].

The optimal treatment for CR is not well established. Crysdale *et al* [15] showed a recurrence rate of 0% only with the excision of the ranula, 61% with marsupialization and 0% with ranula and ipsilateral sublingual gland excision. In our literature review, for CR, it was observed that surgical excision and marsupialization showed a good prognosis when recurrence was evaluated. In the case reported here, the surgical excision of the lesion associated with the salivary gland was performed and a good prognosis was observed. In addition, Zhao *et al* [16] when evaluated the recurrence index after treatment observed that excision of the sublingual gland is the best choice of treatment, since the recurrences correspond just 1,2% of all cases.

The differential diagnosis of a neonate swelling in the mouth's floor includes teratoma, dermoid cyst, lymphatic malformation, congenital ranula, heterotopic gastric cyst and thyroglossal duct cyst[17,18]. Nevertheless, several complementary examination may help to perform the diagnosis, such as sialography, fine needle aspiration cytology, MR, CT and ultrasonography[17]. However, the best choice to differentiate the lesions are histopathological exam and fine needle aspiration cytology. In the present case report, the clinical features, medical history, and histopathological analysis confirmed the diagnoses of the CR.

CONCLUSION

Congenital ranula is seen as an uncommon disease, which may be discovered in the prenatal period, generally after 21 weeks of gestation. There is a need for the careful evaluation of a congenital ranula, based on clinical and radiological examinations, to discern etiology and provide appropriate treatments[19,20]. Atresia of the salivary gland ducts and the location of the CR, considering the sublingual gland evolvement, is the main cause of this pathology in the congenital type and the best therapeutic treatment is surgical excision.

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