



Congenital Granular Cell Tumor: A Case Report with a 9-years of Follow-up

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Abstract

Congenital granular cell tumour or congenital epulis is an uncommon benign tumor of the oral cavity of newborns. The condition may interfere with respiration, feeding or lip closure. Antenatal ultrasound diagnosis is possible and it can avoid unexpected situations. Surgical excision is the first line treatment but, occasionally, the lesion may present spontaneously regress postpartum. The aim of this case report is to describe the clinical presentation and treatment of the congenital granular cell tumour in the maxillary arch of a 2-hour-old female newborn.

Keywords: Congenital epulis; Granular cell tumor; Newborn

Introduction

Congenital Granular Cell Tumour (CGCT), also known as congenital epulis or congenital gingival granular cell tumor, is an uncommon benign tumour that is usually diagnosed clinically at birth or by antenatal ultrasound [1,2], was first reported by Neumann in 1871, for this reason can be called as Neumann tumor as well [3]. The condition is seen 3 times more frequently in the maxillary alveolus than in the mandibular alveolus [4,5]. Females are affected from 9 to 1 time more frequently than males [6]. The typical location is the alveolar ridge of the maxilla near the lateral incisor and canine [7,8], with variation in size from a few millimeters to as large as 9 cm. Usually, it is presented as a single mass in a round or ovoid shape, pedunculated or sessile, and it may interfere with feeding and respiration; however, some cases of multiple lesions have been reported [9]. The recommended treatment plan consists of surgical resection of the mass under General Anesthesia (GA) or Local Anesthesia (LA), considering that spontaneously regression is rare. There are no reports showing CGCT recurrence or malignant transformation. Nowadays, surgery procedure with a high power laser is an option for resection, besides with this technique suturing is not necessary, because the laser itself cauterizes the peripheral vessels [10,11]. The aim of this case report is to describe the clinical presentation and treatment of the congenital granular cell tumour in the maxillary arch of a 2-hour-old female newborn.

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Case Presentation

A healthy female newborn was referred to the Oral & Maxillofacial Surgery team for diagnosis and treatment of a large mass protruding from the mouth. The mother did not perform appropriate antenatal ultrasound and no remarkable medical history was noted. The infant was born by normal vaginal delivery. At birth, a firm pedunculated mass was observed by the medical team protruding from the oral cavity (Figure 1). Two hours after birth, a clinical examination performed by the Oral & Maxillofacial surgical team revealed a single nodular mass, reddish and firm on palpation, measuring about 3 cm in diameter. The mass was pedunculated and it was arising from the maxillary over the lateral incisor and canine area (Figure 2). The patient presented obstruction of airway and difficulty in alimentation (the mass interfered while breastfeeding). An urgency surgery was scheduled on the same day. The surgical resection of the mass was done under GA with oral endotracheal intubation (Figure 3). A scalpel was used for excision, and there was a minimal blood loss (Figure 4). In post excision, any kind of alveolar defect was noted (Figure 5). Postoperative recovery was uneventful. The female newborn was breastfed immediately after surgery with any complications (Figure 6). Healing was uneventful and the 9 years follow-ups demonstrate a normal development



Figure 1: Overview for the congenital epulis.

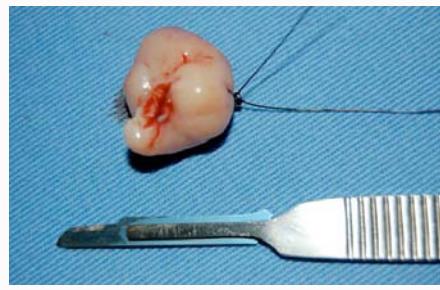


Figure 4: Resection mass of congenital granular cell tumor.

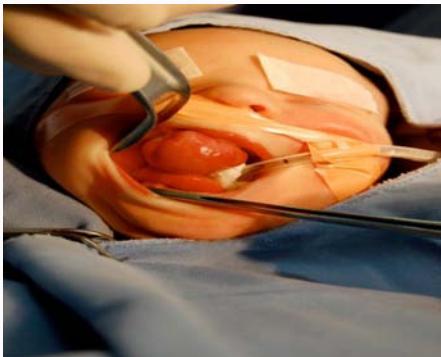


Figure 2: Intra oral view of the congenital epulis.



Figure 5: Alveolar ridge post excision.



Figure 3: Pre-op view of 2-hour newborn.



Figure 6: Breastfed post-op.



Figure 7: Follow-up of nine years without recurrence.

of the buccal tissues without recurrence (Figure 7). Histopathological analysis revealed a fragment of mucosa coated by atrophic squamous stratified epithelium showing sheets of large polygonal cells with abundant granular cytoplasm and small lightly basophilic eccentric nuclei, consistent with the diagnosis of congenital granular cell epulis or congenital epulis (Figure 8).

Discussion

Congenital epulis is also known as granular cell tumour of the gingiva is encountered exclusively in newborns. The etiology of the lesion was still unclear and controversial [1]. A couple of theories suggests that the tumor may has myoblastic, odontogenic, neurogenic, fibroblastic, histiocytic and endocrinologic pathogenesis [4,8]. This last one could be responsible for stimulation of the intrauterine endogenous hormone that could explain a marked female preponderance of 8 to 1 male [12,13]. Histopathological findings of CGCT include large round cells with granular, eosinophilic cytoplasm and small eccentric nuclei and a delicate fibrovascular network. These characteristics are similar to those found in the adult granular

cell tumor with differentiation in a pseudo-epithelialomatous hyperplasia, lesser vascularity and more conspicuous nerve bundles than congenital epulis [2,14]. In addition, CGCT are present at birth while the adults or infants granular cell tumor appear at any stage of live [15]. The diagnostic of CGCT is based on clinical characteristics of the tumor added the histopathological examination. Even so, some authors suggest that immunohistochemistry could be used as a complementary method as well. The markers vimentin and S-100, whose positive marking mesenchymal and neural cells origin respectively, were successfully used in 20 cases. In greek epulis means “of the gums”. It’s interesting to note that in a large

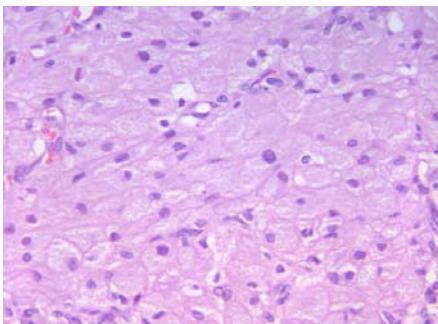


Figure 8: Cells with abundant granular eosinophilic cytoplasm. HE 400x.

of cases the tumor usually arises at the maxillary alveolar ridge without dental involvement [5,7,8], with an average size of 1.5 cm. Although unusual, some authors also have described a few cases of CGCT on the tongue as well. The clinical presentation consists of a single mass, sessile or pedunculated with a smooth mucosal surface, reddish in color [16,17]. As reported by Saki [6], Hiradfar [18], Damante [19] and Dzieniecka [20] some cases of multiple lesions are described at the same patient. The differential diagnosis should be included congenital malformations, such as encephalocele, as well as benign and malignant neoplasms, such as lymphatic malformation and rhabdomyosarcoma or non-specific soft tissue masses such as a fibroma or a vascular malformation or other developmental anomalies such as CGCT or neuroectodermal tumors of infancy. Clinical aspects like site of location, size and growth potential may discard such lesions. Diagnosis is mainly clinical postnatal. However, the tumor can be visualized by antenatal ultrasound or with better accuracy in a Magnetic Resonance Imaging (MRI) from the 26th week of gestation. Despite the cost of MRI technique, one of the major disadvantages must consider that the image findings are nonspecific. Even so, once visualized, the imaging exams can anticipate surgical postnatal surgery planning and delivery as well. In medical history no remarkable problem was related to the pregnancy but the mother was from a remote rural area, so she couldn't maintain a prenatal routine. Even with a successful intervention we hypothesized if an antenatal ultrasound could be anticipate the surgical planning for surgeon's team. CGCT is not associated with congenital abnormalities. Although rare, spontaneous regression has been reported in the literature. The surgical excision represents the first option treatment under LA or GA. In this case report, the mass resection was performed to restore airway and breastfeeding. The procedure restored the anatomy of the premaxilla without damage of a bone tissue, allowing healing of the buccal mucosa, and the future normal tooth eruption. To note, this is the first case report that presented a surgical resection of a CE with a follow-up of 9 years. Even in cases of incomplete resection the prognosis of CGCT is good and no reports about recurrence were made until now. Now days, scalpel techniques have leading to a little more painful and uncomfortable post-surgical period, mainly because of the presence of sutures [21]. Alternatives techniques such as the use of laser – assisted oral surgery may be advantageous in allowing reduction of surgical time, the absence of bleeding and consequently promote a good vision of the surgical site, often avoiding infiltrative anesthesia and suture with a better and faster healing process [22,23].

Conclusion

The tumor is often misdiagnosed before surgery because of its rarity and lack of awareness among clinicians. It can be alarming for

parents and for the obstetrics team. In this case report, it's important to emphasize the performance of the multiprofessional team, with an urgent action, that favors a correct diagnosis and successful treatment with a favorable prognosis of nine-year follow-up.

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