

Congenital granular cell epulis: report of a case and review of the literature.

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ABSTRACT

Congenital granular cell tumour of the new born is a rare benign lesion of soft tissues which normally affects the maxillary alveolar ridge in neonates. Though benign, it can cause respiratory embarrassment and prevent feeding causing physical distress to the infant and psychological distress to the parents or guardian. Very few cases are reported in the literature. We aimed to highlight the clinical presentation, management and a brief review of the literature of the lesion.

A case of a 3-day old baby who presented to the Oral and Maxillofacial Surgery Clinic University of Benin Teaching Hospital (UBTH) was used to illustrate the clinical presentation and management of congenital epulis.

Congenital epulis of the new-born is a rare occurrence, though it can grow to a frightening size, it still remains a benign lesion with little or no recurrence following surgical excision. The role of counselling to the parents cannot be over-emphasized.

Keywords: Congenital, epulis, granular, newborn.

INTRODUCTION: Congenital granular cell epulis (CGCE) is a benign soft tissues lesion of gingival origin occurring in the neonates^{1,2}. CGCE is very rare and unique^{2,3} and was first described in 1871 by a German pathologist, Ernst Christian Neumann⁴. It is an intriguing lesion with unclear aetiology, histogenesis and natural history. Several theories have been suggested, namely myoblastic, odontogenic, neurogenic, fibroblastic, histiocytic, and endocrinologic origin^{3,5}. Out of the several theories proposed, the most favoured are the odontogenic and gingival epithelial theories, which support its origin from mesenchyme⁵. There are usually no associated dental abnormalities or congenital malformations except for occasional reports of a hypoplastic or absent tooth and possibility of mild mid face hypoplasia⁶. Also, polyhydramnios has been noted and reported due to the blockage of the oral cavity and inability to

swallow⁷. Due to its rare occurrence, we therefore aim to report a case and review the available literature. A new born female child was referred to the Department of Oral Maxillofacial Surgery, University of Benin Teaching Hospital, Benin City, Edo State from a private facility three days after delivery on account of a protruding mass from the mouth. On clinical examination, a round, soft, pedunculated mass measuring 10cm by 6cm was found to be located on the anterior maxillary alveolar ridge. Swelling was lobulated with a bigger right lobe (Figure 1). The mass prevented normal mouth closure, interfered with breast feeding but did not pose any obvious threat to the airway. There was no history of spontaneous bleeding. Parents were counselled and patient was booked for excisional biopsy and blood samples were collected for basic haematological and biochemical investigations. Results obtained were within the normal range. Following discussion with paediatrician as regard fitness for surgery, surgical excision of the mass was done by the use of cautery under local anaesthesia (Figure 2). On the seventh day postoperative followup, the wound healed without complications (Figure 3). The examination of the resected specimen showed a bilobular, encapsulated, smooth mass (figure 4) and sections of the specimen appeared pale and gritty-like. On histological examination, the tissues were seen to be composed of sheets of large polygonal granular cells with distinct

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Received:05/03/2019

Accepted:22/08/2019

<https://dx.doi.org/10.4314/ajoh.v9i1.6>

borders, having abundant granular eosinophilic cytoplasm and predominantly eccentrically located vesicular nuclei with conspicuous nucleoli (Figure 5). There were no signs of recurrence after 5 months of follow-up.

DISCUSSION

Our review of the literature regarding this condition was complicated by somewhat confusing terminology used up to date. It could be referred to as congenital epulis of the newborn (CENB), congenital granular cell tumor (CGCT), congenital granular epulis (CGE), congenital granular cell epulis (CGCE) congenital granular cell myoblastoma (CGCM), congenital granular fibroblastoma (CGF), and congenital Neumann's tumor (CNT). The Greek term epulis literally means swelling on the gingiva⁸ and is used in dentistry to nonspecifically refer to hyperplastic gingiva tumour mass⁹. Thus a more specific terminology of "CONGENITAL GRANULAR CELL EPULIS" is recommended by the World Health Organisation^{10,11}.

Epidemiology: Due to the infrequency of CGCE occurrence, it has mostly been noted in the literature via case reports and literature reviews. Since its first description in 1871⁴, more than 170 cases have been reported since then. Two of the larger works include that of Dash et al¹² including fifty patient reviews and Lack et al.¹³, including 21 patient reviews noting multiple commonalities. The incidence rate was found to be 0.0006% at a centre in Wales and epulis accounted for 10.8% of the entire oral lesion in a centre in India. There is a noted higher incidence in the Caucasian population.

Aetiology: The aetiology is unknown, several theories have been suggested, namely myoblastic, odontogenic, neurogenic, fibroblastic, histiocytic, and endocrinologic^{3,5}. Out of the several theories proposed, the most favoured are the odontogenic and gingival epithelial theories, which support its origin from mesenchyme⁵. There are usually no associated dental abnormalities or congenital malformations except for occasional reports of a hypoplastic or absent tooth and possibility of mild mid face hypoplasia⁶. Also, polyhydramnios has been noted and reported due to the blockage of the oral cavity and inability to swallow⁷.

Clinical presentation: Clinically, the lesion

usually presents as a smooth surface sessile or pedunculated mass with a normal to reddish colour. Its size varies from several millimetres to a few centimetres in diameter and may interfere with feeding and respiration^{6,8,14,15}. Mostly solitary but, large and multiple lesions have been reported⁵. The lesion has a site predilection for the maxillary alveolar process, lateral to the midline in the region of the primary canine and lateral incisor.^{2,5,6,8,14,15} It has an 8-10:1 sex predilection for females^{1,3,5,8,15}. Less frequently, it has been reported in the mandibular alveolus, tongue, palate, skin, the subcutaneous tissue, skeletal muscles, and vocal cords⁸. The case reported was a female with lesion on the canine region. The case presented in this paper has a typical appearance of what has been reported in several literatures; bilobed, smooth-surfaced, pedunculated mass located at the anterior maxillary alveolar ridge in a 3 day old female (Figure 1). The lesion is usually diagnosed at birth clinically, although difficulty may occur when the index of suspicion is low or when the origin of the tumour is hard to determine¹⁵. Also, if the lesion is large, it may be diagnosed in utero by 3D ultrasound and magnetic resonance imaging examinations^{6,16}. In-utero diagnosis is important in choosing the delivery method, since large lesions may compromise a normal virginal delivery and a caesarean operation may be necessary¹⁷. Treatment is usually surgical excision done under general or local anaesthesia depending on the size of the lesion. Few cases of spontaneous regression have been reported following a watchful waiting especially for very small lesions although very rare^{6,18}. After surgical removal, no recurrence or malignant changes have been reported even after incomplete excision¹⁹. No recurrence has also been reported due to incomplete surgical excision⁷. Excision with carbon dioxide laser has also been reported²⁰. Also, in the present case, the lesion affected breast feeding as it was the presenting complaints aside the unsightly protruding mass from the mouth. There was no threat to the airway in the index patient. Reported cases have been excised under local anaesthesia as well as under general anaesthesia^{5,7,19}. In our case the lesion was excised under local anaesthesia due to



Figure 1. Growth attached to the alveolar ridge of the maxilla.



Figure 3. Growth site, seventh postoperative day



Figure 2. Immediate postoperative clinical image demonstrating the site of wound



Figure 4. A Bi-lobed and multi-nodular pre-sectioned surgical specimen

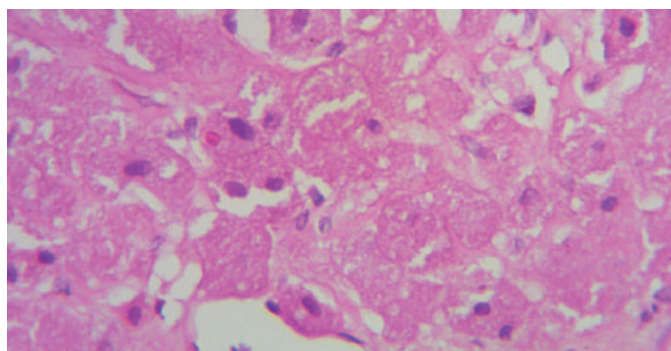


Figure 5. High power view of a congenital granular cell epulis showing large cells with abundant eosinophilic granular cytoplasm. (H&E, $\times 400$)

Differential diagnosis

It differs from other granular cell tumour encountered in adults by its exclusive origin from the neonatal gingival, the scattered presence of odontogenic epithelium, the more elaborate vasculature and the lack of interstitial cells with angulate bodies¹⁵. Clinical differential diagnoses for congenital lesions of oral mucosa depend on site of involvement, size, velocity of growth, and possible accompanying lesions. This includes teratoma, haemangioma, fibroma, Choristoma, hamartoma, melanotic neuroectodermal tumour of infancy, rhabdomyosarcoma, rhabdomyoma, lymphangioma, osteogenic and chondrogenic sarcoma, and granular cell tumour^{6,16,21}.

Immunohistochemistry

Congenital epulis is a rare lesion of uncertain histogenesis²². The immunohistochemistry of the tumour is diverse in newborns. Congenital granular cell tumour/epulis is S-100 negative and does not show differentiation to specific cell type²³. Possible histological origins of the epulis may include epithelial and undifferentiated mesenchymal cells, pericytes, fibroblasts, smooth muscles, nerve related cells and myofibroblasts⁸. It has been suggested to be a non-neoplastic, degenerative, or reactive lesion⁸.

Histopathology

Characteristic histological findings shown by this lesion includes large round cells with granular eosinophilic cytoplasm and small eccentric nuclei and a delicate fibro-vascular network separating the cells. Histopathologically, it is known that congenital epulis consists of granular cells and is similar to the adult granular cell tumour, but there are some differences such as pseudo-epitheliomatous hyperplasia, lesser vascularity, more conspicuous nerve bundle than congenital epulis²⁴. Coarse cytoplasmic granularity and absence of lipid droplets help to distinguish it from rhabdomyoma and leiomyoma⁵. Fibroblastic origin of the lesion has been described based on the description of patients affected by neurofibromatosis⁵.

CONCLUSION

The congenital epulis is an oral mass that presents at birth in neonate. Presence of such oral mass at birth is usually very distressing to the parents, thus parents should be counselled on the benign nature and surgical excision with no documented report of recurrence or malignant transformation. Prompt surgical excision is also advocated especially for lesions that are large and that have adverse effect on

feeding and possible threat of airway obstruction. Good knowledge of differential diagnoses of similar growths in the oral cavities of newborns (teratoma, haemangioma, rhabdomyoma, schwannoma, leiomyoma, and fibroma) is important by the clinicians as treatment modalities will be different in each case.

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