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Case report

Congenital granular cell epulis—a case report

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ABSTRACT

Congenital granular cell epulis (CGCE) is an uncommon benign lesion found in newborns. It has predominance for females with an 8:1 ratio in relation to males and is exclusively encountered in the oral cavity. The most affected oral site is located around the canine/incisor region of the maxillary alveolar ridge, where the lesion arises from the soft tissue as a solitary pedunculated mass. CGCE's histogenesis remains obscure and controversial. We present a rare case of 2 separate CGCE lesions adjacent to each other measuring 23 × 18 × 10 and 15 × 10 mm, positioned facially on the right maxillary alveolar process. The patient, a 2-day-old female newborn, did not experience any serious difficulty regarding breathing or deglutition. Complete surgical excision was the treatment of choice in this case, and the procedure was performed under both general and local anesthesia. Histologic and immunohistochemical analysis confirmed the diagnosis of CGCE. The patient showed satisfactory postoperative healing and excellent health at both the 10-day recall appointment and the 6-month follow-up.

1. Introduction

With <250 reported cases to date, the congenital granular cell epulis (CGCE) verily is a rare lesion. The credit of first describing this lesion in literature goes to the German pathologist Dr. Franz Ernst Christian Neumann who in 1871 presented it as a "congenital epulis." Epulis is a word derived from the ancient Greek language and translates into "swelling on the gingiva" [1]. In medical literature, this lesion is known by many names such as Neumann tumor, Abrikosov tumor, granular cell myoblastoma, and so on; however, the recommended terminology by the World Health Organization is "congenital granular cell epulis" [2].

The CGCE is a benign tumor located in the alveolar ridge of the newborn child. Its histologic origin is still unknown and debated [3]. Clinically, this lesion presents itself in the form of a broad-based, firmly attached solitary-like polypoid nodule with a predominant labial aspect of the gingiva [4]. Because of the size of the tumor and risk of interference with the newborn child's feeding and respiration, the treatment of choice is often acute surgical excision [5].

2. Presentation of case

A 2-day-old female newborn weighing around 3600 g was referred to the maxillofacial department for evaluation and possible treatment of a "large soft mass" positioned facially on the right maxillary alveolar process. Clinical examination of the tumor presented a pink-colored, smooth mass measuring around 20 × 20 mm with no prominent blood vessels. Obtained clinical information was determined as sufficient and resulted in the preliminary clinical diagnosis: congenital granular cell tumor.

The young female was healthy with no additional medical problems. Despite the large size of the lesion, there was no interference with breathing or feeding, and the child stayed within optimal figures regarding neonatal weight and growth. Complete surgical excision was the treatment of choice, and the procedure was performed under both general anesthesia and local anesthesia. During the surgical procedure, a second smaller tumor of the same kind was detected and excised. Thereafter, both tumors were subjected for histopathologic analysis (Figures 1 and 2).

The patient showed satisfactory postoperative healing and excellent health at the 10-day recall appointment. Follow-up at 6 months revealed a small scar formation on the alveolar crest, but seemingly normal development of the tissues at the former site of the tumor (Figures 3 and 4).

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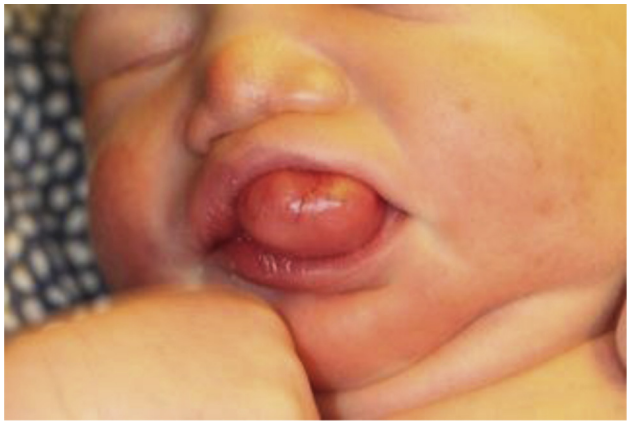

Figure 1. Congenital granular cell epulis in a 2-day-old neonate.

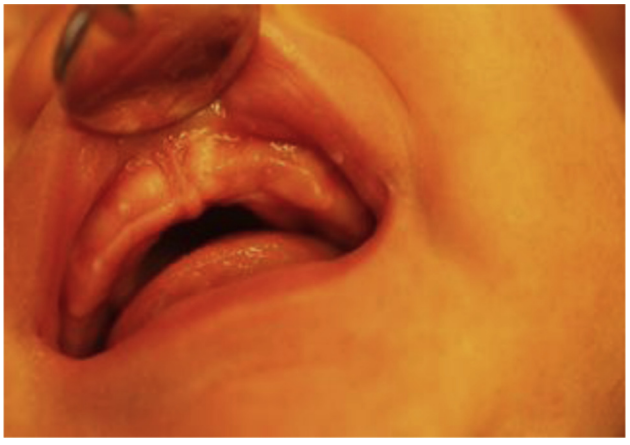

Figure 3. Six-week postoperative healing.

The macroscopic evaluation depicted a large nodular soft mass with a smooth whitish surface, homogenous in terms of texture and measuring 23 × 18 × 10 mm. The microscopic examination illustrated an unencapsulated tumor covered with thin surface squamous epithelium. The tumor consisted of nests and trabeculae of homogenous, large, polygonal cells with eosinophilic granular cytoplasm, which was slightly positive for Alcain Blue-periodic acid-Schiff Stain and had indistinct cell borders. The nuclei were medium to large in size, centrally located, basophilic, hyperchromatic, and vesicular with distinct nucleoli. The resection margins were free. The mitosis rate was variable with an average value of <2 per 10 high-powered field. Immunohistochemically, the lesion showed a strong positivity for staining with vimentin and laminin, but weak positivity to cluster of differentiation 68 (CD68), neuron specific enolase, and microphthalmia transcription factor and negativity to S100, actin, human melanoma black 45, melan A, and Sry-related HMG-BOX gene-10. These histopathologic and immunohistochemical findings confirmed the preliminary clinical diagnosis of congenital granular cell tumor.

3. Discussion

This benign lesion, CGCE, predominantly affects female infants with an 8:1 ratio in relation to male infants and is encountered 3 times more often on the maxilla than on the mandible [6]. Clinically, a majority of cases appear around the canine/incisor region of the maxillary alveolar ridge as a solitary pedunculated mass [7]; however, multiple tumor presentation is seen in 10% of cases [8]. In accordance with this tumor description, our case proves to be very rare with a presentation of 2 different lesions adjacent to each other.

The histogenesis of CGCE is still unknown; however, it is postulated that mesenchymal cell, fibroblasts, and Schwann cells may be candidates of origin [6]. The spatial dimensions of this lesion vary, but a tumor size of up to 90 mm has been reported [8]. CGCE growth patterns show a significant tumor enlargement during the third trimester of the pregnancy and a sudden stop of growth after birth. Owing to this fact and the correlation to female newborn predominance, it is believed that maternal hormones stimulate tumor growth. Studies on mice seem to support this claim; however, paradoxically, no estrogen or progesterone receptors have been detected in CGCE cells [9]. In our case, the primary tumor had an average size and measured approximately 23 × 18 × 10 mm.

Macroscopically, CGCE illustrates a defined intraoral firm mass with a lean reddish surface. The tumor is limited to the soft tissue and does not involve hard tissue such as bone and teeth [4]. Microscopically, it is a circumscribed entity made up of medium to large cells in a nestlike densely packed order with a homogenous and polygonal presentation (Figures 5 and 6). The cells have an eosinophilic granular cytoplasm, eccentric nuclei, small nucleoli, and a low-grade to absent mitosis activity [4]. Pseudoepitheliomatous hyperplasia can often be seen by the overlying squamous epithelium [4,10,11]. In our case, however, there was an absence of pseudoepitheliomatous hyperplasia. Malignant characteristics of

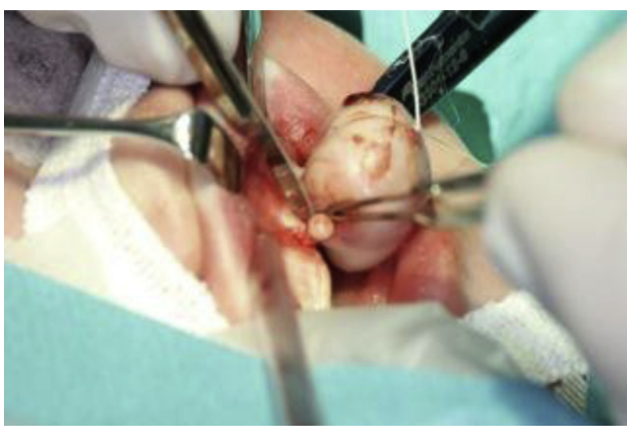

Figure 2. Surgical excision of the multiple tumors.

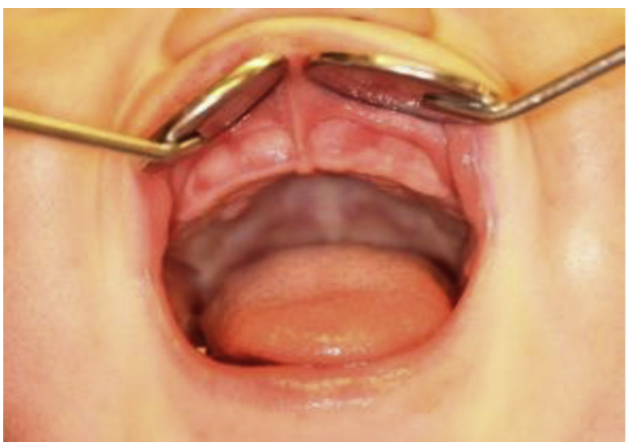

Figure 4. Six-month healing.