Congenital granular cell tumour: report of a case

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Abstract

Congenital granular cell tumour of the newborn is an uncommon benign tumour of uncertain origin. Mostly, it occurs as a single tumour but rarely as multiple. The lesion arises from the mucosa of the gingiva either from maxillary or mandibular alveolar ridge. This paper reports a three-day old female patient who presented at the dental centre of General Hospital, Lagos with a pedunculated soft tissue mass arising from the incisal region of the maxillary alveolar ridge. The lesion was excised under local anasthaesia when the baby was four days old. Microscopic examination of the lesion showed stratified squamous epithelium overlying sheets of large polygonal cells with granular cytoplasm and a diagnosis was congenital granular cell tumour.
In view of the fact that cases reported in the scientific literature are not too many, there is justification for report of this case and reports of more cases are recommended

Key words: Congenital granular cell tumour, Maxilla.

Introduction

Congenital granular cell tumour, is a rare benign soft tissue lesion of unknown etiology. It commonly occur along the alveolar ridge of the maxilla in the neonate [1-10]. The lesion was described for the first time in 1871 by Neumann [2]. To date, fewer than 200 cases have been described in the literature [3, 6]. The lesion which usually present at birth is multiple in 10% of the cases. The lesion usually appears as a protuberant mass, sometimes pedunculated or sessile, firm in consistency with a smooth or lobulated surface. The size of the tumor varies from several millimeters to centimeters in diameter [5]. Females are affected ten times more than the males [4-7]. The tumor is seen twice as often in the maxilla than in the mandible and usually in the incisor-canine region. Surgical excision is advocated as the treatment of choice for CGCT, preferably immediately after detection. Spontaneous regression is rare and large tumour may cause airway obstruction and feeding difficulties [3-10]. The aim of this paper is to report a case of this rare tumour in a 3 day old-girl, managed in our institution.

Case report

A 3 day old healthy female child was presented at the oral and maxillofacial surgical out patient clinic of Dental Centre, General Hospital Lagos with an unusual swelling over the central incisors of the maxilla present since birth. (Figure 1) The mother claimed that the swelling had been interfering with normal breast feeding of the new born baby. Delivery had otherwise been normal at full term and maternal and paternal histories were non contribution.

Figure 1. Clinical presentation of the lesion in the maxillary anterior region

Medical history was not relevant. The lesion was firm, non tender, measured 3x1.5x3 cm and was the same colour as of oral mucosa and pedunculated in nature (Figure 2).
The lesion was surgically excised under local anesthesia using 2% xylocaine, in the dental chair. The blood loss was minimal; the intraoral wound was sutured with black slick suture which was removed after one week post operatively. The patient was placed on antibiotics, mild analgesic post operatively for 5 days and mother counseled to continue breast feeding. The immediate postoperative period was uneventful. The histopathology of the specimen revealed a mass of tissue consisting of parakeratinised stratified squamous epithelium overlying sheets of large polygonal cells with pale granular cytoplasm and small compact nuclei (Figure 3). A diagnosis

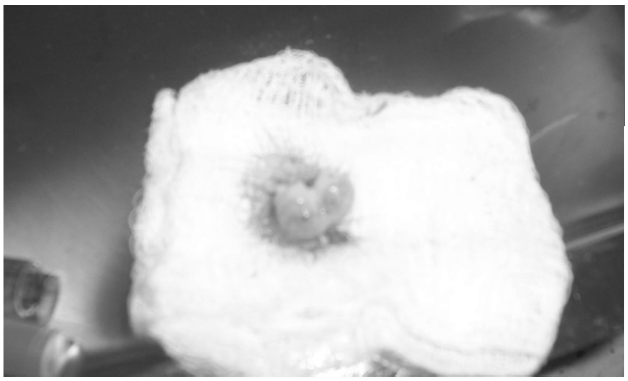

Figure 2. Excised specimen

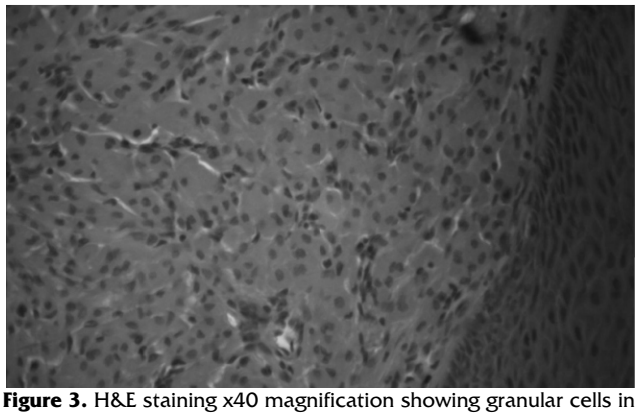

Figure 3. H&E staining x40 magnification showing granular cells in the subepithelial stroma

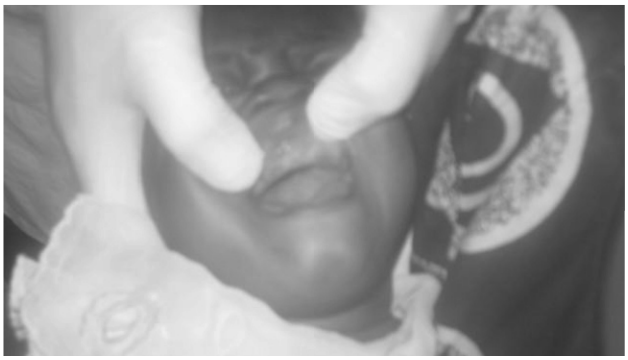

Figure 4. Post operative picture of patient after 9 month

of Congenital Granular cell tumour was made.

The patient presented at 9 months (Figure 4) and 16 months for post operative follow up with no evidence of recurrence. The left central incisor had erupted by the 16th month when she presented for follow up.

Discussion

Congenital granular cell tumour of the gingiva (CGCT), congenital granular cell myoblastoma, or Newman's tumour, following the first published case is encountered exclusively in newborns [2]. CGCT is usually referred as congenital epulis in the medical literature. Epulis however is a non -specific term used to designate hyperplastic gingival tissue or gingival tumor masses. Since there are cases which are not exclusively related to the gingiva it seems the term CGCT would be more appropriate term for this disease entity [6,7,11].

Less than 200 cases of CGCT have been described in the literature [3,6,9]. This report describes a rare case of congenital granular cell lesion with involvement of maxillary alveolar ridge in a three day old female child (Figure 1). Females are affected ten times more than the males [1,3-9]. An endogenous hormonal influence has been proposed to explain the female prevalence and the intrauterine growth, but this theory is not proved since detectable oestrogen and progesterone receptors within the lesions are absent [4]. This present case, like most cases that have been reported in the literature appeared as a solitary lesion, occurring on the maxillary alveolar ridge. The maxillary to mandibular ratio is 3:1. However, CGCT is reported to occur in both maxillary and mandibular alveolar ridges in about 10% of the cases in literature [3-8] . The canine/incisor region, which is reported as the most common site of the disease in literature was also the site of presentation in this case. There are also reports of occurrence of CGCT in multiple sites in the oral cavity involving alveolar and extralveolar sites, for example, the tongue [1, 9]. The frequent occurrence in the maxillary canine and incisor region has been attributed to the fact that the maxillary anterior region is a common site for supernumerary teeth. Additional congenital or underlying bone or dental anomalies are usually not presented in this disease, although there are reports of a hypoplastic or absent underlying tooth [9].

The etiology of the condition is unknown. Several theories have however been suggested, namely; myoblastic, odontogenic, endocrinologic neurogenic, fibroblastic and histiocytic [6, 10]. These latest findings however suggests the presence of cells with different potentiality and capacity for multiple pathway of differentiation, CGCT presents a wide range of differential diagnoses, such as granular cell tumor, fibroma and melanotic neuroectodermal tumor of infancy, among others. However, the clinical diagnosis is usually simple due to the typical occurrence in the maxillary alveolar mucosa of newborn girls. The most important lesions to different it from however, is the oral melanotic neuroectodermal tumor of infancy. Other possible differential diagnoses that should be considered include, fibroma, lipoma, leiomyoma, rhabdomyoma, rhabdomyosarcoma, peripheral giant cell granuloma, pyogenic granuloma, cysts of oral mucosa, fordyce's spot, natal teeth, eruption cysts, neurofibroma, myxoma, hemangioma, lymphangioma, and congenital ranula [7-10].

Prenatal imaging of congenital lesion of oral mucosa is possible by ultrasound and magnetic resonance imaging and can be modified in planning the delivery [9, 10]. Post natally computed tomogragh or magnetic resonance imaging of the head is useful in demonstrating the extent and differential diagnoses of congenital maxillofacial lesional mass and for planning surgical treatment [7, 9, 10]. Surgical excision is the treatment of choice as was done in this case especially when it is interfering with feeding. There are sporadic reports of spontaneous regression of CGCT. Most studies however favour a conservative approach to surgery as it minimizes danger of damaging underlying alveolar bone and developing tooth buds [9,10].

Conclusion

The present case report describes a case of congenital granular cell tumour in the incisal region of the maxillary alveolar ridge. There was indication for excision of the lesion because it interfered with the normal breastfeeding of the patient and caused psychological trauma to the parents. In view of that fact that the lesion is rare, it is recommended that more cases be reported.

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