

Congenital Epulis: Case Report and Literature Review

REMA NAGPAL, PRADEEP SURYAWANSHI, NANDINI MALSHE, ALOK GODSE, VIJAY KALRAO

ABSTRACT

Congenital epulis of the neonate (CE) or congenital gingival granular cell tumor (CGCT) is a rare, benign, mostly single tumor. This arises from gingival mucosa, most commonly from maxillary alveolar ridge, with

predominance in females. The histogenesis of the lesion remains unclear. The present report describes a case of congenital epulis in the maxillary alveolar ridge. This lesion was causing feeding difficulties and was excised under general anaesthesia.

Key Words: Congenital epulis (CE), Congenital Granular Cell Tumor (CGCT), Neumann's Tumor

INTRODUCTION

Congenital Epulis is a rare, benign, soft tissue gingival lesion [1,2,3]. Neumann described the first case of CE in 1871, hence it is also known as Neumann's tumor [1,4,5]. The word 'Epulis' is derived from a Greek word, meaning 'on the gum' or gum boil. CE commonly arises from the future site of the maxillary canine or lateral incisors [6] of newborn, and is usually not associated with any other congenital malformations [4,5]. CE has a female predilection with 8-10:1 ratio [1,4,5,7]. Zuker and Buenecha in a review (1993) described only 167 reported cases in the literature [2]. To date, there are about 200 reported cases in literature [5].

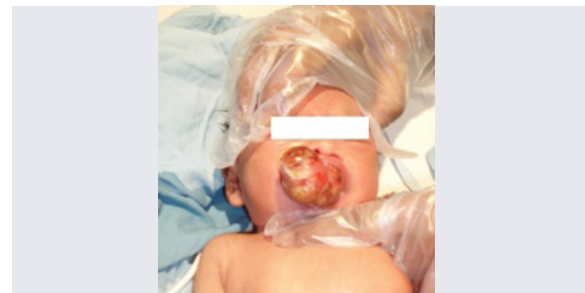
This report documents the presentation and management of a congenital granular cell tumour of the left maxillary alveolar ridge, found in a female newborn, and treated with prompt surgical excision. We did a review of Indian literature and were able to document 23 reported cases of CE in India, indicating its predominant Caucasian distribution and relative rarity in the Indian Subcontinent.

CASE REPORT

A term female neonate presented at 4 hours of life to our center for evaluation of a firm pedunculated mass protruding from mouth [Table/Fig-1]. The neonate was born to a G₂ P₁ L₁ unregistered mother by normal vaginal delivery with an unremarkable antenatal history. The birth weight was 2700 grams. Maternal & paternal medical histories were non-contributory. The mass was attached to the gingiva of the left maxilla, measuring 4.0 x 2.0 x 4.0 cm, was of the same colour as the oral mucosa, smooth-surfaced and pedunculated. On palpation, it was firm, not compressible or reducible, and non-tender. The mass hindered breast-feeding, but did

not cause airway obstruction or respiratory distress. No other structural deformities were noted. The neonate required no mechanical respiratory support, since it did not obstruct the airway.

Postnatal MR imaging on day 2 of life revealed a lobulated, well-defined mass arising from the maxillary ridge without extension into the nasal airway, soft palate, floor of the mouth, mouth, nose or cranium [Table/Fig-2]. The unerupted upper teeth appeared normal. The history, clinical features & MRI of the lesion suggested its benign nature.



[Table/Fig-1]: Postnatal photograph shows lobulated mass, originating from the gum pad of the maxillary ridge



[Table/Fig-2]: MR image shows the smooth, well-defined mass arising from the maxillary ridge without extension into the nasal airway, soft palate, floor of the mouth, mouth, nose or cranium

Three days after birth, the infant was operated on, under general anaesthesia, with oral intubation. The operative and postoperative course was uneventful. Oral feedings were instituted on the first postoperative day and the child was discharged on the following day. Healing was uneventful.

Histologic examination of the specimen revealed an unencapsulated lesion covered with squamous epithelium. The lesion was composed of homogeneous cells with granular eosinophilic cytoplasm and basophilic centrally located nuclei. These findings were consistent with congenital granular cell tumour of the newborn.

DISCUSSION

CE occurs in the mucosa of the maxillary/ mandibular alveolar ridge as a smooth surfaced pink mass. A MEDLINE search conducted by Kayiran et al., [8] revealed 8 reported cases on the tongue, either in isolation, or along with gum lesions. It is usually not associated with any congenital abnormalities. However large lesions can cause hypoplastic jaws or teeth, and mid-face hypoplasia and is called 'Binder Syndrome' [4,6,9].

In a review of 113 cases between 1871-1971, Fuhr and Krogh [10] noted an 8-10:1 female predilection, and 3:1 maxillary alveolar site predilection [4,7], usually a solitary mass, although 10% cases may occur as multiple lesions [4,5,7]. The size of the CE reportedly varies from few mm to 9 cm [1,7]. Large lesions can interfere with fetal deglutition, resulting in polyhydramnios, respiratory obstruction, and difficulty in feeding postnatally [3,4]. The size of the lesion, in our case, was four cm in length, and was interfering with feeding.

The proposed cells of origin include odontogenic epithelium, undifferentiated mesenchymal cells, pericytes, fibroblasts, smooth muscle cells, nerve related cells and histiocytes [11]. Histologically, CE shows highly vascularised fibrous tissue, with nests of polygonal cells with large clear and granular cytoplasm, and a small nucleus, with a normal overlying epithelium [11]. While the granular cells of CE are quite similar to the cells of the true granular cell tumors (granular cell myoblastoma), the histology and epidemiology of the two lesions differ. CE occur only on the gumpads of neonates, while granular cell tumors occur only in adults (20-60 years age), and may involve multiple organs- tongue (30-50%), skin (30%) [11]. Other sites of the adult tumors include supraglottic and infraglottic airways, bronchus, mastoid, orbit, breast, muscle and lip [12]. Granular cell tumours are less vascular, often have a component of pseudoepitheliomatous hyperplasia, and contain more nerve bundles than does congenital epulides [12]. Less than 1% of granular cell tumours are malignant, and some authors argue that those are actually misdiagnosed sarcomas, histiomas and rhabdomyosarcomas [12].

Electron microscopic study of CE showed granular cells containing heterogeneous electron dense granules, lysosomes and cytoplasmic lipid droplets. The cells have

irregular cytoplasmic borders with small extensions [1]. CE shows positive staining for vimentin either in the intercellular spaces or in the cytoplasm of granular cells, explained by the abundance of collagen and its precursors. Positive reaction for desmin, a 53 kD fibrillar protein, is a specific marker of muscle cells. Ultra structural signs of smooth muscle differentiation were first detected in CE in 1983 and recently confirmed by other authors [4,5].

Sonographic prenatal diagnosis of congenital abnormalities is an invaluable tool in the management of pregnancy, delivery and postnatal treatment of the affected newborn. The earliest antenatally diagnosed case was reported in a 31 week fetus. Prenatal diagnosis of CE is possible with ultrasonography, after 25th week of intrauterine life, when the images of the fetal face reveal a homogenous well-circumscribed, non-septate oral mass, with doppler finding inconsistent with hemangioma [1,13,14]. A marked blood supply through the vascular pedicle is characteristic [15]. Potential impairments may necessitate rapid intervention requiring EXIT procedures [14,15].

The postnatal MR imaging findings of a mass isolated to the gingiva, without involvement of the unerupted teeth, are also non-specific, and would include other non-specific masses such as fibromas, and vasoformative lesions.

Small lesions (<2cm) may regress spontaneously, as reported in approximately 8 of the 200 reported cases [1,16]. Larger lesions need resection, as they often interfere with airway patency, and cause feeding difficulties. Removal of CE by surgery under local or general anesthesia [1,7,17] electrocautery [17] and carbon dioxide laser set at 15 watts continuous wave, has been reported [1]. Complete surgical resection is curative. Recurrence of the lesion and damage to the future dentition has not been reported, suggesting that radical excision is not warranted [1].

CONCLUSION

Congenital Epulis is a rare, benign, gingival lesion seen exclusively in neonates. It is relatively commoner in Caucasians, and less frequent in Asians. Cases of spontaneous regression, lack of recurrence even after incomplete removal and lack of malignant counterpart suggest CE is a non-neoplastic lesion and needs to be differentiated from granular cell tumours. Prenatal ultrasonographic diagnosis of CE is possible after 25th week of intra uterine life. Surgical excision is curative, with no reported recurrences.

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