

# A Large Congenital Granular Cell Tumor: A Case Report

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(Received: September, 2014)

(Accepted: January, 2015)

## ABSTRACT:

Congenital granular cell tumor (CGCT) is a rare benign neoplastic growth of the anterior maxilla of neonates. The tissue origin of this lesion whether it is from neural or muscular; is still debatable although many immunohistochemical markers studies have been documented. We present a case report of a female neonate with a large CGCT along with a brief review of literature.

**KEY WORDS:** congenital, epulis, granular, neonate, tumor

## INTRODUCTION:

Congenital granular cell tumour (CGCT) is an uncommon congenital benign epulis of the neonate. The original first description of the lesion was dated in 1871 by Neumann.<sup>[1]</sup> CGCT is most frequently seen on the alveolar median ridge of the maxilla than alveolar ridge of mandible(1:3) in the incisor-canine area. Characteristically CGCT has shown a female predominance. The presentation of the lesion is unique as it is seen at birth as a firm fibrous mass arising from the gingival mucosa. This anterior jaw tumor can be obstructing and can complicate respiration as it may block the nasal and oral airway passage. Feeding of the neonate may be an issue of concern as lesion may obliterate the entire mouth opening. Surgical excision has to be done on priority basis to avoid further complications.<sup>[2]</sup>

## CASE REPORT:

A 2-day-old new-born girl was referred from a rural hospital to the General Surgery Department of People's hospital, Bhopal, India. The baby presented with a large mass protruding from the mouth with compromised respiration (Figure 1). The infant was

born in 36th week of gestation after a period of normal pregnancy. The girl infant was weighing 2.45 kg and exhibited healthy clinical parameters. The mother was a primigravida but as she belonged to a rural place, no antenatal ultrasonography check-up was done. Details of medication during gestation were not recorded and there was no family history of any congenital anomaly.

Clinical examination revealed a large pedunculated, bilobed, exophytic fibrous growth arising from the upper right anterior alveolar ridge measuring 4 × 3 × 3 cm. The growth was having irregular reddish-black surface showing areas of crustation and haemorrhages. The mass was completely obstructing the oral cavity and partially the left nostril. The infant was being fed with a nasogastric tube.

The tumor was excised under general anaesthesia. An elliptical incision was given to the pedicle and the tumor was mobilised and removed in toto (Figure 2). Haemostasis was achieved with diathermy followed by wound closure. The postoperative recovery was uncomplicated and remarkably splendid and uneventful (Figure 3).

The tumor was sent for histopathological examination. Hematoxylin and eosin stained section showed the overlying normal gingival oral mucosa without hyperplasia (Figure 4). The connective tissue showed vascular-rich stroma with large sheets of medium sized, ovoid and polygonal granular cells. The cells were packed with fine granular eosinophilic

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cytoplasm and had vesicular nuclei. Areas of haemorrhage were seen. The diagnosis was confirmed as congenital granular cell tumor.

Patient was recalled after few days post-surgery and was kept on regular follow-up with no sign of recurrence in a period of 6 months, whereas no recurrence was observed in a follow-up period of 2 years.

## DISCUSSION:

CGCT or congenital epulis is an uncommon tumour of the neonate. Approximately 250 cases have been reported since first case reported in 1871.<sup>[3]</sup> Other



**Figure 1:** The girl neonate presenting with a firm protruding mass from oral cavity associated with compromised respiration and feeding.



**Figure 2:** Excised specimen was a single lobulated firm fibrous mass measuring approximately 4 × 3 × 3 cm.

terminologies for CGCT described in literature are *congenital epulis*, *Granular cell rhabdomyoma*, *congenital myoblastoma*, or *Neumann's tumour*. "Congenital granular cell tumor" (CGCT) is nowadays frequently employed terminology in the literature and the same is used in our case.<sup>[4]</sup>

Estimation of Incidence of the CGCT in world population is 0.0006 %<sup>[5]</sup>. CGCT appears sporadically, and with no familial tendency. It most commonly originates from the anterior alveolar ridge, with the maxilla being involved twice as much as the mandible though the lesion can arise in small number cases from the tongue, palate, skin, the subcutaneous tissue, skeleton muscles and the vocal chords.<sup>[6]</sup> This peculiar tumor has a smooth or lobular surface and a firm rubbery consistency.<sup>[7]</sup>

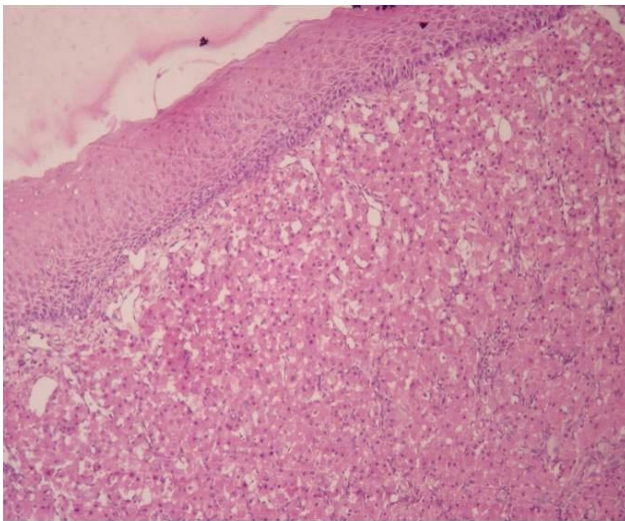
Prenatal diagnosis of CGCT can aid in counselling the parents as to the nature and treatment of the abnormality, as well as assessing the potential risk of airway obstruction and intraoperative complications. Ultrasonography (USG) or Magnetic resonance Imaging (MRI) prenatal imaging of congenital lesions of oral cavity can be helpful in such cases.<sup>[8]</sup> Prenatal imaging is important in the choice of delivery method, since large tumors may compromise a normal vaginal delivery and a caesarean operation may be necessary.<sup>[9, 10, 11]</sup> Prenatal diagnosis of the lesion on with USG was reported in the literature as early as 26<sup>th</sup> week to 38 weeks which mostly coincides with third trimester of the pregnancy. The lesion is not detectable till that period; reason for this is unclear.<sup>[11]</sup>

CGCT has unique female preponderance. Based on the theory supported by the experimental production of CGCT in a mouse following injection of endogenous hormone, this characteristic finding was suggested to be attributable to the presence of endogenous hormones and sudden regression of mass after birth.<sup>[12]</sup> This was subsequently proved failed as there was no detectable presence of these endogenous hormone receptors with the lesion tissue. Trauma, due to finger sucking in utero, was also suggested by some authors.<sup>[9, 13, 14]</sup> Recently, it was suggested that it may be a local metabolic or reactive change.<sup>[15]</sup>

CGCT mostly presents as an isolated mass in an otherwise healthy infant. Some authors have reported association with other congenital anomalies like bilateral transverse facial cleft, polydactyl, congenital goitres and also with neurofibromatosis.<sup>[16, 17]</sup>



**Figure 3:** Post-operative appearance and recovery was remarkably splendid.



**Figure 4:** Large sheets of polygonal granular cells in the stroma were the diagnostic feature. (H and E stain)

Clinical differential diagnosis is broad based and depends upon the site of involvement; size and velocity of growth. Various similar presenting lesions that can be considered include fibroma, lipoma, leiomyoma, rhabdomyoma, peripheral giant cell granuloma, pyogenic granuloma, myxoma, neurofibroma, haemangioma, lymphatic malformations, melanoameloblastoma of infancy, granular cell tumor, oral teratoma, dermoid cyst etc.<sup>[18]</sup> The accurate diagnosis can be established histopathologically in most of the cases. Histopathological similarity is seen with the Granular cell myoblastoma which is also called as granular cell tumor (GCT) of adults which is seen in

tongue. CGCT can be separated from Granular cell tumor (GCT) by location, patient's age, and few minor histopathologic differences (Table 1).<sup>[19]</sup>

**Table 1:** Difference between the Adult granular cell tumor and congenital Granular cell tumor.<sup>[19]</sup>

Adult Granular cell tumor	Congenital Epulis (CGCT)
Occurs in adult between 20-60 years	Occurs in New born
Involves Multiple organs	Only in Gum pads
A malignant variant is reported	Does not recur and no malignant potential
Histopathology (HP) pseudoepitheliomatous hyperplasia present	Absent
HP shows more conspicuous nerve bundles.	Less conspicuous nerve bundles.
HP shows less vascularity	More vascularity in a plexiform arrangement
Immunohistochemistry study express S-100 protein markers	Negative for S-100 protein

The histogenesis of CGCT has also been debatable as various authors suggested different source of origin of the tumour. The proposed source of origin includes undifferentiated mesenchymal cells, odontogenic epithelial, pericytic, and fibroblastic, histiocytes, nerve-related, smooth muscle, and primitive mesenchymal cells. The histogenesis remains still unclear despite documentation by several studies. Immunohistochemical analysis of this case was negative for the neural tissue marker S100.

Treatment of choice of CGCT is prompt surgical excision, although spontaneous regression has been reported in isolated few cases.<sup>[20]</sup> Delay in surgical removal can lead to airway obstruction and major feeding difficulty for the infant. Recurrence or malignant changes in CGCT has not been documented in literature.<sup>[13]</sup>

**CONCLUSION:**

CGCT should be given considerable clinical importance as it has potential to complicate feeding and respiration of affected infants if it is not promptly treated.

The lesion still poses scope for further research

in its etiopathogenesis especially by immunohistochemical methods. The diagnosis by various methods during prenatal period also should be assessed for this entity.

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**How to cite this article:** Mhaske A, Mhaske S, Yuwanati M: A Large Congenital Granular Cell Tumor: A Case Report. *PJSR* 2015; 8 (1):46-49

**Source of Support:** Nil, **Conflict of Interest:** None declared.