



Case Report

Congenital Granular Cell Tumor in an Infant

Mudit Agarwal

Director Lab Services, Nishtha Pathology Lab, New Delhi, India

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ABSTRACT

Congenital granular cell tumor (CGCT) is a rare, benign soft tissue tumor seen in newborns. The tumor is quite uncommon, with an incidence of only 0.0006% and less than 250 cases have been reported in the literature so far. The lesion has a marked female preponderance, with the female:male ratio being 8-10:1. It usually occurs on the gingival of the anterior alveolar ridge, more frequently in the maxillary alveolus than in the mandibular alveolus.

In this case report, we present a congenital granular cell tumor, sent by a paediatric surgeon as histopathological tissue from the oral cavity of 1 day old male baby, delivered with a solitary polypoidal nodule projecting from the mouth and presenting as difficulty in breast feeding. The mass was sent in toto and was diagnosed on histopathological examination.

***Corresponding author:**

Dr Mudit Agarwal; Director Lab Services, Nishtha Pathology Lab, Shop No 3 Ground Floor, Main Road, Opposite Water Tank, ASHok Vihar Phase 1, New Delhi, 110052 India
Phone: +91 011 2040 7807

Introduction

Congenital granular cell tumor (CGCT) is a rare, benign soft tissue tumor seen in newborns.¹ It was first described by Neumann in 1871 and therefore it is also known as Neumann's tumor.² Additional nomenclature for this tumor includes congenital granular cell epulis, congenital granular cell lesion, gingival granular cell tumor, granular cell tumor of the newborn, granular cell myoblastoma and granular cell fibroblastoma.³

The tumor is quite uncommon, with an incidence of only 0.0006% and less than 250 cases have been reported in the literature so far.⁴ The lesion has a marked female preponderance, with the female:male ratio being 8-10:1.⁵ It usually occurs on the gingival mucosa of the anterior alveolar ridge, more frequently in the maxillary alveolus than in the mandibular alveolus.⁶

In the present case report Congenital Granular cell tumor was sent as histopathological tissue from the oral cavity by a paediatric surgeon of 1 day old male baby delivered with a solitary polypoidal nodule projecting from the mouth and presenting as difficulty in breast feeding was sent in toto and was diagnosed on histopathological examination.

Case Report

A 2-day -old female child was referred with the chief complaint of difficulty in feeding since a few days. On examination, a 1.5 × 1.5 cm, pedunculated growth was observed arising from the left side of maxilla in the canine region. The lesion was pink in color, similar to that of the surrounding mucosa, surface was smooth without any ulcerations and was firm in consistency. The patient was uncomfortable because of this small growth which was possibly hindering proper suction during breast feeding



Fig. 1: Gross Examination before surgery

(Figure 1). Surgical excision of the growth was performed under local anesthesia. There was minimal bleeding during the procedure and the infant was comfortable with breast feeding from the very next day. The gingival mass was fixed in formalin and sent for histopathological examination.

Gross examination showed a tumor with a firm round surface projecting from the mouth cavity with a narrow stalk. The surface was pink, measured 2 cm in diameter, and showed variegated appearance (Figure 2).

Low power microscopic examination of the sections from the polyp showed a well circumscribed tumor covered with a keratinized squamous epithelium. The tumor was composed of lobules and nests of spindle to polygonal, uniform-looking cells, having abundant eosinophilic, granular cytoplasm. The nuclei showed no features of atypia. A rich vascular network was seen surrounding the tumor cells. (Figure 3A, 3B).

Discussion

Congenital granular cell tumor is a rare neoplasm, usually seen in newborns and infants. It characteristically presents at birth as a solitary, polypoid nodule which is firmly attached to the dental ridge by a narrow or broad stalk.⁷ The tumor is typically 1 to 2 cm in diameter but sizes up to 9 cm have been described.⁸ It is often covered by a smooth, pink mucosa which may sometimes be erythematous or ulcerated.³ They are usually not associated with any dental abnormalities or congenital malformations.⁹

The tumor can be diagnosed prenatally by obtaining intrauterine images by ultrasonography or magnetic resonance imaging. These may be used to guide in the choice of delivery method since presence of large tumors may require delivery by caesarean section.¹⁰



Fig. 2: Gross Findings

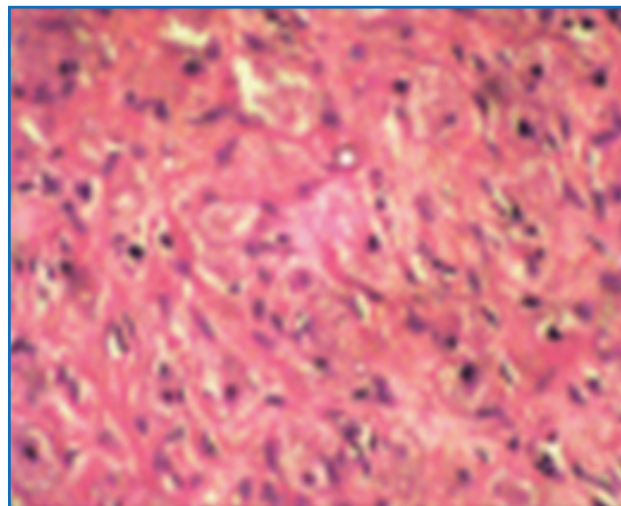
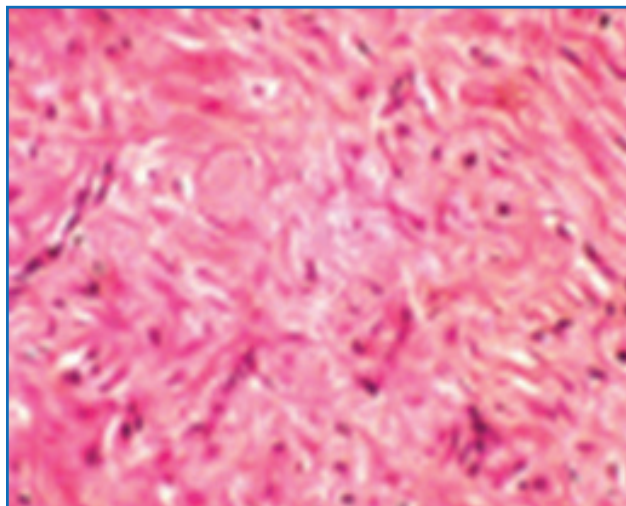


Fig. 3A,3B: Microscopic Examination revealing the granular cells (H& E stain 40 X)

Spontaneous regression of the tumor is rare and large lesions may cause airway obstruction and feeding difficulties.¹⁰ Therefore, the recommended course of action is to surgically excise the tumor promptly after diagnosis.¹¹ A conservative and non-mutilating approach is preferred as there are no reports of local recurrence after excision and the tumor carries an excellent prognosis.^{5,12}

The histogenesis of CGCT is unclear and multiple theories of derivation have been proposed, including origination from odontogenic epithelium, neuroendocrine progenitor cells or undifferentiated mesenchymal cells. Post degenerative or reactive changes have also been suggested.³ Due to its female preponderance, another theory proposed is that of endogenous hormonal stimulation in-utero. However, this theory is unaccepted due to lack of estrogen or progesterone receptors within the tumor.^{4,10}

Light microscopy of CGCT shows keratinized stratified squamous epithelium with the deep dermis shows fairly well-circumscribed tumor, composed of nests and ribbons of lightly packed, medium to large, polygonal to slightly spindled, uniform looking cells with an eosinophilic granular cytoplasm. Nuclei are eccentrically placed with no atypia and prominent nuclei. The tumor has a prominent capillary network.^{3,13,14}

Immunohistochemically, the granular cells in CGCT are positive for vimentin, HLA-DR antigen, and occasionally positive for neuron-specific enolase and carcino-embryonic antigen. They are typically negative for S-100 protein, ruling out a Schwannian origin. They are also negative for alpha feto-protein, actin, laminin and specific macrophage markers.^{5,15}

The main histopathologic differential diagnosis of CGCT is adult granular cell tumor.³ However, it is seldom seen

in children and microscopically shows less vascularity than CGCT and the overlying epithelium shows pseudoepitheliomatous hyperplasia. Additionally, adult GCT is positive for S-100 protein on IHC as shown in (Figure 2).¹⁶

Other important differential diagnoses to be considered are melanotic neuroectodermal tumor of infancy, hemangioma, fibroma, embryonal rhabdomyosarcoma, granuloma, malignant granular cell myoblastoma, chondrogenic and osteogenic sarcoma and schwannoma.^{3,4}

Conclusion

To conclude, the congenital granular cell tumor of infancy carries a life threatening problem and needs to be differentiated from Adult congenital granular cell tumor as the two differ in their treatment approach and the former carries an excellent prognosis.

Thus congenital cell tumor of infancy can present as incidental finding at birth if not diagnosed on USG or MRI Scan antenatal and needs immediate surgical intervention and can be diagnosed on microscopic examination without any significant gross abnormality and needs to be distinguished from adult congenital tumor in view of appropriate treatment.

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Competing Interests

None declared

References

1. Bosanquet D, Roblin G. Congenital Epulis: A Case Report and Estimation of Incidence. *International Journal of Otolaryngology*. 2009;2009:1-3.
2. Zeren T, Sencimen M, Altun C, Altug H. Congenital granular cell tumor in newborn. *European Journal of Dentistry*. 2013;7(4):497.
3. Conrad R, Perez M. Congenital Granular Cell Epulis. *Archives of Pathology & Laboratory Medicine*. 2014;138(1):128-131.
4. Kumar R, Jaiswal S, Singhal A, Garg R. Congenital granular cell lesion: A rare tumor of new born. *Journal of Oral and Maxillofacial Pathology*. 2013;17(3):440.
5. Fister P, Volavsek M, Novosel Sever M, Jazbec J. A newborn baby with a tumor protruding from the mouth. *Acta Dermatovenereol Alp Panonica Adriat* 2007;16(3):128-30.
6. McGuire TP, Gomes PP, Freilich MM, Sandor GKB. Congenital Epulis: A Surprise in the Neonate. *J Can Dent Assoc* 2006; 72(8):747-50.
7. Weiss SW, Goldblum JR. *Enzinger & Weiss's Soft Tissue Tumors*. 5th ed. Philadelphia, PA: Mosby-Elsevier; 2008:878-889.
8. Eghbalian F, Monsef A. Congenital epulis in the newborn, review of the literature and a case report. *J Pediatr Hematol Oncol*. 2009;31(3):198-199.
9. Koch BL, Myer C 3rd, Egelhoff JC. Congenital epulis. *AJNR Am J Neuroradiol* 1997; 18(4):739-41.
10. Silva GCC, Vieira TC, Vieira JC, Martins CR, Silva EC. Congenital granular cell tumor (congenital epulis): A lesion of multidisciplinary interest. *Med Oral Patol Oral Cir Bucal*. 2007 Oct 1;12(6):428-30.
11. Lapid O, Shaco-Levy R, Krieger Y, Kachko L, Sagi A. Congenital Epulis. *PEDIATRICS*. 2001;107(2):e22-e22.
12. Dash JK, Sahoo PK, Das SN. Congenital granular cell lesion congenital epulis – report of a case. *J Indian Soc Pedod Prev Dent* 2004;22(2):63-7.
13. Regezi JA, Sciubba JJ, Pogrel MA. *Atlas of Oral and Maxillofacial Pathology*. Philadelphia, PA: WB Saunders; 2000:66.
14. Wenig BM. *Atlas of Head and Neck Pathology*. 2nd ed. Philadelphia, PA: Saunders-Elsevier; 2008:289-290.
15. Ugras S, Demirtas I, Bekercioglu M, Kutluhan A, Karakok M, Peker O. Immunohistochemical study on histogenesis of congenital epulis and review of the literature. *Pathol Int* 1997;47(9):627-32.
16. Gokhale UA, Malhotra CJ. Congenital epulis of the newborn. *Indian J Pathol Microbiol* 2009; 52(3):436-7.