DOI: 10.7860/IJNMR/2016/20265.2174

Case Report

Neonatology Section

Congenital Granular Cell Tumor of a Newborn: A Case Report

CHHANDA DAS, MADHUMITA MUKHOPADHYAY, SUCHARITA SARKAR, BEDABRATA MUKHOPADHYAY

ABSTRACT

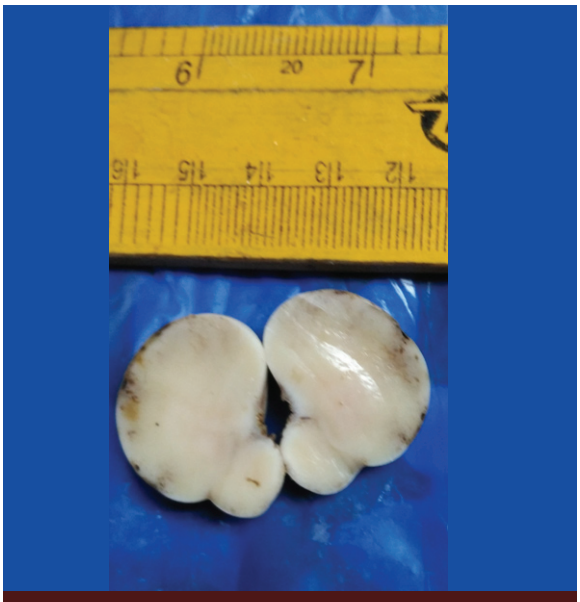
Congenital Granular Cell Tumour (CGCT) is a rare benign soft tissue tumour of newborn, different from adult GCT. The tumour has a strong predilection for the maxillary alveolar ridge with a female preponderance. Here we report an unusual case of congenital granular cell epulis in the mouth of a 2 hour old female newborn. She had a round, soft, pedunculated mass measuring (2x 2x1) cm. which was causing feeding difficulty. This case was reported as congenital granular cell epulis. We are presenting this case because of its rare nature and uncommon presentation at this age.

Keywords: Epulis, Intraoral, Pedunculated.

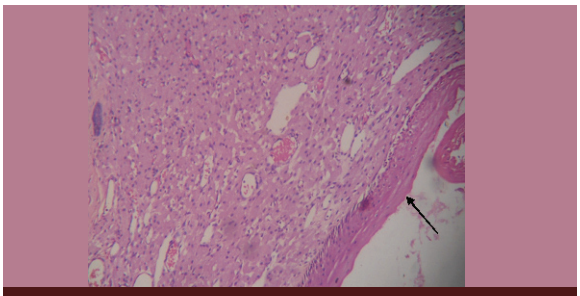
CASE REPORT

A full term newborn female child delieved 2 hours ago in our hospital, presented with a complaint of huge intraoral mass of tissue arising out of the lower gum. Her birth history was uneventful .

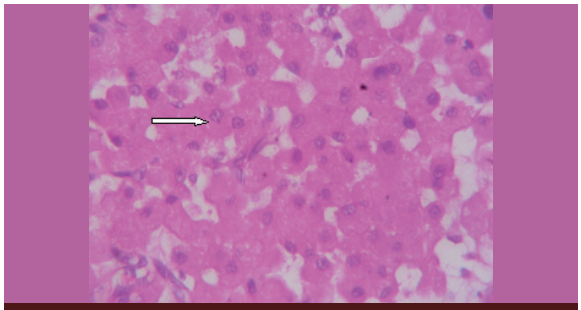
Clinical examination revealed a large mass of soft tissue projecting outward from lateral maxillary alveolar area. The intraoral mass was pedunculated and firmly connected to the alveolar ridge. The mass was slight pale yellow in colour and underlying mucosa was reddened. The mass was obstructing the feeding as it was causing difficulty in sucking process. Though, no airways obstruction was there. The child was operated within few hours after birth under general anaesthesia, after taking consent from her parents. Excised mass was sent to Department of Pathology, for histopathological examination. On gross examination the mass was described as single globular tissue with the total measurement of 2x2x1cm. On cut sections solid white homogenous tissue identified. No bony tissue was seen [Table/Fig-1]. Multiple sections were taken from the different areas of the mass for routine histopathological examination. After processing 4 µm-thick sections were cut from paraffin-embedded tissue blocks and stained with haematoxylin and eosin. On microscopic examination of H & E stain showed clusters of large polygonal cells with abundant granular, eosinophilic cytoplasm and oval basophilic nuclei. The overlying mucosa showed a well-differentiated, attenuated squamous epithelium [Table/Fig-2&3]. Histopathology was compatible with granular cell tumour (epulis). Immunohistochemistry was done. It showed lack of staining for S-100 [Table/Fig-4]. Follow-up of this case revealed no recurrence, the child started was gaining weight and no feeding difficulty.



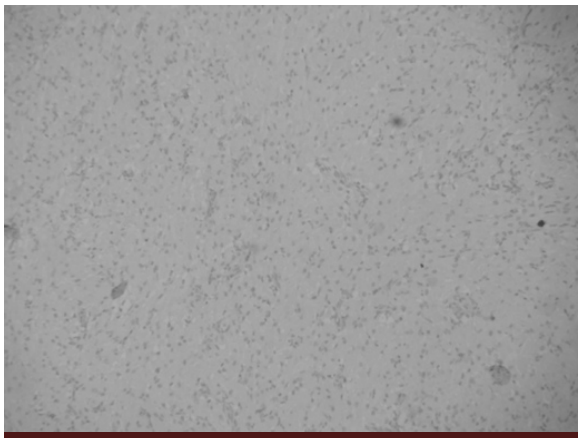
[Table/Fig-1]: Gross picture of the tumor.



[Table/Fig-2]: Histological examination of tumor showing attenuated epithelium (x100).



[Table/Fig-3]: Clusters of polygonal cells with abundant granular, eosinophilic cytoplasm (x400).



[Table/Fig-4]: IHC shows S-100 negativity (x100).

DISCUSSION

Congenital granular cell tumour or congenital epulis (meaning- on the gum) was first described by Neumann in the year of 1871 [1], a benign soft tissue tumour almost exclusively found in neonate, commonly manifest within first month of life [2]. Lesions protruding from the neonate's mouth presents with either normal coloured or reddish and sessile or pedunculated mass. Gingiva is the most common site, varying in size from several millimetres to a few centimetres [3]. Surgical excision is a treatment of choice for CGCT. Spontaneous regression may occur even in case of incomplete resection [3]. Large tumours cause airway obstruction and feeding difficulties. The tumour has predilection for the upper jaw and characteristically found in labial aspect of dental ridge [3] .There are several controversy about the histogenesis of CGCT. The tumour has an mesenchymal origin (fibroblast, myofibroblast, histiocytes, or Schwann cell [3]. On the other hand, some researchers suggest that the tumour is neuroectodermal in origin [4]. Multiple lesions are found in 10 % of cases [5]. CGCT is a rare type among gingival growth which has an incidence of just 0.0006 % [6]. The usual location is alveolar mucosa, more frequently in the anterior maxilla in the midline in the region of the primary canine and lateral incisor, less frequently over mandible and tongue [7]. Maxillary/ mandibular ratio is 3:1 [8,9]. Female babies are more affected the males approximately 90% of case [3] with a female: male ratio of 8:1 [10]. The tumour has no tendency to local recurrence or malignant transformation [3]. Grossly the tumour may present with a firm pedunculated mass arising out of alveolar mucosa by means of a stalk [11]. Adult GCT is the main differential diagnosis which is rarely seen in the first decade of life, it is most frequently diagnosed between the 3rd to 6th decades of life [3] with no sex predilection [12]. The CGCT showed positive immunohistochemical staining to HLA-DR antigen, vimentin, NKI/C3, and PGP9 and occasionally NSE and CEA and negative for S-100 protein , NGFR/P75 and PGP 9.5 in contrast to adult variant [3,13]. Histologically this tumour is different from adult granular cell tumour by absence of pseudoepitheliomatous hyperplasia of surface epithelium, prominent vascularity and presence of scattered remnants of odontogenic epithelium [3].

Other important differential diagnosis are congenital dermoid cyst, congenital cyst choriostoma, congenital lipoma, heterotopic gastrointestinal cyst, rhabdo-myoma, hemangioma, lymphangioma and congenital fibrosarcoma osteogenic and chondrogenic sarcomas [14,15]. Associated malformation as reported are Triple X-syndrome, neurofibromatosis, polydactyly and polyhydraminos and maxillary hypoplasia [15]. Though diagnosis can be confirmed by histopathology and immune profiling but immunohistochemistry cannot confirm histogenesis of the tumour [16].

CONCLUSION

CGCT is relatively uncommon benign tumour arising from the gingival mucosa typically from the anterior maxilla as a midline lesion. The female newborn presented in this case is consistent with the age and sex predilection of CGCT. Reporting of such rare cases will expect to facilitate the narrowing down of differential diagnosis of oral lesions presenting at or immediately after birth and proper diagnostic approach and treatment planning.

REFERENCES

[1] Neuman E. Elin Fall von Congenitaler Epulis. *Arch Heilk*. 1871;12:189.

[2] Kumar R, Jaiswal S, Singhal A, Garg R. Congenital granular cell lesion: A rare tumour of newborn. *J Oral Maxillofac Pathol*. 2013;17(3):440-42.

[3] Rosai J. Peritonium, retroperitoneum, and related structures. In: Juan Rosai – Rosai And Ackerman's Surgical Pathology. vol 1 .10th ed. New Delhi: Reed Elsevier India Private Limited,2012:252.

[4] Liang Y, Yang YS and Zhang Y. Multiple congenital granular cell epulis in a female newborn: a case report. *Journal of Medical Case Reports*. 2014; 8:413.

[5] Congenital (gingival) Granular cell tumour: Sharon W. Weiss, John R. Goldblum et al editors Enginger & Weiss's Soft Tissue Tumours, 5th ed, Elsevier Inc, 2008:

[6] Bosanquet D, Robin G. CGCL: A case report and estimation of incidence. *Int J Otolaryngol*. 2009;31:198-99.

[7] Hasanov A, Musayev J, Onal B, Rahımov C, Farzaliyev I. Gingival granular cell tumor of the newborn: a case report and review of literature. *Turk Patoloji Derg*. 2011; 27(2):161-63.

[8] Loyola AM, Gatti AF, Pinto DS Jr, Mesquita RA. Alveolar and extra-alveolar granular cell lesions of the newborn:

report of case and review of literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.* 1997;84(6):668-71.

[9] Olson JL, Marcus JR, Zuker RM: Congenital epulis. *J Craniofac Surg*. 2005;16:161–64.

[10] Kanotra S, Kantra SP, Paul J.Congenital epulis. *J Laryngol Otol*. 2006;120:148-50.

[11] Silva GC, Vieira TC, Vieira JC, Martins CR, Silva EC. Congenital granular cell tumour (congenital epulis):a lesion of multidisciplinary interest. *Med Oral Patol Cir Bucal*. 2007;12:E428-30.

[12] Aparna HG, Jayanth BS, Shashidara R , Jaishankar P. Congenital epulis in a newborn :A case report, immunoprofiling and review of literature. *Ethiop J Health Sci*. 2014;24(4):359-62.

[13] Adeyemi BF, Oluwasola AO, Adisa AO. Congenital epulis. *Indian J Dent Res.* 2010; 21(2):292-94.

[14] Szlachetka K, Bern EML, Ozcan T. et al. Prenatal diagnosis of a rare gingival granular cell tumour of the fetal mouth. *J Ultrasound Med*. 2012;31(1):125-27.

[15] Gokhale UA, Malhotra CJ. Congenital epulis of the newborn. *Indian J Pathol Microbial*. 2009; 52(3):436-37.

[16] Ugras S, Demírtas I, Bekerecíoglu M, Kutluhan A, Karakök M, Peker O. Immunohistochemical study on histogenesis of congenital epulis and review of the literature. *Pathol Int*. 1997; 47(9):627-32.

AUTHOR(S):

1. Dr. Chhanda Das
2. Dr. Madhumita Mukhopadhyay
3. Dr. Sucharita Sarkar
4. Dr. Bedabrata Mukhopadhyay

PARTICULARS OF CONTRIBUTORS:

1. Assistant Professor, Department of Pathology, The Institute of Post Graduate Medical Education and Research, West Bengal, India.
2. Professor Department of Pathology, The Institute of Post Graduate Medical Education and Research, West Bengal, India.
3. PGT, Ist year, Department of Pathology, The Institute of Post Graduate Medical Education and Research, West Bengal, India.
4. PGT, 3rd year, MD Department of Biochemistry, Institute of Medical Sciences Banaras Hindu University, Banaras, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Chhanda Das,
31 Eastern Park, First Road, Santoshpur.
Kolkata-700075, West Bengal, India.
Email: chhhdas@gmail.com

FINANCIAL OR OTHER COMPETING INTERESTS:
None.

Date of Publishing: **Jul 01, 2016**