Congenital Granular Cell Epulis (Neumann's Tumour): A Rare Case Report

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Abstract- Neumann's Tumour/Congenital Granular Cell Tumour (CGCT)/Granular Cell Epulis Of Infancy is a rare lesion of newborn. It is benign in nature, arising from the alveolar ridge of the gingiva. It is seen more frequently in female neonates. It may cause respiratory and feeding problems.Herewith, we present a case report of CGCT arising from the upper gingival margin of female neonate, treated with surgical excision. We report this rare case in view of its unusual resemblance with Granular Cell Myoblastoma though the histogenesis is different.

Index Terms- Neumann's Tumour, CGCT, Adult granular cell tumour, myoblastoma

I. Introduction

Congenital granular cell tumour (CGCT) of the newborn, also known as Neumann's Tumour is an uncommon lesion, described for the first time in 1871 by Neumann [1],[2]. To date, fewer than 200 cases have been described in the literature [3]. CGCT usually presents at birth has a distinct predilection for females and is most frequently located on the anterior maxillary alveolar ridge [4-8]. It is presented as multiple lesions in only 10% cases[9].

The size of the mass varies from a few millimeters to 9 cm in diameter on gross[9].They can interfere with feeding and respiration. The recommended treatment is surgical excision under local or general anesthesia, although spontaneous regression has been reported in very few cases. There are no reports of recurrence, even if incomplete margins are excised, malignant change, or future disruption to teeth or gums.[9]

The lesion usually appears as a protuberant mass, sometimes pedunculated. Histologically it shows characteristic large cells with granular cytoplasm and spindle cells resembling fibroblasts. The exact pathogenesis of CGCT is still uncertain, as is its growth and progression.

We report a case of CGCT observed in a newborn female, with immunohistochemical confirmation. We also have highlighted the difference between the adult granular cell tumour (Granular cell Myoblastoma) and CGCT.

II. Case Report

A 14 days old, otherwise healthy female neonate was brought with complaint of a mass protruding from her mouth (Figure 1). The baby was born at the 36th week of gestation. Antenatal period and the delivery were uneventful. The birth weight was 2600 grams. A firm, pedunculated swelling arising from alveolar ridge of right maxillary central incisor area was present since birth. The mother reported that the baby did not have feeding problems or airway obstruction. . Clinical diagnosis of benign soft tissue tumour was made and the swelling was excised under local anesthesia and sent for Histopathology.

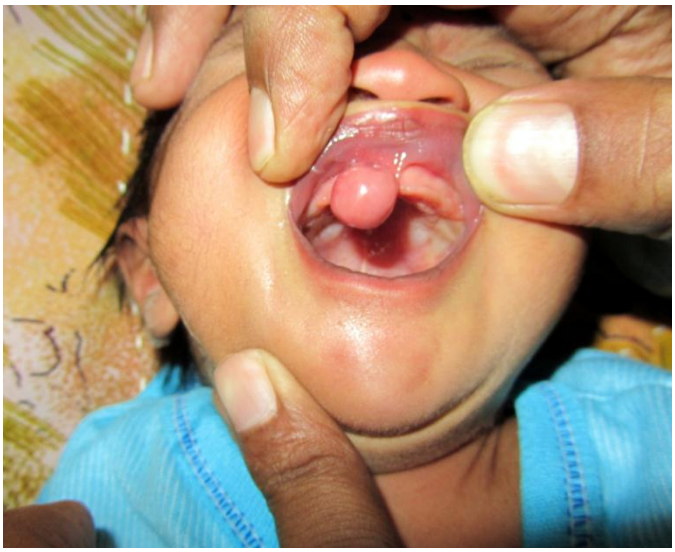

Figure 1: Clinical photograph of patient showing pedunculated swelling arising from alveolar ridge of right maxillary central incisor area.

Pathology

Gross : A single, small, round, pedunculated, grayish white, firm mass with smooth outer surface was received. Diameter was 0.8cm.On cut section the mass was grayish white, homogenous.

Microscopy: The mass comprised almost entirely of sheets and clusters of large, round to polyhedral, closely packed cells with small, uniform, dark oval nuclei and abundant fine granular eosinophilic cytoplasm[1],[10],[11]. Multiple vascular channels were evident in between granular cells. Fibrous stroma was minimally present. Tumour cells extended to the overlying stratified squamous epithelium (Figures 2,3). With this classical histological pattern, a diagnosis of congenital granular cell tumour was rendered. Immunohistochemistry further confirmed the diagnosis as the tumour cells were diffusely and strongly positive for Vimentin and negative for S-100 protein .(Figures 4,5)

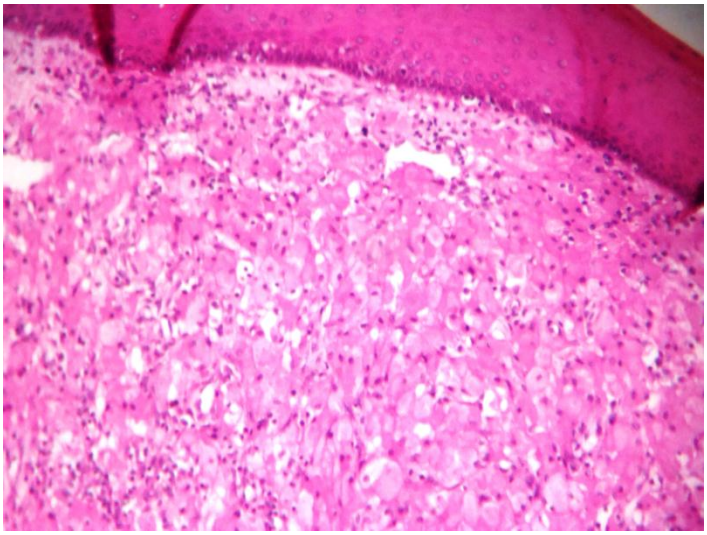

Figure 2: Clusters of large, round to polyhedral cells with small, uniform, dark oval nuclei and abundant fine granular eosinophilic cytoplasm (H & E ,10X)

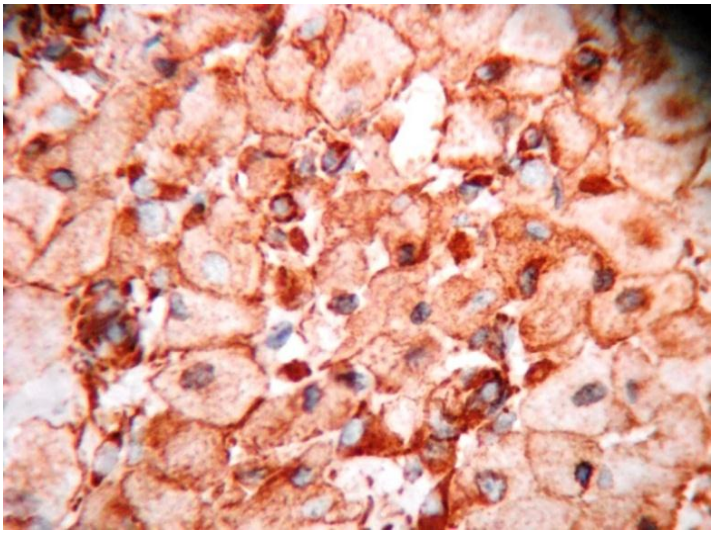

Figure 4: IHC staining for Vimentin showing diffuse cytoplasmic positivity (40X)

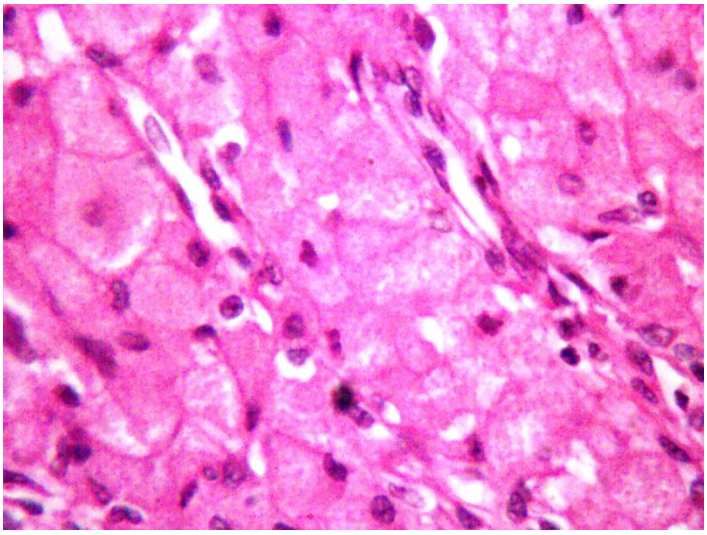

Figure 3: Clusters of large, round to polyhedral cells with abundant fine granular eosinophilic cytoplasm (H & E ,40X)

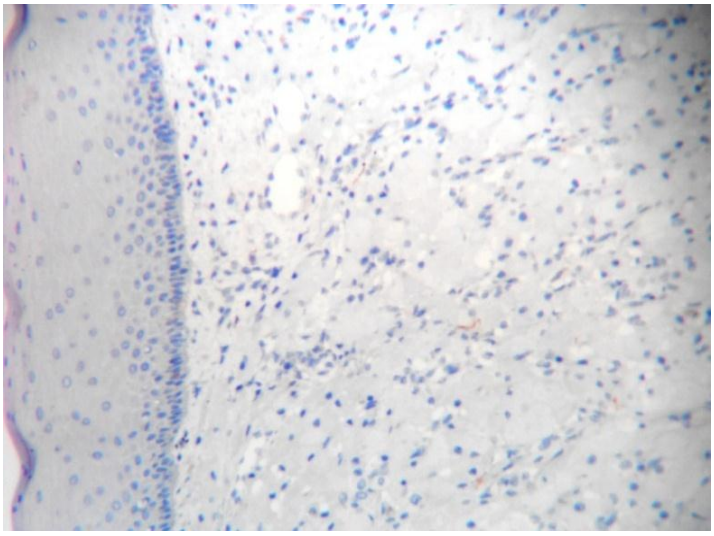

Figure 5: IHC staining for S-100 – Negative (10X)

III. Discussion

CGCT normally arises from the anterior part of the maxillary alveolar ridge of the newborn and frequently occurs lateral to the midline in the area of the developing primary lateral incisor and canine[1],[4]. Congenital granular cell tumour is seen three times more frequently on the anterior alveolar ridge of the maxilla than from the mandible with a female to male ratio of 8 to 10:1. Usually, it appears as a single lesion, but in 10% cases it may arise from multiple locations simultaneously. The condition is not associated with any other dental abnormalities or congenital malformations. Larger lesions may cause prenatal hydramnios due to ineffective deglutition and postnatally feeding and breathing difficulties.

CGCT resembles histologically the more common acquired/adult granular cell tumours (Myoblastoma) of any other site. Both have large cells with eosinophilic granular cytoplasm. The differences are enumerated in the table 1.

Despite two spontaneous regressions reported in the literature [12], surgery is the treatment of choice. Radical resection is not warranted as it is likely to damage the underlying alveolar bone and developing tooth buds. No recurrence has been reported even when the excision has been incomplete [13],[14].

Table I : Difference between CGCT and adult granular cell tumour (Granular cell myoblastoma)[1],[15],[16]

Sr. No./title	**Neumann's Tumour (Congenital Granular Cell Tumour)**	**Adult Granular Cell Tumour (Myoblastoma)**
1.Age of presentation	Just after Birth (Neonate)	Adulthood (30 to 60 years)
2.Site of involvement	Alveolar ridge of gingiva (Maxilla > Mandible)	Whole body, more than half cases are reported in oral cavity (maximum on tongue)
3. Gender	Female >> Male 8 to 10:1	No gender Predilection
4. Number	Rarely multiple	15% patients- multiple lesions, as many as 50 all over the body
5.HPE findings	No pseudoepitheliomatous hyperplasia, Less fibrous septae	Pseudoepithelio -aous hyperplasia seen, More Fibrous septae
6.Immunohisto -chemistry	**Vimentin- Positive S 100**– Negative	**S 100 – Positive NSE- Positive** [15]
7.Tissue of origin	Thought to be mesenchymal	Thought to be Schwannian.[1]

IV. Conclusions

CGCT has cells which are histopathologically identical to the more ubiquitous granular cell tumour (Myoblastoma) found in adults, but the early onset, unique location and pedunculated appearance differentiate the CGCT from adult granular cell tumour.

Our case provides additional evidence that CGCT derives from a mesenchymal origin, unlike the GCTof any other site.

REFERENCES

[1] Lapid O, Shaco-Levy R, Krieger Y, Kachko L, Sagi A. Congenital epulis. Pediatrics 2001;Feb107:e22

[2] Neumann E. Elin Fall von Congenitaler Epulis. Arch Heilk. 1871;12:189.

[3] Fister P, Volavsek M, Novosel Sever M, Jazbec J. A newborn baby with a tumour protruding from the mouth. Acta Dermatoven APA 2007;16:128-9.

[4] MB Diniz, MA Giro Elisa, CC Zuanon Angela, CA Costa, J Hebling,Congenital epulis: A rare benign tumour in the newborn. Journal of Indian Society of Pedodontics and Preventive Dentistry, Vol. 28, No. 3, July-September, 2010, pp. 230-233

[5] Kannan SK, Rajesh R, Congenital epulis - congenital granular cell lesion: a case report, J Indian Soc Pedod Prev Dent. 2006 Jun;24(2):104-6.

[6] Liviu Feller, Neil H Wood, Avin S Singh, Erich J Raubenheimer,Robin Meyerov and Johan Lemmer, Multiple congenital oral granular cell tumours in a newborn black female: a case report,Cases Journal 2008, 1:13 doi:10.1186/1757-1626-1-13

[7] Eghbalian F, Monsef A. Congenital epulis in the newborn, review of the literature and a case report. J Pediatr Hematol Oncol. 2009 Mar;31(3):198-9

[8] Khalid Khattala, Mohamed Rami, Aziz Elmadi, Leila Chbani, Youssef Bouabdallah. A case report of a huge congenital granular cell epulis operated under local anesthesia,The Pan African Medical Journal. 2012;12:105

[9] David Bosanquet and Graham Roblin ,Congenital Epulis: A Case Report and Estimation of Incidence, Int J Otolaryngol. 2009; 2009: 508780.

[10] 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 RadiolEndod. 1997 Dec;84(6):668-71.

[11] Parmigiani S, Giordano G, Fellegara G, Brevi B, Magnani C. A rare case of multiple congenital epulis. J Matern Fetal Neonatal Med. 2004 Nov;16 Suppl 2:55-8.

[12] Priyanshi Ritwik, Robert B Brannon and Robert J Musselman.,Spontaneous regression of congenital epulis: a case report and review of the literature , Journal of Medical Case Reports2010, 4:331

[13] Gokhale UA, Malhotra CJ. Congenital epulis of the newborn. Indian J Pathol Microbiol 2009;52:436-7

[14] Guilherme Costa Carvalho Silva, Tainah Couto Vieira , Janaina Couto Vieira, Carlos Roberto Martins, Edgard Carvalho Silva,CGCT: A lesion of Multidisciplinary interest, Med. Oral. Path. Cir Bucal 2007 Oct. 1:12(6), E 428-30

[15] Pietro Leocata,Giuseppina Bifaretti, Solidea Saltarelli, Alfonso Corbacelli, Luca Ventura, Congenital (granular cell) epulis of the newborn: a case report with immunohistochemical study on the histogenesis, Annals of Saudi Medicine, Vol 19, No 6, 1999

[16] Rosai and Ackerman's Surgical Pathology, Elsevier, Vol. 2, Pg. 2317-2318

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