Congenital granular cell tumor

in a neonate—a case report

Amar A. Shah^{*} and Anirudh V. Shah

Abstract

Background: Congenital granular cell tumor (CGCT) is a rare benign soft tissue tumor of the new born. It is also called congenital epulis.

Case presentation: We report the case of a 3-day-old male neonate who was postnatally noted to have a 2-cm mass over the maxillary alveolar ridge in the midline. The mass was causing feeding difficulties, and hence, decision for surgical excision was taken. The postoperative recovery was uneventful.

Conclusion: Congenital granular cell tumor (CGCT) is commonly seen on the gingival margins of the maxilla or mandible. Large tumors may cause polyhydramnios in the antenatal period or feeding and respiratory difficulties in the postnatal period. Diagnosis is usually confirmed by histopathology. Malignant change or recurrence has not been reported following incomplete excision of this mass.

Keywords: Congenital granular cell tumor, Neonate, Congenital epulis

Background

Congenital granular cell tumor (CGCT) is a rare benign soft tissue tumor of the newborn. The incidence is 0.0006% [1]. Congenital epulis of newborn is the universally accepted and frequently used term for this pathology [2]. It is also referred to as Neumann tumor after the German pathologist Ernst Christian Neumann who first described it in 1871 [3]. The lesions are known to vary in size from a few millimeters to almost 9 cm [4]. Larger lesions tend to cause feeding difficulty for the child, and surgical excision is indicated.

Case presentation

We report the case of a 3-day-old male neonate. The child was born at term and was postnatally noted to have a 2-cm mass over the maxillary alveolar ridge in the midline [Fig. 1]. The mass was not picked up on the antenatal scan. The mass was causing feeding difficulty to the

*Correspondence: shahamar22@gmail.com Amardeep Multispeciality Children Hospital & Research Centre, 65, Pritamnagar Society, Near Government Ladies Hostel, Near Gujarat

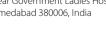
College, Ellisbridge, Ahmedabad 380006, India

child following which medical attention was sought. On examination, the mass arose from the maxillary gingiva and was pink, irregular, lobulated, and rubbery. The rest of the oral cavity was normal. Excision of the tumor was planned in view of the clinical condition and symptoms. Following endotracheal intubation, the base of the lesion was infiltrated with 0.5% lidocaine and 1:100,000 epinephrine [Fig. 2]. The alveolar mucosa around the base of the lesion was elevated, and the lesion was completely excised. The alveolar mucosa was apposed with interrupted polyglactin sutures. The child was commenced breastfeeding 12 h after surgery. This was gradually built up to full feeds over the course of the next 72 h, and the child was discharged home. Follow-up visits were scheduled at 1 week and 4 weeks. On follow-up, the child was thriving well, and the alveolar mucosa had also healed up nicely. Histopathology showed squamous mucosa with large polygonal cells, round nuclei, and abundant granular cytoplasm. No evidence of malignancy was noted. The overall picture was suggestive of congenital granular cell tumor (CGCT). Congenital granular cell tumor (CGCT) is a rare lesion and frequently seen on the alveolar ridges of the maxilla or mandible. It is predominantly a solitary



Open Access

Shah and Shah Annals of Pediatric Surgery (2022) 18:10 https://doi.org/10.1186/s43159-021-00145-0



© The Author(s) 2021. Open Access This article is licensed under a Creative Commons Attribution 4.0 International License, which permits use, sharing, adaptation, distribution and reproduction in any medium or format, as long as you give appropriate credit to the original author(s) and the source, provide a link to the Creative Commons licence, and indicate if changes were made. The images or other third party material in this article are included in the article's Creative Commons licence, unless indicated otherwise in a credit line to the material. If material is not included in the article's Creative Commons licence and your intended use is not permitted by statutory regulation or exceeds the permitted use, you will need to obtain permission directly from the copyright holder. To view a copy of this licence, visit http://creativecommons.org/licenses/by/4.0/.

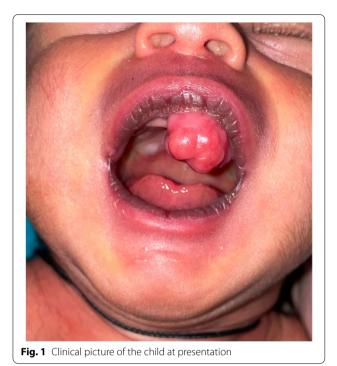




Fig. 2 Per operative picture showing the mass arising from the maxillary alveolar ridge

lesion and more prevalent in females and Caucasians [5–7]. It can be picked up during prenatal imaging as early as 26 weeks. It presents as an irregular, lobulated, rubbery soft tissue mass of varying sizes. A large size may be the reason for polyhydraminios affecting fetal deglutination. Postnatally, it may cause difficulty in feeding [1]. The mass is believed to originate from the undifferentiated mesenchymal cells, pericytes, fibroblasts, odontogenic epithelial, and smooth muscle cells. The diagnosis is usually clinical and confirmed by histopathology. Congenital

granular cell tumor needs to be differentiated from adult granular cell tumor. The adult variant occurs in 30–60 years. It is seen in the tongue and other sites. The overlying stratified squamous epithelium shows pseudoepitheliomatous hyperplasia [1]. The adult granular cell tumor shows positivity for S100, laminin, nerve growth factor receptor, and smooth muscle actin. CGCT shows positivity for only vimentin and neuron-specific enolase. Other differentials for congenital masses in the oral cavity can be dermoid cysts, vascular malformations, encephaloceles, and malignancies like teratoma and rhabdomyosarcomas. Melanotic pigmented neuroectodermal tumor is an osteolytic pigmented infiltrating neoplasm primarily affecting the jaws of the newborns.

Management of a child with CGCT remains debatable. The tumor stops to grow at birth and spontaneous regression can occur. These lesions can be monitored to see for regression. However, surgery is indicated when the mass causes impairment in feeding as was seen in the present case or respiratory compromise. For symptomatic patients, simple excision is preferred as radical excision can damage the developing tooth buds. Malignant change or recurrence has not been reported following incomplete excision of this mass [1].

Conclusion

Fewer than 250 cases of CGCT have been reported to date. Controversy still exists regarding the exact etiology, growth, and progression of CGCT in fetal life. Treatment is planned depending on the size of the lesion and the symptoms that it presents in the neonate.

Abbreviation

CGCT: Congenital granular cell tumor.

Acknowledgements

None

Authors' contributions

AAS: operating surgeon, literature review, interpretation of the data and drafted the manuscript. He is the corresponding author and will be accountable for all aspects of the work. AVS: conceptualized the report and critically revised the manuscript. Both authors approved the final version of the manuscript.

Funding

None

Availability of data and materials

The article is about a case report of a neonate who has been operated on at the hospital owned by the authors.

Declarations

Ethics approval and consent to participate

Ethics approval was waived as the presentation is from a private children's hospital owned by the authors. Also, the study is not an experimental study. This is an uncommon case of a congenital granular cell tumor in a neonate.

Consent for publication

The authors have obtained the necessary approval for the publication of the data from the hospital. The parents of the child have given written consent to operate the child and to share the photograph for academic purposes in the journal.

Competing interests

The authors declare that they have no competing interests.

Received: 6 September 2021 Accepted: 3 November 2021 Published online: 04 February 2022

References

- 1. Patil RN, Helwatkar S, Raut W. Congenital granular cell epulis: a case report. J Med Sci Health. 2017;3(1):41–4.
- Ruschel HC, Beilke LP, Beilke RP, Kramer PF. Congenital epulis of newborn: report of a spontaneous regression case. J Clin Pediatr Dent. 2008;33:167–9.

- 3. Neumann E. Ein fall von congenitaler epulis. Arch Heilk. 1871;12:189–90.
- 4. Gardner P, Rozzelle A. Congenital granular cell tumor: case report and review. Case Rep Pediatr. 2018;2018:3.
- 5. Dash J, Sahoo P, Das S. Congenital granular cell lesion "congenital epulis": report of a case. J Indian Soc Pedod Prev Dent. 2004;22:63–7.
- Lack E, Worsham G, Callihan M, Crawford B, Vawter G. Gingival granula cell tumors of the newborn (congenital "epulis"): a clinical and pathologic study of 21 patients. Am J Surg Pathol. 1981;5(1):37–46.
- Thoma V, Idrissi B, Kohler M, Becmeur F, Viville B, Favre R. Prenatal diagnosis of congenital epulis a case study. Fetal Diagn Ther. 2006;21(4):321–5.

Publisher's Note

Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

Submit your manuscript to a SpringerOpen[™] journal and benefit from:

- Convenient online submission
- ► Rigorous peer review
- Open access: articles freely available online
- ► High visibility within the field
- ▶ Retaining the copyright to your article

Submit your next manuscript at > springeropen.com