

Case report:**Congenital epulis (congenital gingival granular cell tumor)**

Mohammed S. Saeed*, Moutaz Al Ani**

*Department of Pathology, College of Medicine, University of Mosul;

**Department of Pediatric Surgery, Al Khansa'a Maternity Teaching Hospital.

(Ann. Coll. Med. Mosul 2010; 36 (1 & 2): 153-156).

Received: 17th Mar 2010; Accepted: 7th Jul 2010.**ABSTRACT**

Congenital epulis or congenital gingival granular cell tumor is a rare lesion of newborn, also known as Neuman's tumor. Congenital epulis mostly occurs as a single mass but rarely as multiple. It arises from the mucosa of gingiva either from maxillary or mandibular alveolar ridge. The exact histogenesis of congenital epulis is still uncertain, but the biologic behavior of this lesion is compatible with embryonic hamartoma, so that surgical excision is the treatment of choice. The presented case describes a mass in the anterior region of maxillary alveolar ridge in a 15 day old healthy female, which causes a feeding problem and hinders normal closure of the mouth. Tumor was excised surgically under general anesthesia. The excised material was sent for histopathological examination and the diagnosis of congenital epulis was performed.

Keywords: Congenital epulis, congenital gingival granular cell tumor, newborn.

الخلاصة

الورم اللثوي الخلقي أو ورم اللثة للخلية المحببة الخلوية هو ورم نادر الحدوث عند الأطفال حديثي الولادة، كما يعرف أيضا بورم نيومان. الورم اللثوي الخلقي على الأغلب يظهر ككتلة أحادية ولكن نادرا متعدد الكتل. هذا الورم ينشأ من الغشاء المخاطي للثة عند الحافة السنمية لكل من الفك الفمقي أو الفك الأسفل. المنشأ الأكيد لورم اللثوي الخلقي غير معروف لحد الآن ولكن التصرف الإحيائي للورم يؤكد كونه ورم خلقي وعملية استئصاله جراحيا هي الطريقة المثالية لعلاج الحالة المقدمة توصف ورم في المنطقة الأمامية من الحافة السنمية للفك الفمقي في طفلة حديثة الولادة تبلغ من العمر ١٥ يوم. الورم أحدث مشاكل في الرضاعة ويعيق غلق الفم. الورم استأصل جراحيا تحت التخدير العام. أرسلت العينة إلى الفحص النسيجي المرضي حيث تم تشخيصه كورم لثوي خلقي.

الكلمات المفتاحية: ورم لثوي خلقي، ورم للخلية المحببة الخلوية، حديثي الولادة.

Congenital epulis (CE) is a rare lesion of the gingiva of the newborn, also known as congenital gingival granular cell tumor or granular cell tumor of newborn. Neumann first described congenital epulis in 1871, hence also known as Neumann's tumor ⁽¹⁾. Congenital epulis occurs as a well-defined, pedunculated pink mass, with a smooth or lobulated surface of various size from few mm to 9 cm. It commonly arises from the anterior

maxillary or mandibular alveolar ridge of the newborn in a ratio of 3:1, and usually not associated with any other congenital malformations. Congenital epulis has a female predilection, with an 8:1 ratio; an endogenous (intrauterine) hormonal stimulus is proposed, but has been disproved because of the absence of a receptor for estrogen and progesterone ⁽¹⁻⁴⁾.

Case report

A newborn girl was referred to the pediatric surgery center at Al-Khansa'a Maternity Teaching Hospital because of a mass protruding from her mouth. The infant was born on the 38th week of gestation. Pregnancy and parturition were normal. The birth weight was 3400 gm. At birth a firm, non-ulcerated pedunculated tissue mass was found protruding from the mouth. The mass was attached to the gingiva by a stalk measuring 2 X 1 cm. The mass hindered normal closure of the mouth and breast feeding, but did not cause airway obstruction or respiratory distress. On physical examination the neonate was pink and active. There was neither cervical lymphadenopathy, nor other congenital abnormalities. There was no family history of similar lesion, (Fig. 1).



Figure (1): Pedunculated mass from the anterior region of maxillary alveolar ridge.

The infant was operated on under general anesthesia with mask. Surgery was performed using electric cauterization at 20 watt. Blood loss was insignificant. The area of the resection was left open for closure by secondary intention. The operative and postoperative course was normal. Oral feeding was started on the first postoperative day and the child was discharged on following day. Healing was uneventful and the gingiva re-epithelialized within 10 days.

Macroscopically: A firm whitish-gray smooth surface mass. The cut surface of the specimen showed a homogenous, whitish-gray tissue, which on histopathological examination showed a highly vascularized growth covered with mildly acanthotic squamous epithelium and the underlying connective tissue showing highly vascularized stroma with closely packed polygonal cells, with centrally placed uniform nuclei and a markedly granular cytoplasm (Fig. 2).

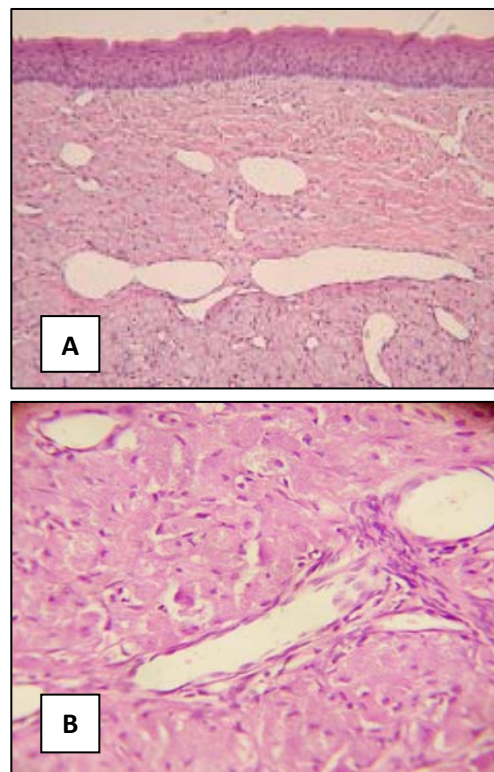


Figure (2): A-The vascularized stroma shows a benign proliferation of round cells, with abundant eosinophilic granular cytoplasm and small nuclei. B-Round granular cells with eosinophilic cytoplasm, with small uniform basophilic nuclei.

Discussion

Congenital epulis or granular cell tumor of newborn is a rare lesion of the gingiva seen only in newborn. Since 1871 after the first published case until 2002, 216 cases of congenital epulis have been reported from the literature⁽⁵⁾. Epulis is most frequently located on the anterior maxillary alveolar ridges as a single mass although in 10% of cases occur as multiple lesions. CE clinically appears as a pedunculated protuberant mass, which may interfere with respiration or feeding. In cases with large lesions mechanical oral and nasal obstruction can impair fetal deglutition and neonatal respiratory efforts resulting in polyhydramnios prenatally or respiratory impairment postnatally⁽⁶⁻⁸⁾. The presented case was 2 week age female with pedunculated single maxillary alveolar ridge mass of 2 x 1 cm. In our case there are no other congenital abnormalities present. CE has an 8:1 female predilection and 3:1 maxillary alveolar site predilection^(2,8). Our case is a female child and lesion occurred in anterior maxillary gum pad. Like our case CE usually occurs as a single mass although 10% cases occur as multiple^(2,8). The size of the CE varies from few mm to 9 cm⁽⁸⁾. Large lesion can interfere with fetal deglutition resulting in hydramnios, respiratory obstruction and difficulty in feeding postnatally⁽²⁾. As the size of the lesion in our case is two-centimeters in diameter, it did interfere with feeding and breathing but fortunately she has no respiratory infection at the time of presentation which might occur secondary to aspiration.

There is a striking histologic similarity of granular cells of congenital gingival granular cell tumors (GGCTs) and the far more common granular cell tumor (GCT) of any other site. There are several distinguishing features of GGCT, such as predilection for newborn females, anterior maxillary location, presence at birth, plexiform arrangement of thin walled blood vessels, and lack of pseudoepitheliomatous hyperplasia^(1,9).

The exact histogenesis of CE is still uncertain. The various proposed cells of origin are of the odontogenic epithelium, undifferentiated mesenchymal cells, pericytes,

fibroblasts, smooth muscle cells, nerve related cells, and histiocytes⁽¹⁰⁾. However, most authors suggest a mesenchymal origin⁽¹¹⁾.

Immunohistochemical stains gave the following results: the tumor cells were diffusely and strongly positive for vimentin, and negative for S100-protein, actin, desmin, laminin, keratin, estrogen, and progesterone receptors⁽¹⁾. Electron microscopic examination showed granular cells containing heterogeneous electron dense granules, lysosomes, and cytoplasmic lipid droplets. The cells had irregular cytoplasmic borders with small extensions. There was no basement membrane associated with the granular cells. There was no evidence of any epithelial differentiation and no evidence of schwannian differentiation as well⁽¹⁾.

Based on the above findings, in addition to the absence of local recurrence of the mass even in incomplete excision, the possibility of spontaneous regression, and the lack of a malignant counterpart, it is concluded that the biologic behavior of congenital epulis is compatible with embryonic hamartoma^(10,12,13).

This tumor is often misdiagnosed before surgery because of its rarity. The differential diagnosis of a large mass in the fetal or neonatal oral cavity should include such congenital malformations as encephalocele, dermoid cysts or teratoma and benign and malignant neoplasms including hemangioma, lymphatic malformations, rhabdomyoma, fibroma, melanotic or pigmented neuroectodermal tumors of infancy^(1,14). The differential diagnoses in this case were hemangioma, teratoma or rhabdomyoma.

Despite two spontaneous regressions reported in the literature⁽¹⁵⁾, surgery is the only possible treatment for these tumors. Surgery should not be radical; it minimizes the danger of damaging underlying alveolar bone and developing tooth buds. No recurrence has been reported despite an incomplete resection⁽⁸⁾.

Conclusion

Congenital epulis is a rare lesion of gingiva of the newborn, appears usually as a single pedunculated firm red smooth or lobulated mass on the maxillary alveolar ridge, mostly in

female. The exact histogenesis of congenital epulis is still uncertain. However; recently most authors suggest a mesenchymal origin and they conclude that biologic behavior of the congenital epulis is compatible with embryonic hamartoma. Lastly surgical excision is the treatment of choice for this lesion.

References

1. Lapid O, Shaco-Levy R, Kringer Y, et al. Congenital Epulis. *Pediatr* 2001; 107 (2): 22-24.
2. Koch BL, Myeriii C, and Egelhoff JC. Congenital Epulis. *AJNR* 1997; 18: 739-41.
3. Leocata P, Bifaretti G, Saltarelli S, et al. Congenital (Granular Cell) Epulis of the newborn: A case report with immunohistochemical study on the histogenesis. *Ann Saudi Med* 1999; 19: 527-9.
4. Salvia GC, Vieira TC, Vieira JC, et al. Congenital granular cell tumor (congenital epulis): A lesion of multidisciplinary interest. *Med Oral Pathol Oral Cir Bucal* 2007; 12(6): 428-30.
5. Reinshagen K, Wessel LM, Roth H, et al. Congenital epulis: a rare diagnosis in pediatric surgery. *Eur J Pediatr Surg*. 2002 Apr; 12(2): 124-6.
6. Eghbalian F, and 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.
7. Koch BL, Myeriii C, and Egelhoff JC. Congenital Epulis. *AJNR* 1997; 18: 739-41.
8. Wittebole A, Bayet B, Veyckemans F, Gosseye S, Vanwijck R. Congenital Epulis of newborn. *Acta Chir Belg* 2003; 103: 235-7.
9. Kannan SK and Rajesh R. Congenital epulis-congenital granular cell lesion: A case report. *J Indian Soc Pedod Prev Dent* 2006; 24(2): 104-106.
10. Sahu S, Maurya RK, Agarwal A, et al. Multiple congenital epulis in newborn - A rare presentation. *JOMFP* 2009; 13(2): 78-80.
11. Kupers AM, Andriessen P, Kempen MJ et al. Congenital epulis of the jaw: a series of five cases and review of literature. *Pediatr Surg Int* 2009; 25: 207-10.
12. Dash JK, Sahoo PK, and Das SN. Congenital Granular Cell Lesion "Congenital Epulis"- Report of a case. *J Indian Soc Pedo Prev Dent* 2004; 22(2): 63-67.
13. Mabongo M, Wood NH, Lemmer J, et al. Congenital epulis. A case report. *SADJ* 2008; 63(6): 350-1.
14. Sándor GK, McGuire TP, Gomes PP, et al. Congenital Epulis: A Surprise in the Neonate. *J Can Dent Assoc* 2006; 72(8): 747-50.
15. Jenkins HR and Hill CM. Spontaneous regression of congenital epulis of the newborn. *Arch Dis Child* 1989, 64 : 145-147.