HK J Paediatr (new series) 2010;**15**:165-169

Congenital Granular Cell Tumour in a Newborn: A Case Report and Literature Review in China

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Abstract Congenital granular cell tumour (CGCT) is a rare benign intraoral tumour found exclusively in the newborns. Over 200 cases have been reported from all over the world. Although CGCT in the newborns has been well reported in the literature, very few cases have been reported in China. In this paper, we report a case of congenital granular cell tumour seen on the anterior mandibular alveolus. The patient was a 2-day-old Chinese girl who presented with a pedunculated 3.5 cm-diameter round soft tissue gingival mass protruding from her mouth. We describe the prenatal diagnosis, clinical and histological characteristics of the lesion, and management.

Key words Congenital epulis; Congenital granular cell tumour; Newborn; Prenatal diagnosis; Treatment

Congenital granular cell tumour (CGCT) is a rare benign tumour of the newborn and is also known as congenital epulis (CE) or Neumann's tumour. The most frequent location of the lesions is the gingiva of the anterior alveolar ridge of the jaws. These lesions are seen in the maxillary alveolus three times more often than in the mandibular alveolus. Females are more frequently affected than males (8-10:1) with a Caucasian predisposition. This tumour is typically seen as a mass protruding out of the newborn's mouth, which may interfere with breathing or feeding. The recommended treatment is prompt surgical resection. These lesions are benign and no recurrence or metastatic lesions have been reported.[1]

Although CGCT in the newborns has been well reported in the literature, very few cases have been reported in China.[2-11] We describe a new case occurring in China with an accompanying literature review.

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Received January 28, 2010

Case Report

A 39-year-old pregnant woman in the 32nd week of gestation came to the Stomatology Department of Children's Hospital of Zhejiang University for consultation and advice. Prenatal ultrasonography showed that her foetus had a hyperechoic mass (volume: 2.3 × 1.9 × 2.4 cm) with marked increased blood flow in the oral cavity and it moved synchronously with the movements of the head (Figure 1). The mass did not interfere with foetal swallowing or mobility of the foetal tongue. No facial abnormalities were identified and the amniotic fluid was normal. There was no family history of hereditary diseases. Our presumptive diagnosis was a congenital teratoma. We suggested that the pregnant woman should be followed up with ultrasound scan periodically, but she did not follow our advice. The woman had an uneventful delivery by caesarean section at 38 weeks gestation. The newborn girl had normal apgar score. The birth weight was 3350 g and the length was 52 cm.

The girl was then referred to our department on the second day after birth. Clinical examination revealed a pedunculated, well defined 3.5 cm-diameter round soft erythematous tissue mass which was superficially ulcerated. It was located to the right side of the midline and was found to be attached to the gingival of the anterior alveolar ridge of the mandible by a thin pedicle 1 cm in diameter. The large mass prevented normal closure of the mouth, displaced the lower lip and hindered breast or bottle-feeding, but did not cause airway obstruction or respiratory distress (Figure 2a). On further examination, there was a cyst near the mass, on the anterior mandibular ridge, 1.5 cm × 0.5 cm in size (Figure 2b). No other facial or general abnormalities were found. The differential diagnosis included CGCT, odontogenic tumour, teratoma, haemangioma and fibroma with a provisional diagnosis of CGCT. Physical examination and laboratory tests were otherwise normal.

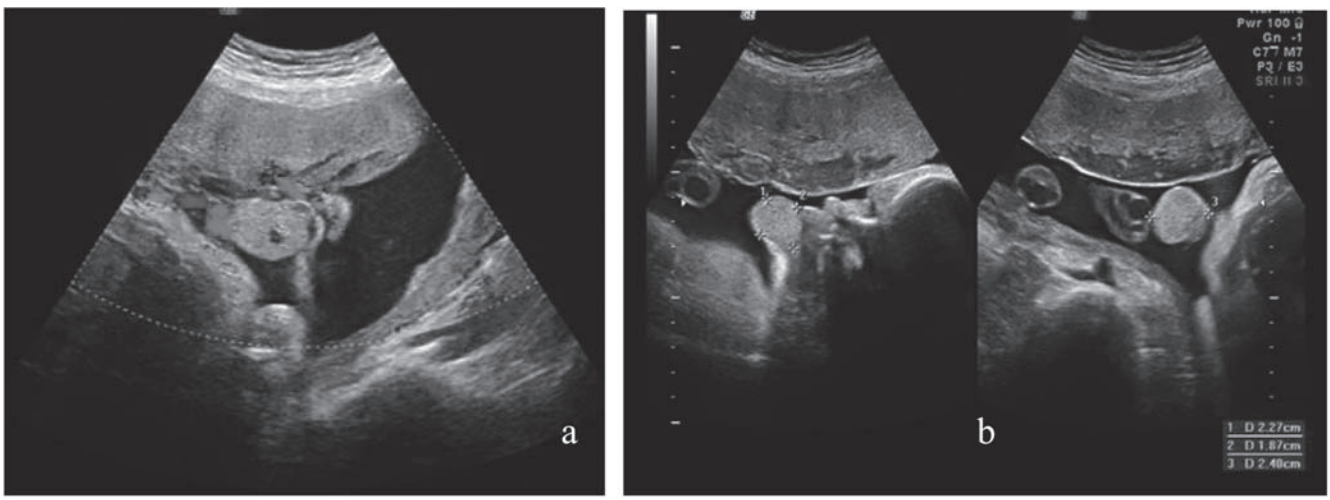

Figure 1 (a) An ultrasound scan performed in the 32nd week of gestation showed a well-defined, smooth, round and hyperechoic mass with marked blood flow in the oral cavity. The mass did not interfere with the swallowing, or the foetal tongue's mobility. (b) Size of the mass 2.3 x 1.9 x 2.4 cm.

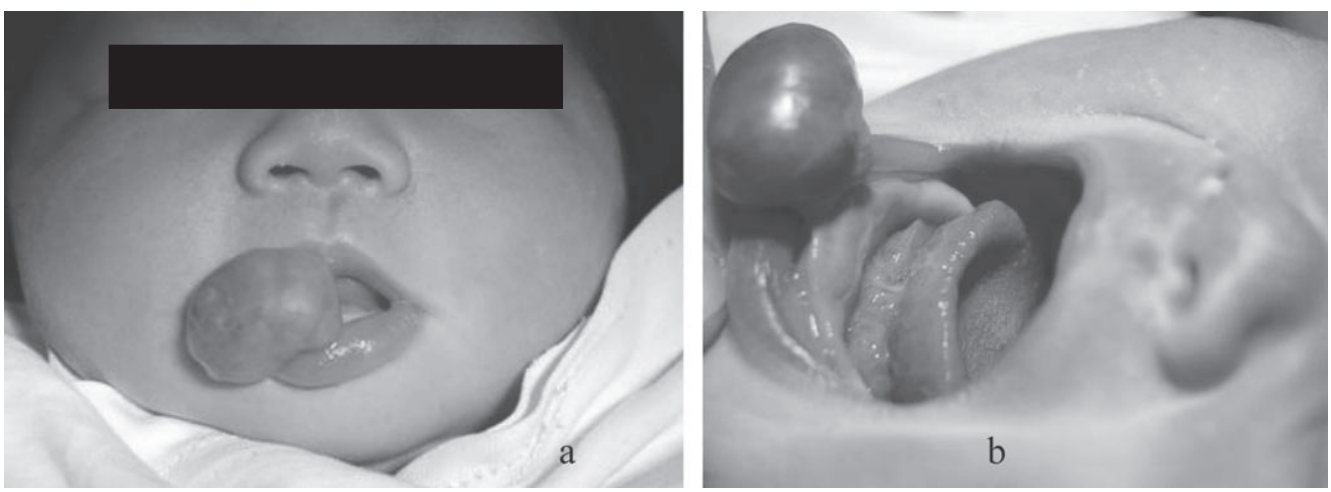

Figure 2 (a) Oral examination, A pedunculated 3.5 cm-diameter round soft tissue gingival mass was noted. It was well defined and exhibited a smooth erythematous and slighty superficially ulcerated surface. The mass protruded from her mouth and prevented normal closure of the mouth and displaced the lower lip. (b) The mass was attached to the right anterior gingival of the mandibular by a thin pedicle measuring 1 cm. The examination of the anterior lower gum also showed a cyst measuring 1.5 cm x 0.5 cm near the mass.

The lesion was completely excised under local anesthesia in the outpatient department when the baby was 3 days old. One month after surgery, healing was uneventful and the anterior alveolar cyst disappeared (Figure 3). No recurrence occurred at 6 months follow-up after surgery.

The mass was sent for histological examination. Histology of the specimen revealed diffuse sheets and clusters of polygonal cells containing small round to oval nuclei and abundant coarsely granular cytoplasm. There was a delicate plexiform network of capillaries (Figure 4). The diagnosis of CGCT was confirmed.

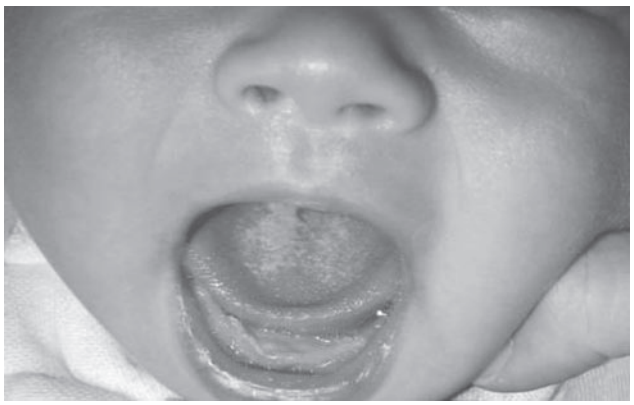

Figure 3 At 1 month after surgical resection, healing was uneventful and the anterior alveolar cyst had disappeared.

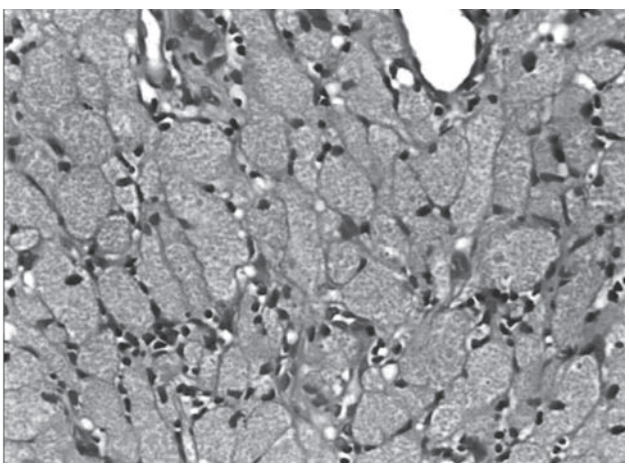

Figure 4 Histology examination of the specimen revealed diffuse sheets and clusters of polygonal cells containing small round to oval nuclei and abundant coarsely granular cytoplasm (H&E stain x 200).

Discussion

CGCT, first described by Newmann in 1871, is a rare benign intraoral tumour exclusively found in the newborns. Over 200 cases have been reported from all over the world.[12] To our knowledge, only 14 cases have been reported in China (Table 1).[2-11] There are some characteristics in Chinese cases which differed from other published cases in the English literature (Table 1). The tumour occurs more commonly in females and there were 11 females and 4 males among the 15 Chinese cases. With the exception of two cases, all were solitary lesions with 7 lesions located in the maxilla and 6 in the mandible. No lesions was reported to occur on the tongue in China. The size varies from 4 millimeters to 4 centimeters in diameter. In the present case, the mass measured 3.5 cm in diameter. It interfered with bottle-feeding or breast-feeding of the baby, but did not interfere with her normal breathing. Although CGCT are not often associated with other congenital abnormalities, they have been described to be associated with neurofibromatosis, polydactyly, midface hypoplasia, and absent underlying teeth.[13] A case associated with bilateral transverse facial cleft had been reported in China.[7] In our case, a cyst was found on the anterior lower gum near the mass, and the cyst spontaneously disappeared one month after operation, suggesting that this cyst could have been associated with the mass.

Although the histogenesis of CGCT is not certain, it is thought to be a non-neoplastic, degenerative or reactive lesion.[1] The distinct visual presentation usually allows for clinical recognition at birth. When the lesion is large and interferes with feeding and breathing, prompt surgical excision under either local or general anesthesia is the optimal management. Complete surgical excision is curative. Recurrence or malignant transformation or damage to future dentition following incomplete excision have not been reported. Wide surgical excision is not necessary.[1,12] Spontaneous regression of some very small lesions (the diameter less than 1.5 cm) had been reported within the first year of life.[14] In our case, the lesion interfered with the feeding and the patient underwent the surgical excision under local anesthesia uneventfully. Follow-up six months after surgery showed complete healing with no evidence of recurrence.

As there is a potential risk of neonatal respiratory distress, prenatal diagnosis is vital for a successful pregnancy outcome. With the development of prenatal ultrasound scanning, the diagnosis of orofacial tumours of the foetus can be made in utero. Most of Chinese cases were not

Table 1 Reported cases of the congenital granular cell tumour in China

Author	Sex	Pattern of occurrence	Location of lesion	Size of tumour	With or without other abnormality	Prenatal diagnosis
Lu et al (2007)[2]	F	Solitary	Mandibular	2.5 x 2 x 2 cm	Without	33 weeks of pregnancy
Zhang et al (2006)[3]	F	Multiple	Mandibular and maxillary	3 x 3 x 3 cm and 1.5 x 1.5 x 1.5 cm	Without	None
	F	Solitary	Maxillary	1.5 x 1.2 x 1.2 cm	Without	None
Dong et al (2002)[4]	F	Solitary	Maxillary	1.5 x 1.2 x 1.0 cm	Without	None
Wang et al (2000)[5]	F	Solitary	Maxillary	0.8 x 0.7 x 0.5 cm	Without	None
Zhang (1998)[6]	F	Solitary	Mandibular	2.8 x 1.7 x 1.4 cm	Without	None
Qu et al (1995)[7]	M	Solitary	Maxillary	2.0 x 1.5cm	With bilateral transverse facial cleft	None
Yu et al (1991)[8]	F	Solitary	Maxillary	1.4 x 0.7 x 0.6 cm	Without	None
Sun (1989)[9]	M	Multiple	Mandibular and maxillary	Like cherry	Without	None
Chen et al (1987)[10]	F	Solitary	Mandibular	4 x 4 cm	Without	None
Liu et al (1984)[11]	M	Solitary	Maxillary	1.5 x 1 x 1 cm	Without	None
	M	Solitary	Mandibular	1 x 0.7 x 0.5 cm	Without	None
	F	Solitary	Mandibular	1 x 0.7 x 0.6 cm	Without	None
	F	Solitary	Maxillary	1.1 x 0.7 x 0.5 cm	Without	None
Present case	F	Solitary	Mandibular	3.5 x 3.5 x 3.5 cm	With a cyst	32 weeks of pregnancy

diagnosed prenatally.[3-11] In our case, prenatal ultrasound scanning of the face of the foetus clearly showed an echogenic mass with marked increased blood flow in the oropharyngeal area. Three-dimensional ultrasound scanning can exactly show the location and size of the mass.[15] The earliest diagnosis was made at 26 weeks gestation.[16] The diagnosis for our patient was made at 32 weeks of pregnancy. For most of the reported patients, prenatal diagnosis was made at about 30~32 weeks of pregnancy, from the end of the second and throughout the third trimester.[17] Tumour size determines the possible interference as well as the treatment. During the prenatal period, the tumour can lead to polyhydramnios by interfering with foetal swallowing. It may interfere with breathing at birth. Although a prenatal ultrasound scan allows a more precise diagnosis, the definitive confirmation remains histological. Prenatal diagnosis is important for the organisation of a multidisciplinary approach to management involving obstetricians, neonatologists, paediatric surgeries and paediatric dentists.[18]

References

1. McGuire TP, Gomes PP, Freilich MM, Sándor GK. Congential Epulis: A Surprise in the Neonate. J Can Dent Assoc 2006;72: 747-50.
2. Lu W, Hu JG, Yu L, Xing FZ, Huang XY. Congenital granular cell epulis in the newborn. Chin J Pediatr Surg (Chin) 2007;28: 335-6.
3. Zhang YZ, Yang F, Li HM, Chen L. Congenital Epulis: 2 cases report. J Clin Exp Pathol (Chin) 2006;22:125.
4. Dong HM, Li MJ, Deng MH. A case of Congenital Epulis. J Shantou University Medical College (Chin) 2002;15:253.
5. Wang YX, Cao MY, Wei SY. Congenital Epulis: 1 case report. J Comprehensive Stomatol (Chin) 2000;16:60.
6. Zhang XD. A case of congenital epulis. Chin J Pathol (Chin) 1998;27:152.

7. Qu PL, Zhang WX, Han BL, Bing HB. Congenital bilateral transverse facial cleft associated with congenital epulis: a case report. J Stomatol (Chin) 1995;15:182.
8. Yu L, Xiao S. A case of congenital epulis. J First Mil Med Univ (Chin) 1991;11:238.
9. Sun MH. A case of congenital epulis in newborn. J Modern Stomatol (Chin) 1989;3:119.
10. Chen LL, Deng QC. A case of congenital epulis in mandibular. J Pract Stomatol (Chin) 1987;3:16.
11. Liu Z, Liu AR, Gao Y. Congenital epulis. J Stomatol (Chin) 1984; 4:203-4.
12. Reinshagen K, Wessel LM, Roth H, Waag KL. Congenital epulis: a rare diagnosis in paediatric surgery. Eur J Pediatr Surg 2002; 12:124-6.
13. Koch BL, Myer C 3rd, Egelhoff JC. Congenital epulis. Am J Neuroradiol 1997;18:739-41.
14. Sakai VT, Oliveira TM, Silva TC, Moretti AB, Santos CF, Machado MA. Complete spontaneous regression of congenital epulis in a baby by 8 months of age. Int J Paediatr Dent 2007; 17:309-12.
15. Shaw L, Al-Malt A, Carlan SJ, Plumley D, Greenbaum L, Kosko J. Congenital epulis: three-dimensional ultrasonographic findings and clinical implications. J Ultrasound Med 2004;23:1121-4.
16. Nakata M, Anno K, Matsumori LT, et al. Prenatal diagnosis of congenital epulis: a case report. Ultrasound Obstet Gynecol 2002; 20: 627-9.
17. Kusukawa J, Kuhara S, Koga C, Inoue T. Congenital granular cell tumour (congenital epulis) in the fetus: a case report. J Oral Maxillofac Surg 1997;55:1356-9.
18. Thoma V, Idrissi B, Kohler M, Becmeur F, Viville B, Favre R. Prenatal diagnosis of congenital epulis: a case study. Fetal Diagn Ther 2006;21:321-5.