CASE REPORT

Congenital granular cell lesion in newborn mandible

O Gupta, R Arora, K Gupta, U Sharma

Department of Pathology, VMMC and Safdarjung Hospital New Delhi, India

Abstract

Congenital granular cell lesion (CGCL) is a rare non-neoplastic lesion found in newborns also known as Neumann's tumor. This benign lesion occurs predominantly in females mostly as a single mass. The histogenesis and natural history of the lesion remains obscure. It arises from the mucosa of the gingiva, either from the maxillary or mandibular alveolar ridge. The lesion is more common in the maxillary alveolar ridge than the mandibular.The present report describes a case of congenital granular cell lesion in an eight-day-old female child who was born with a mass on the anterior mandibular alveolar ridge. The mass was protruding from her mouth and compromised feeding. A clinical diagnosis of teratoma was suggested. Histologically, cells of this lesion are identical to granular cell tumor (neuroectodermal type) and show intense diastase-resistant Periodic Acid Schiff positivity. Immunohistochemically, cells are positive for vimentin but negative for S-100 and desmin, thus suggesting that CGCL is possibly derived from primitive gingival mesenchymal cells rather than having schwannian origin.

Key words: Congenital epulis, congenital granular cell lesion, immunohistochemistry

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Introduction

Congenital granular cell lesion (CGCL) or 'congenital epulis' is a rare lesion found in newborns also known as Neumann's tumor; the word "Epulis" is derived from a Greek word and means "on the gum" or gum boil.[1] Since its first description by Neumann in 1871, around 200 cases have been reported in the worldwide literature so far.[2] The tumor is benign in nature, mostly occurs as a single tumor but rarely as multiple and has a close resemblance to granular cell myoblastoma.The appearance of the lesion is more common in the maxillary alveolar ridge than in the mandibular, with a female predominance.[3] It is an intriguing lesion with unclear etiology, histiogenesis and natural history. Several theories have been proposed to explain its histiogenesis, such as myoblastic,[4] odontogenic,[5] neurogenic, histiocytic and endocrinologic origin.[6]

The definitive histogenesis still remains controversial in spite of additional information provided by electron microscopic and immunohistochemical studies. The other theories of origin include epithelial,[7] undifferentiated mesenchymal cell,[8] pericytes,[9] fibroblastic,[10] smooth muscles,[11] nerve-related cells[12] and myofibroblasts.[1,4]

Case Report

An eight-day-old Indian female child, born at 40 weeks' of gestation weighing 3 kg, was brought by her mother for evaluation of a mass protruding from her mouth, which was causing feeding difficulties, but no airway obstruction. The neonate was born by normal vaginal delivery, with uneventful prenatal and perinatal course, and was otherwise healthy. Intraoral examination revealed a bilobed and pedunculated tumoral mass, with color similar to the adjacent mucosa, arising from the anterior mandibular alveolar ridge. Measuring approximately 2 cm, it had a smooth and firm surface and was protruding slightly from the mouth. A clinical diagnosis of teratoma was suggested, the mass was surgically excised under local anaesthesia using

Address for correspondence:
Dr. Oneal Gupata,
Department of Pathology, VMMC and Safdarjung Hospital,
Sector 111-A, H.NO>23, Ground Floor Rachna Vaishali
Ghaziabad - 201 010, India.
E-mail: oneal80@gmail.com

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2% Xylocaine. The blood loss was minimal and the specimen was fixed in 10% neutral buffered formalin and processed for histopathological examination. The patient was followed for 6 months postoperatively, the period remained uneventful. No recurrence was seen.

Pathological Examination

The excised mass was bilobed, oval, grayish white and firm in consistency measuring 2 × 2 × 1 cm. The external surface was smooth. The cut surface was well-circumscribed, grayish white, and had focal hemorrhagic areas. Hemotoxylin and eosin-stained sections revealed a well-circumscribed mass lying beneath an unremarkable stratified squamous epithelium. There were sheets of monotonous round to polygonal cells with central to eccentrically placed nucleus with condensed chromatin and inconspicuous nucleoli. The cells had abundant granular and eosinophilic cytoplasm. A delicate network of fine capillaries traversed the lesion [Figure 1a]. Nests of entrapped odontogenic epithelium were also seen (Figure 1a [inset]). There was no necrosis or mitosis. The cells were Periodic Acid Schiff (PAS)-positive and diastase-resistant [Figure1b].

The following antibodies were employed: Vimentin (clone: VIM 3B4, Mouse monoclonal, Dako), desmin (clone: [D33] Mouse Monoclonal,Isotype: IgG1, kappa., dako), S-100 (clone: Mouse Monoclonal, Anti-S-100 Protein, Millipore) [Table 1]. Immunohistochemical stains revealed that cells were diffusely and strongly positive for vimentin [Figure 2a] and were negative for S-100 protein [Figure 2b] and desmin.

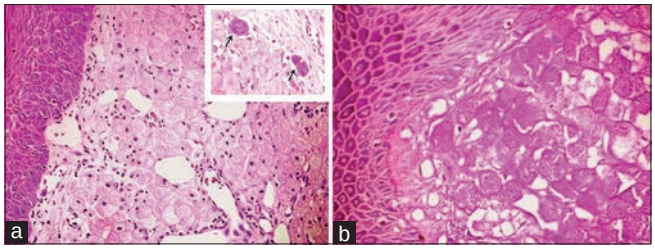

Figure 1: (a) Photomicrograph showing a benign proliferation of round to polygonal cells, with abundant eosinophilic granular cytoplasm and small nuclei [Inset: Entrapped odontogenic epithelium (arrow)] 400×; H and E. (b) Photomicrograph showing intense PAS-positive and diastase-resistant cells (400×)

Discussion

Congenital granular cell lesion (CGCL) or "congenital epulis" occurs typically in newborns. It arises mostly from mucosa over the alveolar ridge, thus interfering with mouth closure and feeding. Most of the cases appear as a solitary lesion occurring predominantly on the maxillary alveolar ridge, the maxillary to mandibular ratio being 3:1, the canine incisor region is most frequently affected. Females are affected more frequently than males with a ratio of 8:1. The cytogenesis and hormonal receptor studies give no clues for female predominance; because of the absence of detectable estrogen and progesterone receptors.[5] The frequent maxillary occurrence in the canine and incisor region can be attributed to the fact that the maxillary anterior region is a common site for supernumerary teeth. In the present report, the lesion was seen in the mandibular incisor region, which is a rare occurrence. When multiple, the tumor may cause respiratory obstruction and prenatal hydramnios due to ineffective deglutition.[3,6] The tumor is often misdiagnosed prior to surgery because of its rarity and lack of awareness, as in our case.

The clinical differentials include teratoma, hemangioma, rhabdomyoma, schwanomma, hibernoma and fibroma.[1,13] The presence of epithelial islands commonly found in the gum pad of newborns and infants, suggests its origin from odontogenic epithelial rests.[5] Congenital epulis strikingly resembles adult granulose cell tumor (myoblastoma) on light microscopy. However, this neonatal counterpart does not have schwannian origin. This is reflected by its S-100 negativity.[3] The CGCL are vimentin-positive. Further, they

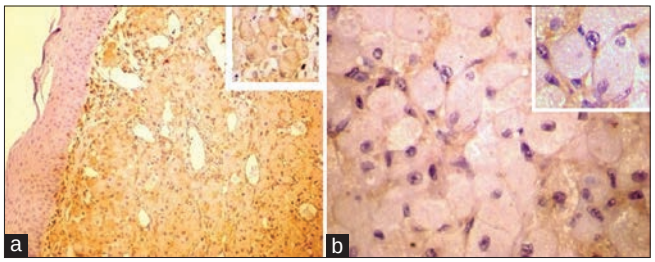

Figure 2: (a) Photomicrograph showing immunoreactiviy of cells to vimentin, 200× [Inset 400×] Immunohistochemistry. (b) Photomicrograph showing a negative reaction of cells to S-100, 200× [Inset 400×] IHC

Table 1: Immunohistochemical results

Primary antibody	Source	Clone	Dilution	Results
S-100	Millipore	Mouse Monoclonal, Anti-S-100 Protein	1:200 using IHC-Tek™ Antibody Diluent (Cat# IW-1000 or IW-1001) to reduce background and unspecific staining and serum blocking step is NOT needed.	Negative
Vimentin	Dako	VIM 3B4, Mouse monoclonal	1:400 using IHC-Tek™ Antibody Diluent (Cat# IW-1000 or IW-1001)	Positive
Desmin	Dako	[D33] Mouse Monoclonal, Isotype: IgG1, kappa.	1:200 using IHC-Tek™ Antibody Diluent (Cat# IW-1000 or IW-1001)	Negative

are negative for neuron specific esterase, actin, desmin, laminin and keratin.[3] Desmin negativity of tumor cells excludes its myofibroblastic origin as seen in our case.[1,4] Out of several theories proposed, the most favored are the odontogenic and gingival epithelial theories, which support its origin from the mesenchyme.[10] A variety of tumors can have similar histiologic features. Coarse cytoplasmic granularity and absence of lipid droplets help to distinguish it from rhabdomyoma and hibernoma.[3] Fibroblastic origin of the lesion has been described based on the description of patients affected by neurofibromatosis along with (CGCL).[5]

In summary, we present a case of CGCL in a female neonate. The lesion demonstrated an immunohistochemical profile, which suggests that CGCL is likely derived from primitive gingival mesenchymal cells rather than having a schwannian origin. We conclude that the CGCL is an intriguing lesion with unclear etiology, histogenesis and natural history.

CGCL is a benign lesion and unlike GCT its malignant counterpart, is unknown, and a complete surgical excision is the treatment.

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