

## Case Report

# Diagnosis and management of congenital epulis of new-born: a rare case report and literature review

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### ABSTRACT

The congenital epulis of new-born is a rare disease of idiopathic etiology and lineage. The literature recorded spontaneous regression of some cases. This finding rendered skeptical questions about its neoplastic nature and posed some speculations about hormonal influences, towards the end of pregnancy, on the growth of the lesion. No malignant potential has been ever reported. Surgery is indicated if the lesion grossly interferes with breast feeding. This paper reports an established diagnosis of congenital epulis of new-born, which was excised surgically with no evidence of recurrence hitherto.

**Keywords:** Congenital epulis, Neonatal lesions, Mucosal swellings, Pediatric oncology

### INTRODUCTION

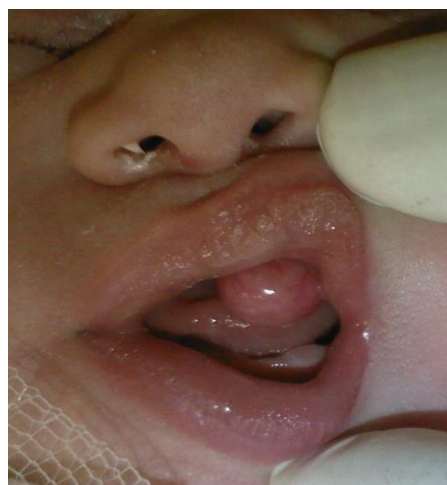
The Congenital Epulis of Newborn (CEN) is a rare congenital tumor of infancy, which affects mainly the alveolar mucosa of the jaws. The exact histogenesis of CEN is controversial. This mucosal growth was first described by Neumann in 1871.<sup>1</sup> Among its other designations, congenital granular cell myoblastoma, granular cell epulis of infancy, granular cell fibroblastoma come atop.<sup>2</sup>

Histologically, the congenital epulis of newborn usually composes of nests of cells with granular cytoplasm set in a scanty to prominent vasculature. Although some cases reported self-involution, the surgical excision is indicated if it grossly interferes with breast feeding.<sup>3</sup>

### CASE REPORT

A ten-minutes-old female neonate was referred to our clinic, from the department of gynecology after delivery, for evaluating the rare maxillary swelling. The baby manifested no other medical problems. Our intraoral

examination revealed a painless sessile nodular lesion whose size was approximately 1 x 1.5 cm. The mucosal color and surface were normal and aspiration was negative (Figure 1).

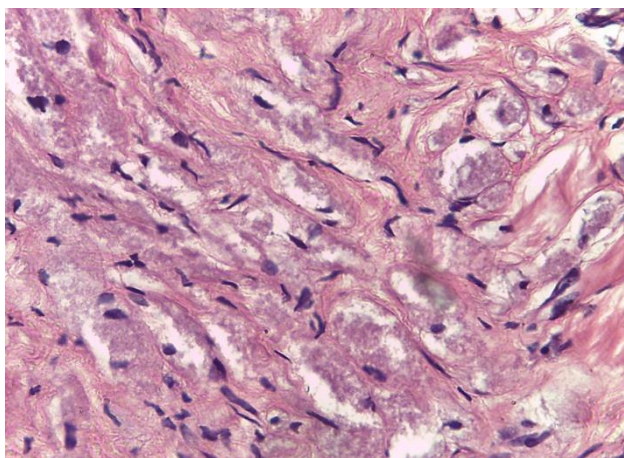


**Figure 1: Clinical picture of the congenital epulis of newborn.**

Melanotic Neuroectodermal Tumor of Infancy (MNET) was excluded via assessing Vanillyl Mandelic Acid (VMA). The patient's mother was assured and educated about the acceptable clinical approach of close observation of this lesion unless interference with feeding or respiration could not be avoided. Thence, the neonate should be scheduled for surgery, under local anesthesia, after ten days since hemoglobin level should be ten and liver should pursue functional consummation.

After ten days, the baby was reexamined. The lesion underwent no regression. Her mother reported an encumbrance of mammary feeding and consented for surgical removal. It was when the lesion was excised under local anesthesia (topical ointment and 0.8 mL lidocaine®) and sent for (immuno)-histological examinations. The differential diagnosis included both congenital epulis of the newborn and mucosal cyst of newborn since melanotic/primitive neuroectodermal tumor has been previously ruled out. Microscopic examination exhibited large, rounded and polyhedral, histiocyte-like cells with small, dark oval nuclei and abundant eosinophilic granular cytoplasm. No cystic cavitations were observed. Neither mitotic figures nor malignant criteria were observed (Figure 2).

Again, the specimen expression of CD99 was negative; ruling out the neuroectodermal tumor. The diagnosis was established as congenital epulis of newborn.



**Figure 2: An H&E stained photomicrograph showing rounded to spindle cells with an abundant eosinophilic granular cytoplasm. Original magnification 40x.**

## DISCUSSION

The congenital epulis is a rare benign soft tissue lesion, which prominently affects the anterior alveolar ridges of both jaws of neonates. With regard to gender and site, most cases have a striking predilection for females and maxilla.<sup>4</sup> The usual size is small but lesions of more than seven centimeters were reported. Classically, CEN is a soft, pedunculated and perhaps lobulated nodule of normal color and texture.<sup>3</sup> Despite its solitary occurrence,

multiple synchronous incidence has been reported including unusual site.<sup>3,5</sup> In a multiple case of CEN, tongue affection was traced.<sup>6</sup>

Concerning its uncertain histogenesis, odontogenic epithelium, fibroblasts,<sup>7</sup> Langerhans cells or histiocytes,<sup>8</sup> smooth muscle, nerve-related cells,<sup>9</sup> endothelial cells, pericytes, myofibroblasts, and undifferentiated mesenchymal cells<sup>10</sup> have been implicated in the pathogenesis of CEN, which, however, remains undefined.<sup>11</sup>

Microscopically, CEN is composed of sheets of large to medium-sized, round to polyhedral cells that have a discrete cellular membrane, abundant granular cytoplasm, and a single darkly staining nucleus with an eccentrically positioned nucleolus. The covering epithelium is thin and attenuated. Odontogenic epithelium rests may be found among the granular cells.<sup>3</sup> Spindle cell epulis, as a variant, was reported in an 8-month-old child whose histological picture displayed ovoid cells with vesicular nuclei and scant, non-granular eosinophilic cytoplasm.<sup>12</sup>

Vascular channels vary between granular cells, but fibrous stroma is minimally conspicuous.<sup>13</sup> Unlike the granular cell tumor (myoblastoma), the atrophic overlying epithelium never demonstrates the pseudoepitheliomatous hyperplasia. Granularity of the lesion is observed in other neoplastic maxillofacial or odontogenic lesions including granular ameloblastoma, verruciform xanthoma and others. However, age and clinical parameters can easily distinguish them all.

Immunohistochemically, granular cells are positive for vimentin and, in most cases, for Neuron-Specific Enolase (NSE), but negative for Leu7 and S-100 protein. Moreover, lesional cells do not immunoreact for laminin, as do the granular cells of the granular cell tumor. Granular cells are reactive only toward vimentin. Such cells are, moreover, strongly positive for acid phosphatase.<sup>9-11,13</sup>

Slootweg et al.<sup>14</sup> concluded that peanut lectin binding protein might be considered a common feature of granular cells regardless of their histogenesis. Lysosomes are supposed to represent the intracellular binding sites for this marker. Moreover, it was shown that granularity, in congenital epulis of newborn and granular cell ameloblastoma, posed no significant difference in their histogenesis since the former are of mesenchymal derivation while the latter, from their intermediate filament protein types which appear to originate from the epithelium.

Differential diagnosis of CEN includes mucosal cyst of newborn and MNET if premaxilla is encountered. To exclude MNET, VMA test is mandatory since CEN, on the one hand, does not change the level of VMA. On the other hand, high levels of VMA are common findings in the individuals with neural cell tumors such as

neuroblastoma, retinoblastoma, pheochromocytoma, and ganglioneuroblastoma, indicating neural crest origin of the tumor.<sup>15</sup>

The surgical excision is the classical treatment in large lesions. CEN never recurs even if the removal is incomplete. However, no treatment is required, in smaller lesions, once the diagnosis is confirmed and aggressive lesions are excluded.<sup>3</sup>

## CONCLUSION

This paper reports a typical case of CEN whose size was moderate. The lesion was excised under local anesthesia at day TEN of age. The diagnosis was histologically and immunohistochemically confirmed. The assurance of parents cannot be overemphasized. Sonographers should pay a rapt attention of similar lesions especially larger ones during the interval of prenatal delivery.

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