

Congenital Epulis

Oren Lapid, MD*; Ruthy Shaco-Levy, MD‡; Yuval Krieger, MD*; Leonid Kachko, MD‡; and Amiram Sagi, MD*

ABSTRACT. Epulis is a rare tumor of the newborn, also known as granular cell tumor of the newborn or Neumann's tumor. This tumor arises from the mucosa of the gingiva, most commonly from the anterior part of the maxillary alveolar ridge, and is typically seen as a mass protruding out of the newborn child's mouth, which may interfere with respiration or feeding. Epulis is seen only in the newborn and is a different entity from other granular cell tumors. The tumor has a marked female preponderance of 8:1. The recommended treatment is prompt surgical resection. Recurrences of the tumor and damage to future dentition have not been reported, suggesting that radical excision is not warranted.

A newborn female with such a mass is described. The tumor was resected using a carbon dioxide laser; the postoperative course was uneventful. On histologic examination, it was composed of diffuse sheets and clusters of polygonal cells containing small round to oval nuclei and abundant coarsely granular cytoplasm. The tumor cells stained positive for vimentin, and negative for S100-protein, actin, desmin, laminin, keratin, estrogen, and progesterone receptors. Electron microscopic examination showed granular cells containing heterogeneous electron-dense granules, lysosomes, and cytoplasmic lipid droplets. The clinical and microscopic features of such tumors are reviewed. *Pediatrics* 2001;107(2). URL: <http://www.pediatrics.org/cgi/content/full/107/2/e22>; congenital epulis, granular cell tumor, gingival granular cell tumor.

ABBREVIATIONS. GCT, granular cell tumor; GGCT, gingival granular cell tumor.

Epulis, or congenital granular cell tumor (GCT), is a rare tumor of the newborn. It is seen as a mass arising in the mouth from the alveolar ridge; this mass may interfere with respiration or feeding. A case of congenital epulis is reported; the tumor was resected using a carbon dioxide laser. The clinical aspects as well as the morphologic, histologic, and electron microscopic features of this lesion and its treatment are reviewed.

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CASE REPORT

Presentation

A newborn girl was referred to the plastic surgery department at the Soroka Medical Center immediately after birth because of a mass protruding from her mouth (Figs 1 and 2). The infant was born on the 36th week of gestation. Pregnancy and parturition were normal. The birth weight was 2700 g (Appgar: 8–10). Ultrasound scans performed at the 25th week of gestation did not reveal any abnormality. At birth a firm pedunculated tissue mass was found protruding from the mouth. The mass was attached to the gingiva of the left maxilla by a stalk measuring 2 cm by 1 cm. The mass prevented normal closure of the mouth and hindered breast or bottle-feeding, but did not cause airway obstruction or respiratory distress. No other pathologic finding was noted. Nasogastric feeding was instituted and 2 days after birth, the infant was operated on under general anesthesia with nasal intubation. Surgery was performed using the carbon dioxide laser set at 15 watts continuous wave (Sharplan Lasers Inc, Migdal Haemek, Israel). 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 feedings were instituted on the first postoperative day and the child was discharged on the following day. Healing was uneventful and the gingiva reepithelialized within 10 days.

Pathologic evaluation of the tumor confirmed the diagnosis of



Fig 1. The mass is seen protruding from the girl's mouth (lateral view).

Fig 2. The mass is seen protruding from the girl's mouth (anterior view).

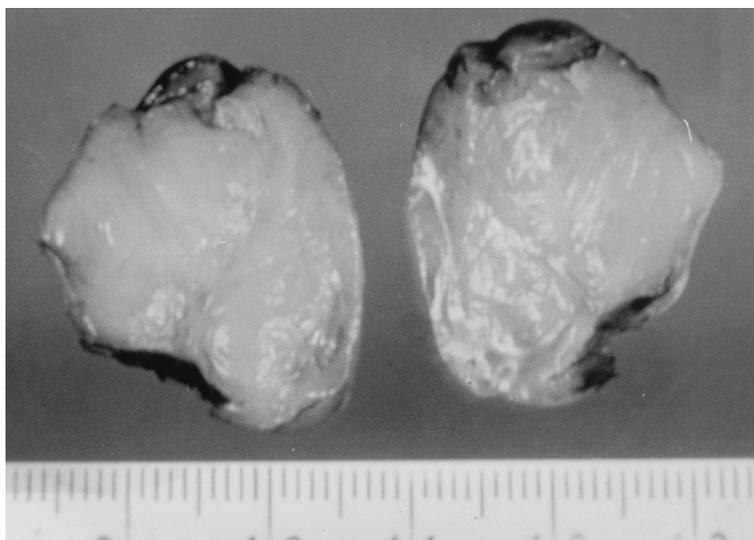


Fig 3. The mass cut in 2. Note the homogenous appearance.

a congenital GCT. Macroscopically, the external surface was slightly irregular and the cut surface was homogenous and tan (Fig 3). Histologically, it was composed of diffuse sheets and clusters of polygonal cells containing small round to oval nuclei



Fig 4. Low-power microscopy $\times 40$. At the top of the slide, one may see the normal squamous epithelium and below it the tumor, which is composed of polygonal cells with a meshwork of delicate blood vessels.

and abundant coarsely granular cytoplasm. There was a delicate plexiform network of capillaries (Figs 4 and 5).

The tumor cells stained positively with Periodic acid-Schiff reagent and the staining was not removed by pretreatment with diastase.

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.

Portions of the tumor were obtained fresh and immediately fixed in 2.5% cold buffered gluteraldehyde and then processed routinely for electron microscopy. Electron microscopic examination showed granular cells containing heterogeneous electron-dense granules, lysosomes, and cytoplasmic lipid droplets (Fig 6). 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.

DISCUSSION

Epulis is also known as GCT of the newborn, or as Neumann's tumor after the first published case.¹

This tumor is rare, as reviewed by Zuker and Buenecha² in 1993, who described only 167 reported cases.

The tumor is seen only in the newborn and is a

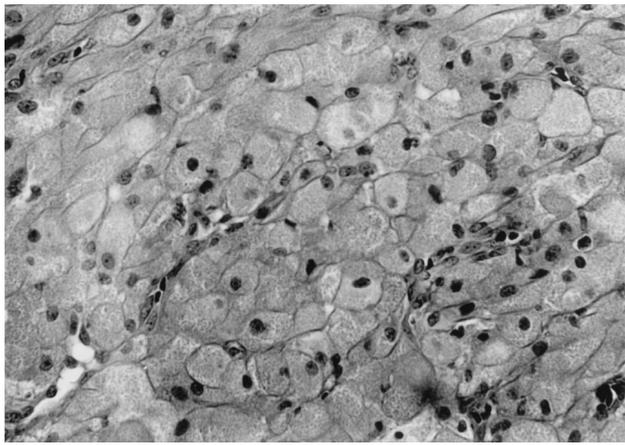


Fig 5. High-power microscopy $\times 400$. Polygonal cells with granular cytoplasm and small round nuclei.

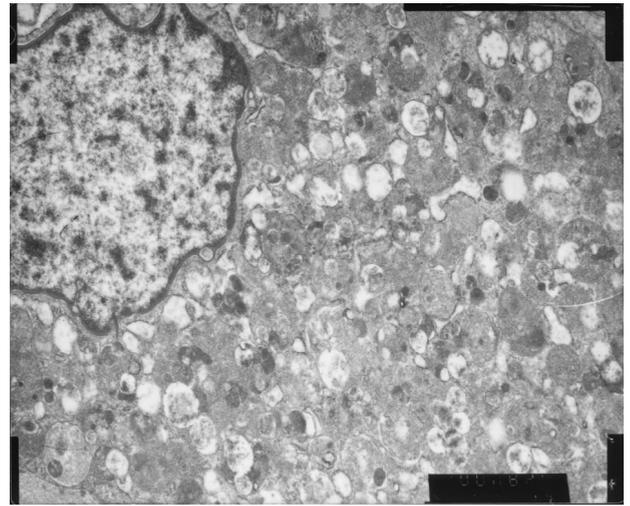


Fig 6. Electron microscopy $\times 10\,000$. Electron-dense granules and numerous lysosomes within the cytoplasm.

different entity from other GCTs. The tumor has a marked female preponderance of 8:1. It arises from the mucosa of the gingiva most commonly from the anterior part of the maxillary alveolar ridge, although it has been described as arising from the mandibular gingiva as well as from several locations simultaneously. Reported size varies from several millimeters to 7.5 cm.

There is a striking histologic similarity of granular cells of gingival granular cell tumors (GGCTs) and the far more common 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 capillaries, and lack of pseudoepitheliomatous hyperplasia.³ However, GGCTs contain cells that are virtually indistinguishable by light microscopy alone from those seen in GCTs. Although Schwann cells are considered to give rise to most GCTs, and, hence, these tumors show diffuse and strong staining for S100-protein, the histogenesis of GGCTs remains unclear. A few theories exist as to the origin of GGCTs, and the most popular ones favor gingival stromal (mesenchymal) origin and odontogenic epithelial origin.⁴ The ultrastructural and immunohistochemical findings in our case are in agreement with those supporting a mesenchymal origin; the cells have features of histiocytes and fibroblasts. The ultrastructural findings of the tumor that we describe are clearly different from those in GCT (irregular cell borders with extensions, lack of basement membrane, and absence of Schwannian cells). Furthermore, as in the case described by Larralde et al,⁵ the tumor cells in our case were reactive for the macrophage marker CD68. They were also diffusely and strongly positive for vimentin. The tumor cells in our case did not react with S100-protein, and this further supports the contention that GGCT and GCT arise from different cells of origin.

Because GGCT is a congenital tumor in which spontaneous regression has been reported,^{6–8} we performed immunohistochemical stainings for estrogen and progesterone receptors, to see whether the tumor is influenced by maternal hormones. The re-

sults of both of these stainings were negative. Although the development of the tumor in the uterus and the higher incidence in females imply a hormonal mechanism, it probably takes place through different pathways.

CONCLUSION

Our case provides additional evidence that GGCT derives from a mesenchymal origin, unlike the GCT of any other site.

The recommended treatment is prompt surgical resection. Recurrences of the tumor and damage to future dentition have not been reported, suggesting that radical excision is not warranted.

The tumor is often misdiagnosed before surgery because of its rarity and lack of awareness by clinicians.² The differential diagnosis includes: hemangioma, lymphangioma, fibroma, rhabdomyoma, and heterotopic gastrointestinal cysts.⁷

In our case the tumor was not diagnosed by ultrasound on the 25th week of gestation, suggesting that it may have developed later. There have been reports of prenatal diagnosis and in those cases, in which the tumor was detected late in gestation.⁹

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