CLINICAL COMMENTARY

Obstructive congenital epulis

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Summary

Introduction: Congenital epulis, known as a congenital gingival granular cell tumor, is a benign tumor and very rare in newborns. Voluminous or multiple tumors can cause mechanical obstruction of the oral cavity and may result in postnatal feeding and respiratory problems.

Discussion: We report the clinical case of a female full-term newborn who presented a tumor on the upper gum protruding into the oral cavity discovered at birth. The pregnancy was followed normally with three prenatal ultrasounds, which did not show abnormalities. The mass was excised under local anesthesia on the second day of life. The outcome was good after surgery and regular feedings were started on the second postoperative day. Histological examination confirmed the diagnosis of gingival tumor with granular cells and absence of signs of malignancy.

Conclusion: Prenatal diagnosis is fundamental in the therapeutic approach to this rare lesion but remains difficult because the findings are non specific and the generally late development of the tumor.

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Introduction

Congenital epulis, or congenital gingival granular cell tumor, is a benign, very rare tumor of the oral cavity in newborns. There is a single tumor in most cases, most often arising from the maxillary gingival mucus tissue and is 10 times more frequent in girls than in boys [1]. Large or multiple tumors can threaten the vital prognosis of the newborn because of the mechanical obstruction of the oral cavity interfering with respiration and oral feeding. The diagnosis is often made postnatally during neonatal care. However, prenatal diagnosis is important to decide on the route of delivery and plan early multidisciplinary postnatal management [2]. Based on a case of obstructive congenital epulis, we discuss the diagnostic, therapeutic, and progressive aspects of this rare neonatal tumor.

Case study

The patient was a newborn girl hospitalized at birth in our neonatology unit for a tumor mass in the oral cavity discovered in the delivery room. Her mother was 19 years old,
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GII, PII, with no notable medical history. The pregnancy had been monitored with three fetal ultrasound examinations that did not demonstrate abnormalities. The full-term infant was delivered vaginally without incident, with an Apgar score of 8, then 9 at 1 minute and 5 minutes, respectively. The clinical exam found normal trophicity and the infant weighed 3700 g, measured 48 cm, had a cranial perimeter of 35 cm, a bilobed tumor mass obtruding from the oral cavity, measuring 3 × 4 cm, pink, with a firm consistency, a smooth surface, sessile, and implanted at the alveolar ridge of the upper maxillary. This mass prevented closure of the mouth and oral feeding, with no repercussions on respiration; the infant was given a venous perfusion of 10% glucose solution (Fig. 1).

Transfontanellar and abdominal ultrasound examinations searching for associated malformations were normal. The tumor was excised on the second day of life under local anesthesia with no complications and bottle feeding introduced on the second postoperative day. The infant left the hospital on her fourth day of life with normal closure of the mouth and good suckling (Fig. 2).

The anatomopathological examination of the surgical specimen showed a bilobular, encapsulated, smooth mass with sessile implantation (Fig. 3).

Discussion

Congenital epulis or gingival granular cell tumor is a very rare neonatal tumor. Since the first description made by Neumann in 1871, approximately 200 cases have been reported in the literature [1]. It is also known as congenital myoblastoma or Neumann tumor [3]. The majority of cases are observed in newborn girls with a 10/1 sex ratio. Recently, one case of congenital epulis in a boy was reported by Nouri et al. [1].

The exact etiology of this benign tumor remains unknown, but several theories have been advanced, i.e., myoblastic, neurogenic, odontogenic, fibroblastic, histiocytic, and endocrine. The female predominance argues in favor of the endocrine theory [4,5]. Histological and
The diagnosis of large congenital epulis is often made at birth, as in our patient, or immediately after birth during the systematic examination of the newborn when the tumor is very small. Classically, there is a single firm tumor with a quite regular surface, sometimes multinodular or sessile or pediculated, pink or red in color, and non painful on palpation [1,2]. The tumor size varies from a few millimeters to around 10 cm at its widest diameter [1,6]. The upper maxillary location opposite the canines or incisives is the most frequent, but the mandibular region can also be involved. Multiple lesions or maxillary and mandibular locations have been described in 5—16% of cases [1,3,7]. The clinical manifestations depend on the size and place of the lesions. Postnatally, large tumors can threaten the newborn’s vital prognosis because of a mechanical occlusion of the oral cavity, hindering the closure of the mouth, disrupting respiration, and preventing oral feeding [8].

The classical clinical aspect of the tumor mass often suggests the diagnosis. However, congenital epulis raises the differential diagnosis of other neuroectodermal lesions of the oral cavity in the newborn, i.e., teratoma, leiomyma, hemartoma, congenital dermoid cyst, congenital fibrosarcoma, congenital lipoma, hemangiomma, and granuloma [3,9]. In all cases, congenital epulis is not associated with other abnormalities of the oral cavity or congenital malformations [10].

Anatomopathological examination of the tumor specimen confirmed the diagnosis by demonstrating the tumor to be formed with large round or polygonal cells with round or oval nuclei, with granular cytoplasm and tumor stroma (Fig. 4). Congenital epulis is distinguished from other giant cell tumors by its exclusively gingival location, the presence of typical epithelium of the gingival mucus membrane, vascularization of the tumor stroma that is very rich in collagen, the absence of nerve components, and by an immunohistochemical profile characterized by the absence of protein S-100 expression [3,6,11].

Surgical excision under general anesthesia is needed urgently in cases of very large and obstructive tumors interfering with respiration or in cases of substantial hemorrhage [4,8]. If not, surgery can be performed under local anesthesia a few hours after birth, as in our patient, for surgery in optimal conditions [2]. The choice between local and general anesthesia depends on the size of the tumor, whether it is pediculated or sessile, and on the number of lesion locations. Most authors have adopted general anesthesia because of the large size of the tumor or multiple locations [1,2,4]. However, as in our case, other authors have preferred local anesthesia with a single and sessile tumor [6,10]. The main complication of surgical excision is hemorrhage, particularly with a sessile tumor. Complete healing of the gingival mucus is obtained within 2 weeks with no complications. However, the main risk of wide surgical excision is affecting physiological tooth eruption [3,6]. Some authors have used electrocauterization or pulsed CO₂ laser [12]. In all cases, no degeneration or recurrence has been described, even after incomplete tumor excision. In cases with small tumors, therapeutic abstention with regular follow-up can be proposed, because although rare, regression by spontaneous necrosis is possible [1,8].

Prenatal diagnosis is fundamental for the therapeutic approach of granular cell gingival tumors. This is possible but remains difficult because there are no specific signs orienting the examination and the tumor generally develops late, beyond 22 weeks of gestation [1]. In the fetus, large lesions can hinder swallowing with development of hydramnios, sometimes acute, which can diagnose the tumor. Fetal three-dimensional ultrasound and magnetic resonance imaging can provide the diagnosis by the 36th week of gestation [3]. Thus, the route of delivery can be decided in advance and early multidisciplinary management of the newborn can be planned, with the participation of the neonatologist for resuscitation at birth, the maxillofacial or head and neck surgeon or the pediatric surgeon to program the surgical excision of the tumor, and the anesthesiologist to choose the anesthesia modalities. The risk of hemorrhage in large lesions with pediculated attachment may warrant cesarean delivery [1,3].

Conclusion

Congenital epulis is a very rare benign neonatal tumor. The diagnosis is often suspected clinically at birth. Surgical excision should be take place urgently in case of a large obstructive tumor. The long-term results are excellent, even after incomplete excision. Prenatal diagnosis is fundamental to plan for early multidisciplinary management of both the mother and her child.

Conflict of interest

None

References


