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Case report

Congenital Epulis: A Case Report

Khadija Haghegh*, Mohammed Almughrabi

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Department of Pediatric Surgery, Tripoli University Hospital, Tripoli, Libya.
***Correspondence**: kh.haghegh@gmail.com

Abstract

Congenital epulis is a rare and benign tumor affecting the oral cavity which is predominantly observed in female infants. It originates from the gingival mucosa and its size varies from few millimeters to few centimeters. While spontaneous regression is possible, larger congenital epulis lesions can lead to airway obstruction and feeding difficulty, which is why surgical excision is considered the treatment of choice. Our case sheds light on this condition, presenting the management done to a two-day-old female newborn diagnosed with an epulis originating from the anterior maxillary alveolar ridge at birth. The mass was successfully excised under general anesthesia, leading to a positive outcome as evidenced by the return to regular feeding on the first postoperative day. Histological examination verified the presence of large polygonal granular cells with granular eosinophilic cytoplasm and round central nuclei which confirms the diagnosis of congenital epulis. This case underscores the importance of early recognition and multidisciplinary collaboration in managing congenital epulis to ensure favorable outcomes

Keywords*: Congenital Epulis, Granular Cell Tumor, GCT, Mouth Tumors.*

Introduction

Congenital Epulis, also known as congenital granular cell tumor or Neumann's tumor, is a rare benign tumor that occurs in newborns with the incidence of 0.0006% [1]. The initial description of this lesion dates back to 1871 when reported first by Neumann [2]. It mostly occurs in females with a 10:1 female to male ratio [3] and predominantly originates from the maxillary alveolar ridge, being three times more prevalent there than in the mandible. In around 10% of cases, simultaneous involvement of both maxillary and mandibular alveolar ridges has been documented [4]. These growths may present as either sessile or pedunculated, with colors ranging from pink to reddish [5]. The primary differential diagnosis is epignathus, an oral teratoma [6].

These tumors can manifest as large masses potentially leading to airway blockage and feeding difficulties. Which is why the preferred treatment involves early surgical excision [7]. However, there have been instances of the lesion regressing spontaneously, even without surgical intervention [8].

Given its infrequent occurrence, there have been instances of misdiagnosis pre-operatively. Therefore, reporting cases of congenital epulis is warranted to spread awareness of this condition among medical professionals to ensure accurate diagnosis and timely management. [3].

Case report

A healthy 2-day-old girl was born with a large mass protruding from her mouth. The baby was born to a para 6 mother via emergency Cesarean section due to polyhydramnios and a tender scar. The baby was born full-term with a weight of 3.400 kg and an Apgar score of 8-9-9, the prominent oral mass necessitated referral from Tarhouna to P.M.H in Tripoli for surgical management.

Upon examination, baby looked mildly icteric but well and active in the incubator, Respiratory difficulties were not noted. She was well hydrated and perfused on IVF.

Two fleshy, rubbery, slightly ulcerated pedunculated masses were observed originating from both the upper and lower alveolar ridges. The largest one was the upper mass which measured 1.5 × 2 × 1 cm and occupied most of the mouth. (Figure 1). A nasogastric tube was introduced to facilitate feeding which passed easily. At this point, two preliminary differential diagnoses were considered, including Congenital Epulis and mouth Teratoma (epignathus).

The systemic examination revealed no notable abnormalities. Routine laboratory tests were within the normal range. The baby was covered with antibiotic. A pelvic/abdominal ultrasound did not reveal any significant issues. A brain MRV study showed normal cerebral vessels and described an oral soft tissue lesion protruding through the mouth, measuring about 25X30X19 mm, for subsequent histopathological correlation. (Figure 2)

On the eighth day of her life, under general anesthesia using a Miller blade with a retromolar approach, the patient underwent a surgical resection of these masses. Both masses were effectively excised with elliptical incisions made to the peduncles, and hemostasis was achieved using diathermy, leading to minimal blood loss. Notably, the child resumed oral feeding successfully on the first post-operative day, indicating a favorable recovery. (Figure 3)

The two masses were subjected to histological examination. The analysis revealed thin atrophic non-keratinized squamous epithelium with focal ulceration and serum crust formation. In the underlying submucosa, there was evidence of tumor cell growth arranged in confluent to nodular cords and sheets. These cells were characterized by large polygonal shapes, abundant eosinophilic granular cytoplasm, and small central nuclei. Importantly, no signs of malignancy were observed, confirming the diagnosis of Congenital Granular Cell tumor, commonly known as Congenital Epulis. (Figure 4,5,6)

Figure 1: A large pedunculated ulcerative mass arising from the upper maxillary alveolar ridge (white arrow), small smooth mass seen in mandibular alveolar ridge (black arrow).

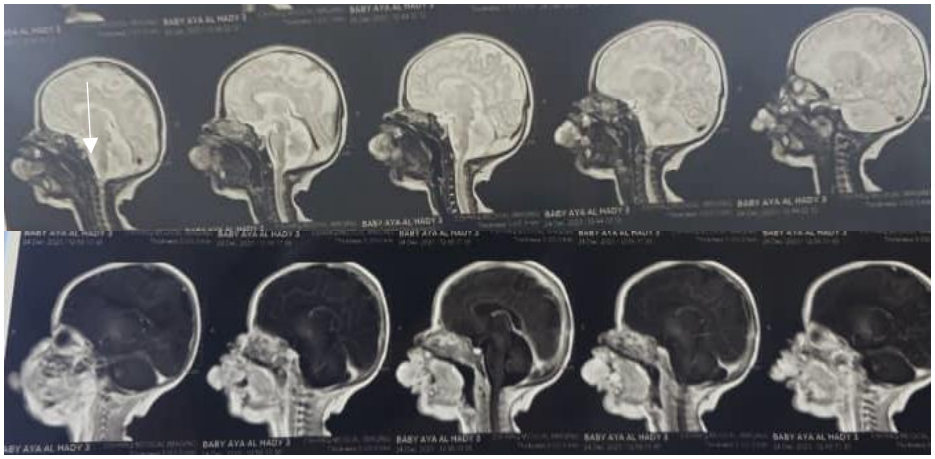

Figure 2: Magnetic Resonance Venography of brain (MRV). Axial T2-weighted MRI section shows well-defined abnormal mixed signal intensity soft tissue lesion protruded through the mouth. measuring about 25X30X19 mm (arrow)

Figure 3: Clinical image. Post-excision.

Figure 4: Gross specimen of upper and lower alveolar arch masses measuring 1.6X2.0 cm and 1.3 cm, respectively.

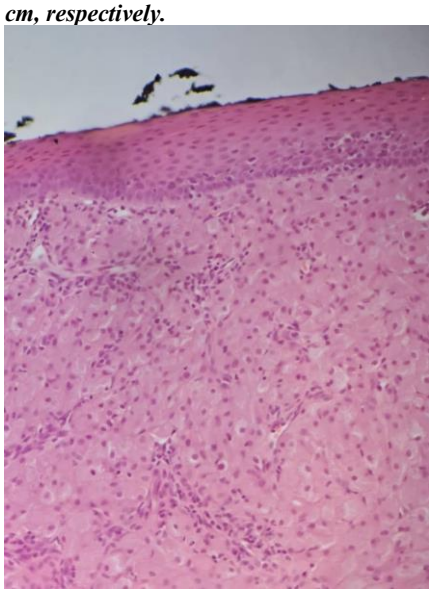

Figure 5: Microscopic examination of excised specimen demonstrating surface squamous epithelium with underlying diffuse sheets of large polygonal tumor cells displaying abundant granular eosinophilic cytoplasm and round central nuclei, confirming granular cell tumor. (H&E, 200x)

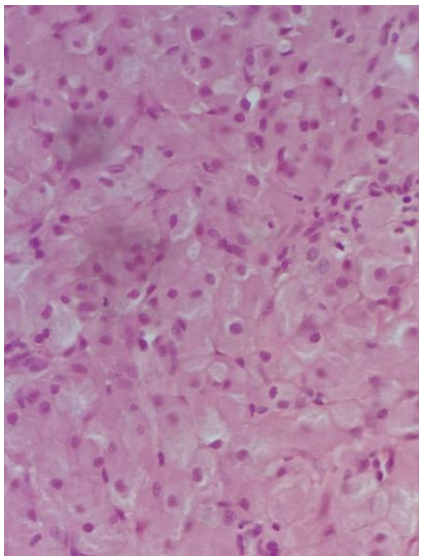

Figure 6: Granular cell tumor; Polygonal tumor cells with round central nuclei and abundant granular eosinophilic cytoplasm. (H&E, 400x)

Discussion:

Congenital epulis, also known as gingival granular cell tumor, is a rare tumor that presents as a pedunculated pink, firm, lobulated mass on the infant's alveolar ridge, more frequently on the maxilla (maxillary/mandibular ratio 3:1). It typically appears in the region between the incisors and canines [9]. Reports of size range from 1 mm to 9 cm in diameter [10]. The tumor mass is in often solitary. However, multiple lesions can occur in up to 10% of cases and therefore total inspection of the oral cavity is needed [11]. It predominantly affects females and is treated promptly with surgical excision, with rare recurrence [4].

Congenital epulis presents sporadically and has no familial tendency, nor association with teratogens (12,13). Although its etiology is unknown, some theories suggest mesenchymal and reactive origins [11,12]. Others suggested that given the female predominance, cessation of growth after birth, and spontaneous regression of the tumor in some cases, the cause may be due to maternal or fetal hormones during pregnancy; however, estrogen and progesterone receptors were not found positive in congenital epulis [13].

In our case, the two preoperative main differential diagnosis were congenital epulis and *epignathus, which* is a form of oropharyngeal teratoma that arises from the palate. Both tumors represent rare benign oral tumors encountered mostly in female neonates. They both can potentially cause respiratory distress with clinical obstructive symptoms such as dyspnea, suffocation, and difficulty in feeding. Despite sharing certain clinical similarities, they possess distinctive etiological and histological characteristics [14-16].

Congenital epulis is frequently characterized as a solitary mass, either pedunculated or sessile, emerging from the gingival mucosa, primarily localized to the anterior maxillary alveolar ridge. Although its etiology remains unclear, histological examination of congenital epulis tumors show distinctive sheets of polygonal cells with round to oval vesicular nuclei and abundant eosinophilic granular cytoplasm [14].

In contrast, epignathus is believed to arise from pluripotent cells situated within Rathke's pouch; an anatomical depression located in the developing oral cavity anterior to the buccopharyngeal membrane. Approximately 60% of epignathus cases originate from the nasopharynx. It is less uncommon compared to congenital epulis with incidence of 1:35,000 to 1:200,000 in live births [15]. Its size can be massively large reaching 30 cm × 40 cm, associated with higher mortality and morbidity rates because of severe airway obstruction and other malformations [16].

Histologically, epignathus tumors exhibit complex features, including well-differentiated cell lines derived from all three germ cell layers. They even might contain well-formed organs and limbs with bony parts and hair which strongly suggests a teratoma. Therefore, some regard epignathus as a parasitic fetus [17-19]. It is also frequently associated with cleft palate [19] and high levels of alpha-fetoprotein which could be used for diagnosis and follow-up post resection [20].

MRI can be used for diagnosis, showing the gingival origin of Congenital Epulis without local invasion, thus, differentiating congenital epulis from epignathus which arise from the oropharyngeal cavity and might show intracranial invasion [20,21].
Similarly, surgical resection is considered the treatment of choice for both congenital epulis and epignathus. No case of malignancy has been reported in neither tumor; and reports of recurrence post-resection are rare [18].
The use of antenatal ultrasonography and fetal MRI can be helpful in diagnosing large oropharyngeal lesions before birth. Due to the potential for causing respiratory difficulty, pregnant mothers of a child with suspected oral mass antenatally should be transferred to a tertiary center, where the delivery can be conducted and supervised by a multidisciplinary team composed of a gynecologist, neonatologist, anesthesiologist, ENT specialist , and pediatric surgeon, who must all work together during the labor and after birth to improve the baby's outcome. Suggested steps during the delivery include maintaining the umbilical cord and fetal circulation to oxygenate the fetus while a rapid examination is done. If the child shows signs of respiratory distress, an EXIT procedure (ex-utero intrapartum treatment) may be required, which involves establishing an airway before the feto-maternal circulation is interrupted. Furthermore, a tracheostomy can be performed if necessary. After a patent airway is secured, clamping of the umbilical cord can be done and the baby is sent to the NICU for further management [14,18,20,22].
In conclusion, we report a rare case of congenital epulis. Early recognition and multidisciplinary collaboration in the management of such case is important to ensure favorable outcomes.

Conflicting Interests
There are no conflicts of interest.

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