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Multiple congenital epuli (congenital granular cell tumours) in the newborn: a case report and review of literature

Monika Dzieniecka[1], Anna Komorowska[2], Adriana Grzelak-Krzymianowska[3], Andrzej Kulig[1]

[1]Department of Clinical Pathology, Research Institute of the Polish Mother's Memorial Hospital, Łódź
[2]Department of Otolaryngology, Research Institute of the Polish Mother's Memorial Hospital, Łódź
[3]Medical University of Łódź, Foreign Language Teaching Center, Łódź

Congenital epulis of newborn is very rare benign intraoral entity of uncertain ethiology. Histologically the lesion is similar to the granular cell tumour of an adult but immunohistochemical stainings prove their different origin. Treatment involves surgical excision, recurrences are rare.

Key words: congenital epulis, granular cell tumour of the newborn, Neumann's tumour, gingival tumour, newborn.

Introduction

Congenital epulis, also known as congenital granular cell tumour (CGCT), granular cell fibroblastoma, congenital granular cell myoblastoma, or Neumann's tumour, is a very rare intraoral tumour. The first diagnosed case was published in 1871. The preferred and accepted term is congenital epulis of the newborn, which points to its non-neoplastic origin. This kind of epulis is a benign, reactive or degenerative lesion, usually single and typically situated on the alveolar mucosa of the jaws, most commonly on the anterior part of the maxillary alveolar ridge (3 : 1 maxillary site predilection). It occurs almost exclusively in Caucasian newborns and has a 8-10 : 1 female ratio predilection, which may indicate the role of a hormonal component in its development. Granular cell tumour is not associated with any other congenital malformations. The histogenesis of the Neumann's tumour is still uncertain and controversial. The prenatal diagnosis of congenital epulis has been recently discussed in the medical literature. Sometimes in the prenatal diagnosis, polyhydramnion is observed, especially when the lesion is large enough to evoke obstruction of the respiratory tract or oral cavity.

We report a three-day-old female newborn with postnatal diagnosis of congenital multiple epulis and review the relevant literature. The aim is to discuss the clinical, microscopic features, the differential diagnosis and complications.

Case report

A three-day-old female newborn was admitted to the Department of Otolaryngology of the Research Institute of the Polish Mother's Memorial Hospital with two round, smooth, pedunculated masses on the anterior ridge of the maxilla. The prenatal ultrasound showed no abnormalities. No family history of hereditary diseases was reported. The infant was delivered in the 40th week of gestation without difficulty, with Apgar score 8 at minute 1, 5 and 10 after birth; the birth weight was 3350 g. The baby girl did not require any mechanical respiratory support because both tumours were small and measured 0.5 and 0.3 cm at their respective greatest dimensions. Both lesions were well defined, firm, round, smooth and pink coloured on the cut surface. Both alveolar masses were excised surgically without complications under local anaesthesia on the 3rd day of the baby's life who was discharged on the same day. The microscopic examination of the lesions revealed nodular formations composed of homogeneous, large, round to oval histiocyte-like cells with distinct cell borders

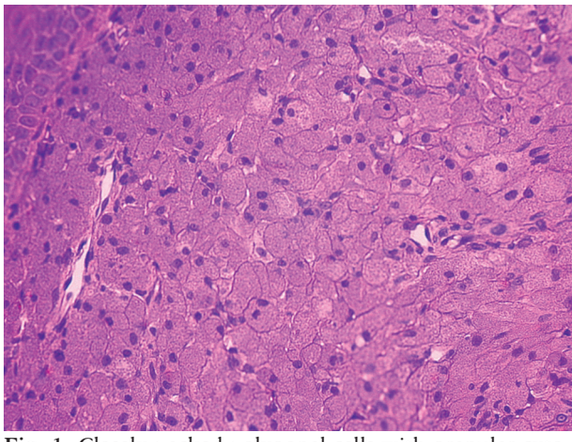

Fig. 1. Closely packed polygonal cells with granular cytoplasm and small round regular nuclei with inconspicuous nucleoli (HE, magnification 400×)

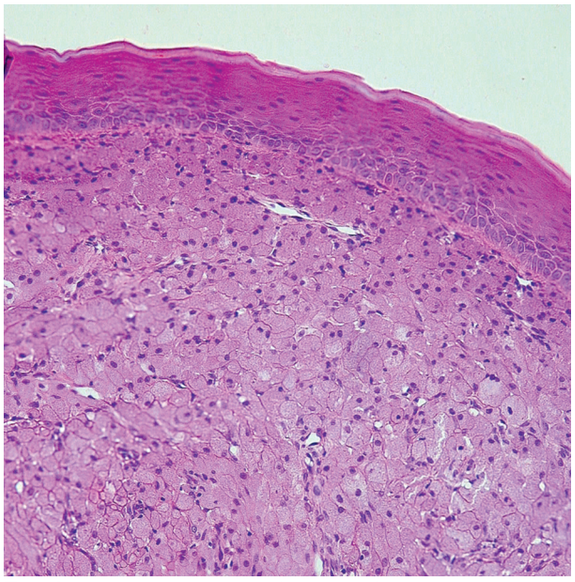

Fig. 2. At the top of the slide there is normal squamous epithelium and below it – the tumour, which is composed of round to oval cells with abundant granular eosinophilic cytoplasm and uniform nuclei (HE, magnification 400×)

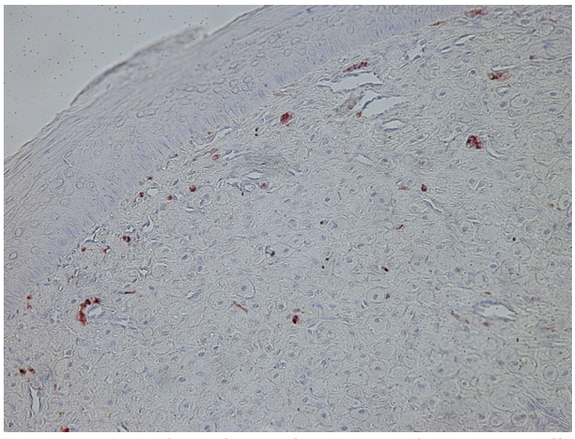

Fig. 3. Immunohistochemical staining. The tumour cells are negative for S 100-protein (magnification 200×)

and abundant granular, eosinophilic cytoplasm, eccentrically placed, small, round, dark nuclei and absent nucleoli. The lesions were covered with normal squamous epithelium.

Immunohistochemically detected cells did not immunoreact to S-100-protein. The diagnostic hypothesis of congenital epulis of the newborn was confirmed based on both the histological details and immunohistochemical profile of the masses.

Follow-up for the next 3 months of the baby's life did not show signs of recurrence.

Discussion

Congenital epulis (CE), more recently renamed a granular cell tumour of the newborn, is a very rare lesion and arises from the mucosa of the gingiva, typically from the anterior part of the maxillary alveolar ridge, but the cases are also reported where the lesion was localized in the mandibular gingiva (with the 3 : 1 maxillary predilection) [1-5]. It is a benign and usually single mass, but can be multiple in 10% of the cases. CE affects mostly white children and has a 8-10 : 1 female preponderance [6-10]. The endogenous hormonal factors might influence growth in utero [11, 12].

Our case is a female newborn with two lesions localized on the anterior ridge of the maxilla. CGCT usually is not associated with abnormality of teeth or other congenital malformations, like in our case. The only exceptions are large lesions which can cause hypoplasia of the jaws, a hypoplastic or absent underlying tooth and the possibility of mild midface hypoplasia, also called Binder syndrome [1, 7]. The tumoral mass protruding through the mouth of a neonate can interfere with respiration and feeding, depending on the size [6]. Only one case report documented associated genital anomalies (hypospodiasis) [13].

The histogenesis is uncertain, and proposed derivations include odontogenic epithelium, undifferentiated mesenchymal cells, pericytes, fibroblasts, smooth muscle cells, nerve related cells, and histiocytes [2, 3]. However, the currently accepted theory is that the epulis represents a reactive entity, and recent immunohistochemical staining and ultrastructural examination favour myofibroblasts as the cell of origin. Clinically, lack of growth, documented spontaneous regression and lack of recurrence even after incomplete resection and lack of malignant transformation further support the reactive theory [7, 14, 15].

The main differential diagnosis for the congenital epulis is adult granular cell tumour (myoblastoma). Both congenital and adult GCT have histopathologically identical cells. Early onset, unique location, pedunculated appearance, absence of pseudoepithe-

liomatous hyperplasia of the overlying squamous mucosa and no nerve bundles make the epulis easily differentiated from myoblastoma [2, 6, 7, 16, 17].

Other differential diagnoses for the congenital epulis include fibroma, rhabdomyoma, granuloma, Epstein's pearls, haemangioma, cephalocele, dermoid cyst, teratoma, melanocytic or pigmented neuroectodermal tumours, lymphatic malformations, schwannoma and heterotopic gastrointestinal cyst [2, 6, 16, 18].

Antenatal images of the lesion may be obtained by ultrasonography or magnetic resonance (MRI) mainly in the last weeks of pregnancy (after 25th week of intrauterine life), but the findings are not specific [2, 6]. In the cases of patients with large lesions, polyhydramnion caused by obstruction of the oral cavity was noted. In our case, the ultrasound performed in the first, second and third trimester (for the last time in the 32nd week of gestation) did not reveal any abnormality.

Surgical excision is curative and no recurrences even following incomplete excision have been reported [7, 19]. Cases of spontaneous regression of very small lesion have been reported [14, 15].

In conclusion, the congenital epulis is a rare, degenerative or reactive lesion of the oral cavity. The diagnosis is suspected clinically and the treatment by simple surgical removal has a curative effect.

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Address for correspondence

Monika Dzieniecka, MD
Department of Clinical Pathomorphology
Research Institute of the Polish Mother's Memorial Hospital
ul. Rzgowska 281/289
93-338 Łódź, Poland
tel. +48 42 271 12 85
e-mail: chicamonika@yahoo.es