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A case of unusual size



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Damiano Tambasco, Annalisa Seccia, Agostino Bruno, Antonio Seccia

Department of Plastic and Reconstructive Surgery, "Catholic University of the Sacred Heart," Rome, Italy

Congenital epulis of the newborn: difficult to diagnose but easy to treat. A case of unusual size

The epulis or giant cell granuloma is a benign tumor of the connective tissue of the gingival mucosa frequent in an advanced period of life but much more rare in its congenital form.

We present the case of a female newborn, otherwise healthy, presenting with a giant swelling protruding from her mouth and originating from upper left alveolar ridge. The size of the mass has created great anxiety in parents and pediatricians, however this clinical presentation suggested us a diagnosis of congenital epulis with a differential diagnosis of teratoma.

We perform surgical resection of the mass under general anesthesia, through diathermy. There wasn't blood loss and post-operative recovery was uneventful. A definite diagnosis of giant congenital epulis was disclosed by histopathological and immunohistochemical analysis.

Although it has not been yet clarified the etiology of this tumor and the role of hormonal influences on its appearance and development and despite have been reported cases of spontaneous regression, the treatment of choice is the early surgical excision.

Nevertheless, particularly in the small centers, due to the rarity and large size of presentation, the diagnosis is often delayed or wrong, exposing so the newborn parents to useless days of waiting and anxiety

In our opinion all pediatricians and surgeons should be aware of this malformation and of its simple, safe and effective surgical treatment, considering the excellent prognosis of this rare disease.

KEY WORDS: Congenita epulis, Early surgical excision, Giant cell granuloma

Introduction

The epulis or giant cell granuloma (so-called giant cell epulis) is a benign tumor of the connective tissue whose etiology and histogenesis are still debated, localized at the level of the gingival mucosa, which originates from tissues that form the hanger apparatus of the teeth (periodontium, alveolar bone, external alveolar periosteum)

and that, in adults, can cause resorption of the alveolar process and the erosion of the root of the teeth.

It's very frequent in an advanced period of life, predominantly in females and may arise from chronic inflammatory exuberant processes, following trauma or chronic local irritation.

A congenital form (congenital epulis of the newborn) is described, which is much rarer, sometimes diagnosed already during intrauterine life^{1,2}.

It appears as a submucosal swelling in the gingival, maximum size of 1-1.5 cm, without any involvement of the underlying bone. Although is a benign lesion, it may recur in 5-10% of cases.

Treatment consists of complete removal of the epulis and the implant base, and in adults, in the Extraction of Teeth Mobilized By The Tumor.

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Correspondence to: Damiano Tambasco, MD, University Hospital A. Gemelli, Largo A. Gemelli 8, 00168 Rome, Italy (e-mail: damtam@hotmail.it)

Case report

A female newborn by caesarean section, was referred to the department of Plastic Surgery immediately after birth for a large mass protruding from her mouth. She was otherwise healthy. On examination there was a giant vegetans swelling, oval appearance (with smooth, pink surface, crossed by fine telangiectasias) and regular, well-defined margins. Dimensions 3,5 x 2,5 x 1,5 cm with a mucous pedicle (base planting 1 x 0,5 cm) originating from upper left alveolar ridge (about 1 cm from the upper labial frenulum). It was interfering with feeding but at that moment the mass didn't create respiratory problems.

This clinical presentation suggested a diagnosis of congenital epulis in differential diagnosis with teratoma. To exclude the presence of other concomitant malformations in 2nd day a skull TC was performed confirming the gingival origin of the swelling and reporting moderate and uneven enhancement after administration of iodinated contrast. No erosive aspects of the maxillary superior or other intracranial abnormalities were detected.

It was therefore decided to perform early (3rd day) surgical resection of the mass under general anesthesia, through diathermy. There wasn't blood loss and postoperative recovery was uneventful.

The material was sent to histopathology and the Haematoxylin and Eosin (H&E) stain revealed sheets of polygonal or ovoid clusters of cells with medium-sized nucleus and nucleoli evident, granular cytoplasm, stroma richly vascularized with capillaries and venules, the absence of mitosis and occasional perivascular inflammation and proliferation of perivascular pericytes. Immunohistochemical analysis was negative for S-100 protein, keratin, desmin, CD31, CD34, CD68. A definite diagnosis of giant congenital epulis was disclosed.

Discussion

Since the first description of Neumann³ of 1871 have so far been described only about 200 cases of congenital epulis of the newborn⁴. and analyzing the scientific literature even recently, we find typically work on a



Fig. 1: Appearance of the lesion at 2nd day of life. Giant vegetans swelling, oval appearance (with smooth, pink surface, crossed by fine telangiectasias) and regular, well-defined margins. Dimensions 3,5 X 2,5 X 1,5 cm.



Fig. 2: Note the mucous pedicle (base planting 1 x 0,5 cm) originating from upper left alveolar ridge (about 1 cm from the upper labial frenulum).

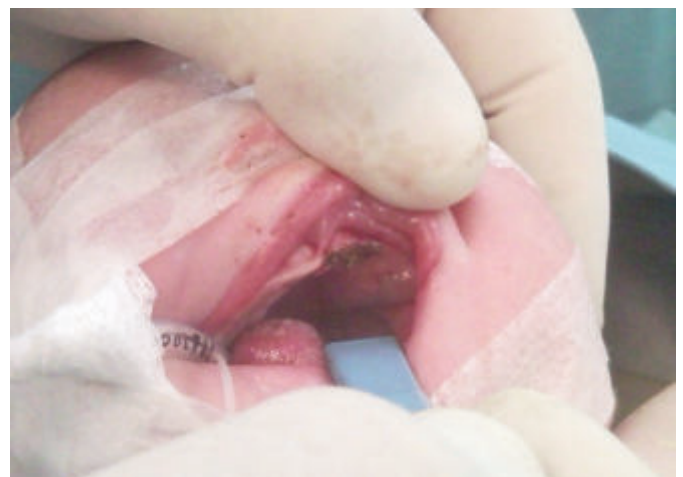


Fig. 3: Post-operative view (3rd day) after surgical resection of the mass under general anesthesia, through diathermy of the implant base.



Fig. 4: Post-operative view at 6 months. No recurrence or gingival distortion were detected.

single case report⁵⁻⁷, although there are also works that report ten cases⁸. These epidemiological data show the rarity of this benign neoplasm, the most frequently diagnosed at birth with a female-male ratio of 8:1^{9,10}. The localization is prevalent in the maxilla, unique, also rarely can have two or more localized lesions for example both at the margin of the alveolar maxilla and mandibular¹¹. In the case of large lesions may occur mechanical disturbances of nutrition and respiration. This lesion is well differentiated from many injuries that occur in childhood teratoma, dermoid cyst, angioma, lymphangioma, fibrosarcoma, leiomyoma, rhabdomyoma, etc.¹².

The tumor size may vary from a few millimeters up to as much as 9 cm and 90% of cases this lesion presents single, interesting, therefore, only the maxilla or mandible (a ratio between the two of 3:1)⁹.

Although it has not been yet clarified the etiology of this tumor (such as the myoblastic, odontogenic, neurogenic, histiocytic and endocrinologic origin^{13,14}) and the role of hormonal influences on its appearance and development and despite have been reported cases of spontaneous regression¹⁵, the treatment of choice is the early surgical excision.

Even in the presence of a non-radical resection of congenital forms, in literature cases of local recurrence have not been so far described.

The decades of experience achieved by our center in the early diagnosis of congenital malformations, due to the high number of births in our hospital and thus to a considerable number of infants for which pediatrics require our advice, led us to straightaway exclude malformations such as hemangiomas, lymphangiomas, fibromas, sarcomas (which sometimes come into differential diagnosis of this lesion¹⁶ and thus significantly speed up the diagnostic and therapeutic process. Nevertheless,

particularly in the small centers, due to the rarity and large size of presentation, the diagnosis is often delayed or wrong, exposing so the newborn parents to useless days of waiting and anxiety

In our opinion all pediatricians and surgeons should be aware of this malformation and of its simple, safe and effective of surgical treatment, considering the excellent prognosis of this rare disease.

RIASSUNTO

L'epulide o granuloma a cellule giganti è un tumore benigno del tessuto connettivo la cui etiologia ed istogenesi risultano ancora dibattute. Si localizza a livello della mucosa gengivale ed origina dai tessuti che formano l'apparato sospensore dei denti (parodonto, osso alveolare, periostio alveolare esterno) e che, nell'adulto, può determinare il riassorbimento del processo alveolare e l'erosione della radice dei denti. È molto frequente in un periodo avanzato della vita, prevalentemente nel sesso femminile, mentre è molto più rara nella sua forma congenita. In questo caso si presenta come una tumefazione sottomucosa, in sede gengivale delle dimensioni massime di 1-1,5 cm senza alcun interessamento dell'osso sottostante.

Presentiamo il caso di una neonata oltretermine da parto cesareo, in buone condizioni cliniche, giunta immediatamente dopo la nascita alla nostra osservazione per una enorme massa sporgente dalla sua bocca, ad origine dall'emiarcata dentaria superiore di sinistra. La massa interferiva con l'allattamento ma al momento non creava problemi respiratori.

Questo quadro clinico suggeriva una diagnosi di epulide gigante congenita in diagnosi differenziale con teratoma. È stata quindi posta indicazione ad asportazione chirurgica della massa in anestesia generale mediante diatermobisturi, con cauterizzazione della base di impianto. Non si è registrata alcuna perdita di sangue intraoperatoria e la degenza post-operatoria è risultata priva di complicanze. Il materiale è stato inviato per esame istopatologico e immunoistochimico che hanno confermato la diagnosi di epulide congenita gigante.

Nonostante non sia stata ancora chiarita l'eziopatogenesi di questo tumore e il ruolo degli influssi ormonali sulla sua comparsa e sul suo sviluppo e sebbene siano stati descritti casi di regressione spontanea, il trattamento d'elezione è l'escissione chirurgica precoce.

Tuttavia, specie nei piccoli centri, per la rarità e le grosse dimensioni di presentazione, la diagnosi è spesso ritardata o errata, esponendo così i genitori del neonato ad inutili giornate di attesa e apprensione.

È importante, pertanto, che tutti i pediatri e i chirurghi abbiano conoscenza di tale malformazione e siano consapevoli della semplicità e dell'efficacia del trattamento chirurgico, considerata la prognosi davvero eccellente di questa rara patologia.

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