

A rare case of congenital epulis of the newborn with multiple lesions

Um caso raro de epúlida congênita do neonato com lesões múltiplas

Abstract

Purpose: To describe a case of congenital epulis with multiple lesions, which was treated combining surgical resection of the largest tumor and monitoring of the smaller lesions.

Case description: A white female newborn showed a large (2.4x1.8x1.1cm), pinkish-red, nodular formation on the left side of the maxillary alveolar ridge, which was preventing mouth closure and causing feeding difficulties. Other small polyps were present on the left side of the mandibular ridge. Excisional biopsy under general anesthesia allowed for the complete removal of the irregular, bilobed maxillary tumor. The diagnostic hypothesis of congenital epulis of the newborn was confirmed based on both the anatomic details and immunohistochemical profile of the lesion. The smaller mandibular tumors were not resected; complete spontaneous remission occurred within 6 months. After surgery, the patient showed substantial clinical improvement, with complete functional and esthetic rehabilitation. No recurrence was observed.

Conclusion: Treatment allowed the reestablishment of vital functions and normal neonatal development. The surgical approach improved the infant's quality of life and restored the parents' confidence and emotional stability.

Key words: Gingival neoplasms; epulis, congenital; surgery

Resumo

Objetivo: Descrever um caso de epúlida congênita com lesões múltiplas, onde foi realizada a exérese total do tumor maior e o monitoramento dos tumores menores.

Descrição do caso: Um sujeito neonato do gênero feminino apresentava uma formação nodular grande (2,4x1,8x1,1cm), vermelha-rosada, no lado esquerdo do rebordo alveolar superior, a qual estava dificultando o fechamento bucal e causando dificuldades de alimentação. Outros pólipos menores estavam presentes no lado esquerdo do rebordo mandibular. A biópsia excisional sob anestesia geral permitiu a remoção completa do tumor superior, com aspecto irregular e bilobulado. A hipótese diagnóstica de epúlida congênita do recém-nascido foi confirmada com base nos detalhes anatômicos e no perfil imunoistoquímico da lesão. Os tumores mandibulares menores não foram excisionados; a remissão espontânea completa ocorreu dentro de seis meses. Após a cirurgia, a paciente mostrou melhora clínica significativa com completa reabilitação funcional e estética. Não houve recorrência nas visitas subsequentes.

Conclusão: O tratamento permitiu o restabelecimento das funções vitais e do desenvolvimento normal do neonato. A abordagem cirúrgica melhorou a qualidade de vida da criança e devolveu confiança e estabilidade emocional aos pais.

Palavras-chave: Neoplasias gengivais; epúlida congênita; cirurgia

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Received: March 12, 2008
Accepted: June 15, 2008

Introduction

Tumors of the oral cavity are not common among newborns (1). Congenital epulis of the newborn, also known as congenital granular cell tumor, is a rare gingival neoplasm that affects the alveolar ridge in the newly born (2-8). Its clinical presentation is usually a single tumor, but multiple tumors have also been reported. The typical location of congenital epulis is on the maxillary anterior alveolar ridge; involvement of the mandibular region occurs at a 2:1 ratio (1,5,9,10). Female newborns are more affected than male subjects (8:1) (1,2,5,6,8).

The lesion may be sessile or pedunculated, with gingival normal or red color, firm consistency and smooth, lobulated surface. Lesion size can vary, and its typical location on the alveolar ridge of newborns may restrict complete closure of the mouth (4,7,10). The tumor usually presents a benign behavior with no further growth after birth, showing spontaneous remission and absence of metastases (4,6,7,10). Surgical resection is indicated as early as possible when breastfeeding or respiratory problems are present. Prognosis is favorable, and no relapses have been reported (1,6,10).

This article describes the diagnosis and treatment of a rare case of congenital epulis in a newborn consisting of multiple lesions. Histogenesis, clinical and histopathological features, and treatment options are reviewed and discussed.

Case description

A white female newborn was brought to the Oral and Maxillofacial Surgery and Traumatology Clinics at Hospital Santa Cruz, Santa Cruz do Sul, RS, Brazil, with a large, pinkish-red, nodular formation on the left side of the maxillary alveolar ridge. The lesion measured approximately 2.4x1.8x1.1cm and was preventing mouth closure. Physical examination revealed the presence of other small polyps on the left side of the mandibular alveolar ridge (Fig. 1A). No other congenital clinical lesions were observed. On the third day of life, after continuous monitoring and

observation by the oral and maxillofacial surgeon, feeding difficulties (suction/breastfeeding) were detected; respiratory function and airway patency were normal. Although the clinical features and the natural history of the lesion were characteristic of a benign process, a surgical intervention was carried out under general anesthesia (Fig. 1B) aiming to reestablish breastfeeding.

Excisional biopsy allowed for the complete removal of the upper tumor (Fig. 1C). Macroscopic analysis revealed an irregular, bilobed appearance. The specimen was fixed in 10% formalin solution and sent for histopathological analysis. The smaller tumors located on the mandible were not resected; complete spontaneous regression occurred within 6 months. After surgery, the patient showed significant clinical improvement, with overall functional and esthetic rehabilitation as a result of maxillary alveolar ridge reconstruction. Hospital discharge took place 3 days after surgery. The surgical approach in this case improved the infant's quality of life and restored the parents' confidence and emotional stability. No recurrence was observed in the regular follow-up appointments.

Histopathological findings

Initial histopathological results were not conclusive due to the non-specific histological profile of the lesion shown as granular cells with eosinophilic cytoplasm, which is similar to other tumors. Differential diagnosis was achieved by assessing immunoreactivity to protein S100, through an antigen recovery system using microwave irradiation and ethylene diamine tetra-acetic acid (EDTA) (1mmol, pH 8.0). A streptavidin-biotin-peroxidase complex was used for amplification, and staining was carried out with hydrogen peroxide and diaminobenzidine. Immunohistochemical results were negative for protein S100. The diagnostic hypothesis of congenital epulis of the newborn was therefore confirmed based on both anatomic details and immunohistochemical profile of the lesion (Fig. 1D).

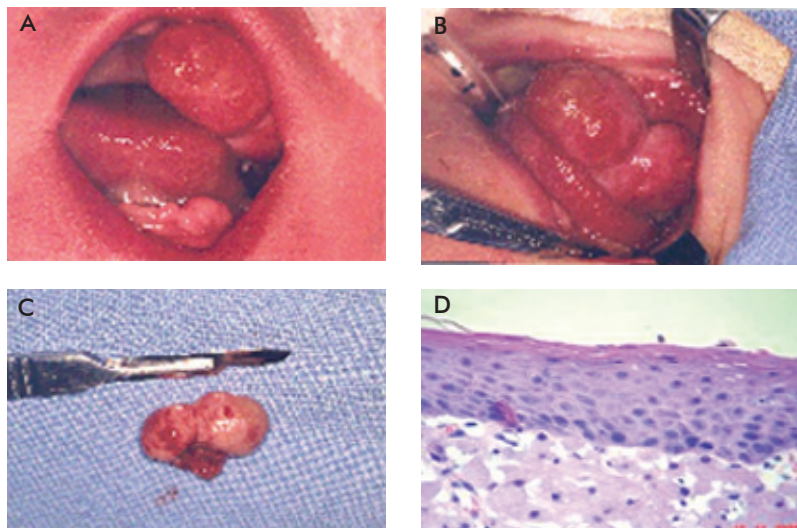


Fig. 1. (A) Largest tumor measuring approximately 2cm, located on the maxillary alveolar ridge; smaller tumors located on the mandibular ridge. (B) Close-up view showing tumor size (note bilobed appearance). (C) Specimen removed and sent for histopathological analysis. (D) Microphotography showing evidence of condensed cells, with eosinophilic granular cytoplasm and covered by a layer of flat epithelium (HE 200X).

Discussion

This paper describes a benign case of congenital epulis of the newborn present at birth in a female child. The predominance of female patients has been described in the literature (1,2,5,6,8,10-12). The typical location of congenital epulis is on the maxillary ridge, particularly in the incisor and canine area (5,7,8,11); the maxilla is affected approximately twice as often as the mandible (4,11,12). The largest tumor in our case measured approximately 2cm, but lesions can vary from a few millimeters up to 9cm in diameter (4,10,12). According to Lapid et al. (8), in most cases the tumor is 2cm or smaller, but tumors of more than 7.5cm may occur and cover the entire oral cavity.

Chattopadhyay et al. (1) have emphasized the possibility of detecting the tumor during pregnancy, via ultrasound. In the present case, however, no previous diagnosis of the tumor had been made so as to allow for any prenatal intervention. A relevant factor was the presence of multiple tumors in the maxilla and mandible. Zarbo et al. (2), Zuker and Buenechea (4) and Loyola et al. (7) have pointed out that multiple tumors, with simultaneous involvement of both maxilla and mandible, are very rare and account for only 10% of the cases.

Many theories have been suggested to explain the histogenesis of congenital epulis of the newborn, including gingival endothelial, mesenchymal, mioblastic, odontogenic, neurogenic, fibroblastic, and histiocytic origins (4,5,7,8,12); however, no consensus has been reached so far. In view of the intrauterine formation of tumors and the higher incidence observed in female newborns, some authors have suggested a possible influence of fetal ovarian hormones in tumor formation, and have recommended the performance of immunohistochemical tests to assess the influence of estrogen and progesterone receptors in the process (5,8,11,12).

Histopathologically, congenital epulis of the newborn is characterized by round, large cells with abundant eosinophilic granular cytoplasm and round to oval, slightly basophilic nuclei. In older tumors, these cells may become elongated and separated by fibrous conjunctive tissue (1). Because these tumors rarely affect the tongue, some authors (7,9) prefer to use the term congenital granular cell lesion. Others use gingival granular cell tumor of the newborn, because of microscopic similarities; however, this terminology should be avoided (10). Despite the histological similarities, these entities have considerably different clinical, histochemical, epidemiological, and pathological features, which justify their classification as distinct lesions (3-5,7,8,11).

Differential diagnosis should be based on the predominance of female patients, tumor location on the anterior maxillary

region, presence at birth, and absence of growth potential, and the possibility of spontaneous remission should be taken into consideration (6). In the present case, there was no evidence of growth after birth, as seen in previous reports (5,10). Histologically, differential diagnosis should rely on absence of pseudoepitheliomatous hyperplasia of the squamous epithelium (5), absence of protein S100 (3,9), and on distinct histogenesis (probably of mesenchymal origin). The immunohistochemical technique is a useful tool in the histogenetic analysis of lesions, and can be used to assess protein S100 immunoreactivity, which is positive in granular cell tumors but absent in cases of congenital epulis of the newborn (2,3,8,11). Finally, differential diagnosis should also be made in relation to other granular cell tumors, hemangiomas, lymphangiomas, fibromas, granulomas, rhabdomyomas, xanthomas, gingival cysts, and heterotopic gastrointestinal cysts (4,10).

Despite its characteristic clinical presentation (a pedunculated gingival tumor emerging from the alveolar ridge), correct preoperative diagnosis is rarely achieved, probably as a result of the few number of cases and absence of awareness by professionals (4,8,10). Nevertheless, precise and early diagnosis of these lesions is of paramount importance to avoid interference with vital functions, such as respiration and feeding, normal development of the jaw and adjacent teeth, and even death (1).

Surgical excision is not recommended to all cases in view of the essentially benign and innocuous behavior of the tumor (spontaneous remission and absence of metastases or relapses) and due to the possibility of affecting unerupted teeth (4,7,8,10). However, trauma associated with feeding or sucking fingers may cause inflammatory reactions and edema and require radical surgical resection of the lesion. Surgery also is indicated in cases presenting large and/or multiple tumors causing respiratory obstructions and/or feeding difficulties, with excellent prognosis (1,4,7,10,12). In our case, surgical excision of the tumor was carried out to restore suction/breastfeeding.

Conclusions

The clinical aspects of the described case, namely presentation of multiple lesions, similarity with other tumors, and possible compromise of vital functions, show the need for implementing a complete examination of the oral cavity of newborns as a routine procedure. Newborns with a diagnosis of congenital epulis of the newborn should be immediately subjected to the most adequate treatment (monitoring or surgical excision) to restore vital functions and improve quality of life.

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