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PRIKAZ SLUČAJA
CASE REPORT

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Rijedak slučaj kongenitalnog fibroznog epulisa kod djeteta

A Rare Case of Congenital Fibrous Epulis in an Infant

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Sažetak
Novorođenče s kongenitalnim epulisom može biti težak prizor kako za roditelje tako i za zdravstveno osoblje uključeno u novorođenačku skrb. Kod djece takve lezije mogu varirati u veličini, a i rizične su zbog opstrukcije dišnih puteva i poteškoća kod hranjenja. Stomatolozi trebaju poznavati takve promjene, no moraju se konzultirati s pedijatrima, anesteziolozima i neonatolozima zbog informacija koje o liječenju trebaju dati roditeljima. Osim toga, kongenitalni epulis važan je i zbog diferencijalne dijagnostike agresivnijih tvorbi u dječjoj dobi. Pregledavajući literaturu na engleskom jeziku pronašli smo samo tri slučaja takve lezije. U ovom radu opisan je slučaj 4-mjesečnog dječaka s neobičnom urođenom oteklinom u prednjem dijelu alveolarnog grebena donje čeljusti, koja je histološki odgovarala kongenitalnom fibroznom epulisu.

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Ključne riječi
gingiva, neoplazme; dojenče

Uvod

Kongenitalni epulis (KE) kod novorođenčeta rijetka je lezija koju je prvi opisao Neumann godine 1871. (1). Od tada se zna za manje od 200 slučajeva, a ponajprije su predstavljeni u literaturi iz područja patologije, stomatologije i otorinolaringologije (2-4). Ta tvorba, obično prisutna od rođenja, višestruka je u 10 posto slučajeva i ima izrazitu predilekciju kod žena te je često locirana u prednjem dijelu alveolarnog grebena gornje čeljusti (3-4).

U diferencijalnoj dijagnostici širok je raspon lezija: tumor granularnih stanica, hemangiom, limfangiom, fibrom, granulom, rabdomiom, ksantom,

Introduction

Congenital epulis (CE) of the newborn is an uncommon lesion described for the first time in 1871 by Neumann (1). Since then, fewer than 200 cases have been described in literature, primarily in the pathologic, dental and otolaryngologic literature (2-4). This lesion, which usually presents at birth, is multiple in 10% of the cases and has a distinct predilection for females, frequently located on the anterior maxillary alveolar ridge (3,4).

A wide range of lesions such as granular cell tumors, hemangioma, lymphangioma, fibroma, granuloma, rhabdomyoma, xanthoma, gingival cysts

gingivalne i heterotopične gastrointestinalne ciste te melanotički neuroektodermalni tumor u dječjoj dobi (5). Kongenitalni fibrozni epulis (KFE) histološki je naziv za kongenitalni epulis i smatra se hamartomoznom tvorbom te, mikroskopski, nema granularnih stanica nužnih za dijagnozu tipičnoga kongenitalnog epulisa. U ovom prikazu opisana je takva rijetka urođena lezija kod 4-mjesečnog dječaka u prednjem dijelu alveolarnog grebena donje čeljusti. Izliječena je bila kirurški - ekscizijom uz lokalnu anesteziju.

Prikaz slučaja

Četveromjesečno zdravo muško dijete opći je stomatolog uputio na Odjel oralne i maksilofacijalne kirurgije Stomatološke klinike Sveučilišta u Chitradurgi zbog neobične urođene otekline u prednjem dijelu alveolarnog grebena donje čeljusti. Tvorba na početku nije smetala pri dojenju, ali se kasnije povećala i otežavala normalno hranjenje. Porođaj je inače protekao uredno i u terminu. U majčinoj i očevoj anamnezi nije bilo ničega sličnoga. Limfni čvorovi glave i vrata nisu bili palpabilni. Medicinska anamneza bila je bez osobitosti. Tvorba je bila veličine 2,0x1,0x1,5 cm i iste boje kao okolna nepromijenjena oralna sluznica, Slika 1. Lezija nije bila peteljkaste građe. Anamneza i klinička obilježja sugerirala su da je dobroćudna. Bila je postavljena klinička dijagnoza kongenitalnog epulisa te je kirurški uklonjen pod lokalnom anestezijom 2-postotnim Xylocainom. Krvarenje je bilo minimalno i cijeljenje je proteklo bez komplikacija. Uzorak je nakon zahvata bio fiksiran u 10-postotnom formalinu i poslan na patohistološku analizu, Slika 2.

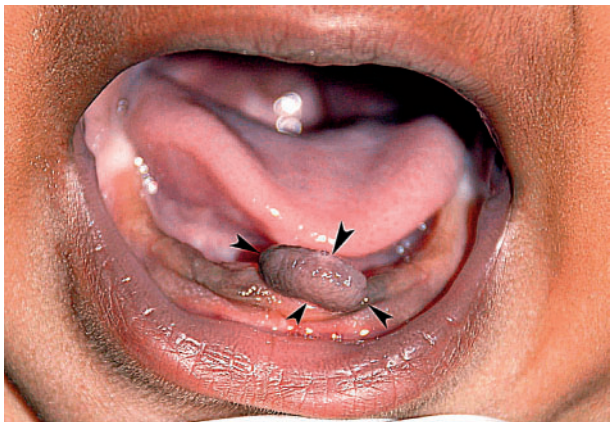
Mikroskopski pregled pokazao je dobro ograničenu tvorbu prekrivenu višeslojnim pločastim epitelom sa zupcima poput pile. Vezivno tkivo karakterizirali su gusto pakirani nepravilni snopovi kolagenih vlakana okruženi plosnatim, uglastim, vretenastim stanicama s dugim tankim produžecima te malo upalnih stanica, Slika 3. Vidjelo se i nekoliko krvnih žila, Slika 4. Uzorak tkiva bio je obojen specijalnom bojom prema von Giesonu, čime je potvrđena njegova kolagena građa, Slika 5. Prema kliničkoj slici i patohistološkom nalazu, lezija je bila dijagnosticirana kao četvrti slučaj kongenitalnoga fibroznog epulisa.

and heterotopic gastrointestinal cysts, melanotic neuroectodermal tumor of infancy may be included in the differential diagnosis (5). Congenital fibrous epulis (CFE) is a histological variant of congenital epulis which is considered as a hamartomatous lesion and microscopically has no granular cells needed for the diagnosis of typical congenital epulis. This case report describes such a rare lesion occurring in a four month old male child seen in anterior region of mandibular alveolar ridge since birth and was treated by surgical excision under local anesthesia.

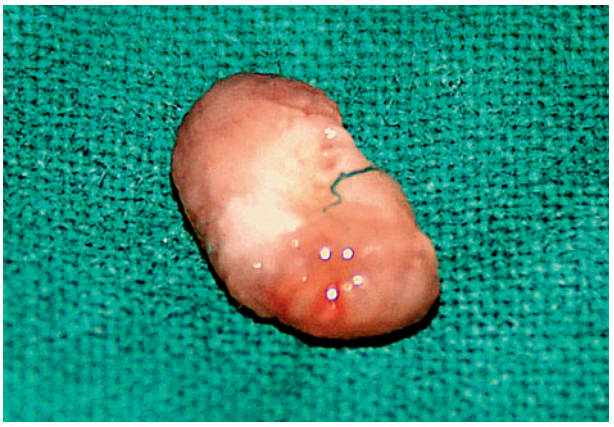
Case report

A four month old healthy male child was referred to the Department of Oral and Maxillofacial Surgery, S.J.M. Dental College and Hospital, Chitradurga, by a general dentist, for an unusual swelling over the mandibular anterior alveolar region noted since birth. It was not interfering with normal breast feeding in the initial stage but later posed difficulty in normal breast feeding as it increased to present size. Delivery of the baby had otherwise been normal at full term. Maternal and paternal histories were non contributory. There were no palpable lymph nodes in the head and neck. Medical history was also not relevant. The lesion measured about 2.0x1.0x1.5cm and was same color as that of adjacent normal oral mucosa (Figure 1). The lesion was non-pedunculated in nature. The history and clinical features of the lesion suggested its benign nature. The clinical diagnosis of congenital epulis was made and surgically excised under local anesthesia using 2% Xylocaine. Bleeding was minimal and the lesion healed with no complications and the specimen (Figure 2) was fixed in 10% neutral buffered formalin and sent for histopathological examination.

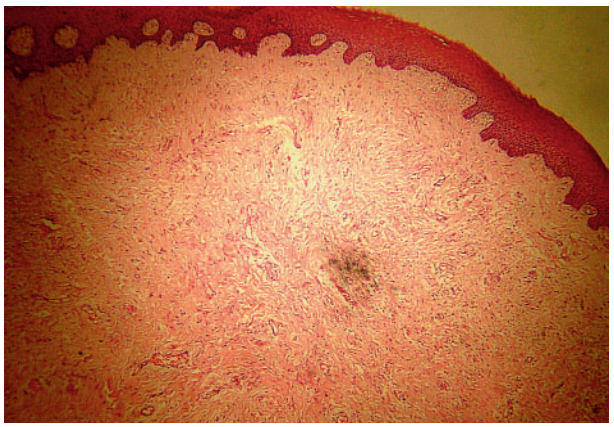
Microscopic examination revealed well circumscribed lesion, covered with keratinized stratified squamous epithelium with retepegs, underlying connective tissue showing and densely packed irregular bundles of collagen fibers surrounded by flattened, angulated, spindle cells with long slender processes and also very few inflammatory cells (Figure 3). Few blood vessels were also seen in the connective tissue (Figure 4). The tissue section was subjected to von Gieson special stain, which confirmed the collagen nature of the tissue (Figure 5). Based on clinical and histopathological findings, the lesion was diagnosed as fourth case of *congenital fibrous epulis*.



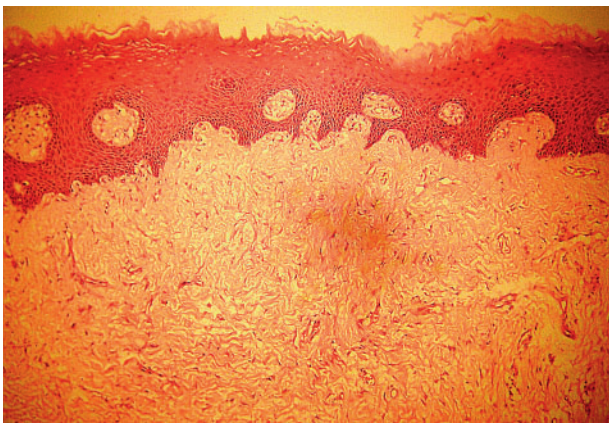
Slika 1. Fotografija prirođene lezije u prednjem dijelu donje čeljusti.
Figure 1 Photograph of the lesion noted in the mandibular anterior area since birth.



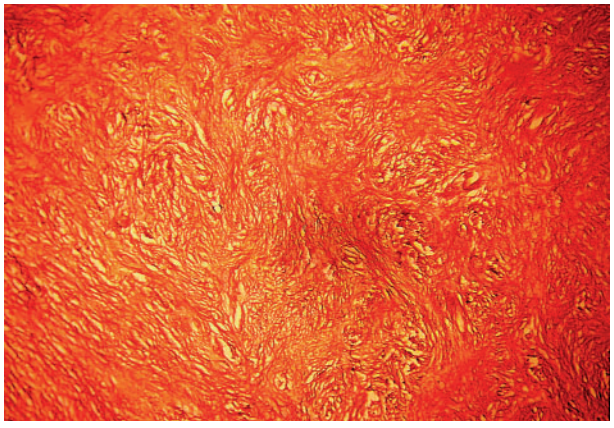
Slika 2. Fotografija kirurški uklonjenog uzorka veličine 2.0 x 1.0 x 1.5 cm.
Figure 2 Photograph of the surgical specimen measuring about 2.0 x 1.0 x 1.5 cms.



Slika 3. Mikrofotografija lezije prikazuje gusto pakirane nepravilne snopove kolagenih vlakana okružene plosnatim, uglastim, vretenastim stanicama i prekrivene keratiniziranim višeslojnim pločastim epitelom (hematoksilin-eozin, izvorno povećanje 40X).
Figure 3 Photomicrograph of the lesion showing densely packed irregular bundles of collagen fibers surrounded by flattened, angulated, spindle cells covered by overlying keratinized stratified squamous epithelium (Hematoxylin & Eosin stain, original magnification 40X).



Slika 4. Mikrofotografija prikazuje nekoliko krvnih žila sa snopovima kolagenih vlakana u stromi vezivnog tkiva (hematoksilin-eozin, izvorno povećanje 100X).
Figure 4 Photomicrograph showing few blood vessels with bundles of collagen fibres in a bland connective tissue stroma (Hematoxylin & Eosin stain, original magnification 100X).



Slika 5. Mikrofotografija prikazuje crveno obojena kolagena vlakna u vaskulariziranoj stromi (bojenje po von Giesonu, izvorno povećanje 100X).
Figure 5 Photomicrograph showing red color stained collagen fibers in a vascular stroma (von Gieson stain, original magnification 100X).

Rasprava

Riječ „epulis“ dolazi od grčke riječi i znači „na gingivi“ ili „čir gingive“. Prvi je tu tvorbu opisao Neumann (1) godine 1871. Taj je tumor rijedak i dosad je u literaturi opisano samo 167 slučajeva prema preglednom radu Zukera i Buenecha iz 1993. (5). Kongenitalni epulis ima spolnu predilekciju u omjeru 8:1 u korist žena i češće je lociran na prednjem dijelu gornjega čeljusnog grebena (omjer 3:1), (2-6). Naš opisani slučaj je neobičan, jer je epulis nađen u prednjem dijelu donje čeljusti kod muškog pacijenta.

Kongenitalni fibrozni epulis smatra se kliničkom i patohistološkom varijantom kongenitalnog epulisa i vrlo je rijedak u dječjoj dobi. Prvi su ga opisali Majid i suradnici godine 1986 (7). Od tada se zna tek za nekoliko slučajeva u azijskoj populaciji. KFE se smatra hamartomatoznom lezijom i - prema mišljenju Inana i njegovih suradnika - nije ni kongenitalni ni fibrozni epulis, nego zaseban entitet (7). Prema našim spoznajama i prema pregledu svjetske literature, opisana su samo četiri slučaja, uključujući i ovaj (7,8). KFE se javlja kod dječaka s prosječnim promjerom od približno 9 mm i manji je od KE-a. U azijskoj populaciji opisana su tri slučaja KFE-a - dva se odnose na dječake, a jedan na djevojčicu (8). U ovom prikazu slična je lezija opisana kod četveromjesečnoga muškog djeteta. Histološki je bila građena od nepravilnih, nasumce poredanih snopova kolagenih vlakana među kojima su nađeni vretenasti i uglasti fibroblasti s malo ili bez infiltracije upalnih stanica.

Za taj se tumor, prije kirurške obrade, često postavlja kriva dijagnoza zato što je rijedak, pa ga kliničari ne prepoznaju odmah. U diferencijalnu dijagnozu mogu se uključiti neuroektodermalni tumori u dječjoj dobi, gingivalna cista, hemangiom, fibrom i granulom (5).

Terminologija vezana za tu tvorbu dosta zbunjuje zbog nepoznanica glede histogeneze tih tumora, a histološka slika slična je mioblastomu granularnih stanica koji se javlja kod odraslih na drugim lokacijama u usnoj šupljini (9). Histogeneza nije poznata u cijelosti, ali predloženo je nekoliko hipoteza, uključujući podrijetlo od epitelnih, nediferenciranih mezenhimnih stanica, pericita, fibroblasta, glatkih mišićnih stanica i stanica živčanog sustava (10-15).

Uobičajeno liječenje kongenitalnog epulisa jest kirurško uklanjanje u razini alveola. Dubljom resekcijom mogli bi se ozlijediti neizniknuli zubi (16). Kod te lezije vrlo je važna točna i rana dijagnoza kako bi se izbjegla opstrukcija dišnih puteva i smetnje

Discussion

The word “Epulis” is derived from the Greek word which means “on the gum” or “gum boil”. The original description of the lesion dated back in 1871 was given by Newmann (1). This tumor is rare and only 167 cases have been reported in the literature as reviewed by Zuker and Buenecha in 1993 (5). Congenital epulis has a female predilection with ratio of 8:1, and is more frequently located on the anterior maxillary alveolar ridge with a ratio of 3: 1 (2-6). Our case was unusual as it occurred in the mandibular anterior region in a male patient.

Congenital fibrous epulis is considered to be a clinical and histopathologic variety of congenital epulis and is an extremely rare tumor of infancy, first described by Majid et al in 1986 (7). Since then only very few cases have been reported occurring in Asian population. CFE is considered as a hamartomatous lesion and according to Inan M et al, it is neither congenital epulis nor fibrous epulis and considered as distinct entity (7). To the best of our knowledge and worldwide literature review showed only four cases, including our present case (7,8). On the contrary, CFE occurs in male infants with an average diameter of approximately 9 mm and smaller than CE. A total of 3 cases of CFE occurred in Asian population with two cases in male infants and one case in female infant[8]. In this present report, a similar lesion in a four month old male infant is described and histologically the lesion showed irregular bundles of collagen fibers arranged in a haphazard manner interspersed by spindle and angulated fibroblasts with little or no inflammatory cell infiltration.

The tumor is often misdiagnosed before surgery because of its rarity and lack of awareness by clinicians. Neuroectodermal tumor of infancy, gingival cyst, hemangioma, fibroma, and granuloma may be included in the differential diagnosis of the lesions of oral cavity (5).

The terminology concerning this condition has been rather confused because of uncertainty regarding the histogenesis of these tumors and similar histological appearance to granular cell myoblastoma occurring in adults at other intraoral sites[9]. The histogenesis is still uncertain, but several theories, including origin from epithelial, undifferentiated mesenchymal cells, pericytes, fibroblasts, smooth muscle cells and nerve-related cells have been proposed (10-15).

The favored treatment for congenital epulis remains surgical removal at the level of the alveolus. Deeper resections may likely damage the underly-

pri hranjenju te osigurao normalan razvoj čeljusti i okolnih zuba. U našem je slučaju bila obavljena kirurška ekscizija tumora pod lokalnom anestezijom, što je omogućilo ponovno dojenje djeteta.

Zaključili bismo da detaljni pregled usne šupljine novorođenčeta treba biti rutinski postupak. Ako se dijagnosticira kongenitalni epulis, potrebno je odmah početi s liječenjem kako bi se uspostavile vitalne funkcije i poboljšala kvaliteta djetetova života.

ing unerupted teeth (16). However, precise and early diagnosis of these lesions is of supreme importance to avoid any obstruction with respiration and feeding, normal development of the jaw and adjacent teeth. In our case, surgical excision of the tumor was carried out under local anesthesia to restore breastfeeding.

To conclude, complete examination of the oral cavity of newborns as a routine procedure is needed. If diagnosed with a congenital epulis, newborns should be immediately treated to restore vital functions and improve quality of life.

Abstract
A newborn infant with congenital epulis can be the most difficult sight for both parents as well as health care professionals involved in neonatal care. These lesions of the infant can be of varying sizes, posing a risk of airway obstruction or interfering with feeding. Dentists should have the knowledge about these lesions as they may necessitate consultation with other practitioners such as pediatricians, anesthetist, and neonatologist and provide information to parents regarding the treatment of these lesions. Furthermore, congenital epulis is an important lesion in the differential diagnosis of more aggressive lesions seen in early life. English literature review showed only three cases of congenital fibrous epulis. In this present report, a four month old male infant is described with an unusual swelling over mandibular anterior alveolar region since birth and histologically the lesion showed features of congenital fibrous epulis.

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Gingival Neoplasms; Infant, Newborn

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