Hereditary Gingival Fibromatosis: a Case Report with Seven-Year Follow-up

**Introduction**

Hereditary gingival fibromatosis (HGF) is a rare disease in both autosomal dominant and recessive types and has a frequency of 1:175,000 (1). Clinically, it is characterized by benign gingival enlargement with normal color and firm consistency and non-hemorrhagic symptomatic illness (2). Although the gingival enlargement does not directly affect the alveolar bone, the gingival swelling may add to the bacterial plaque accumulation, inducing gingivitis, periodontitis, bone resorption and halitosis (1). Histologically, the connective tissue has increased collagen and few fibroblasts and the epithelial tissue presents hyperplasia areas and prominent buds. Despite these characteristics, the histologic features of HGF are not exactly specific, and the definitive diagnosis should be based on family history and clinical findings (1). Some of the clinical complications of HGF can be excessive gingival growth, which results in pseudo pocketing and periodontal disease due to poor oral hygiene, diastemas, delayed...
čajevima hiperplazija gingive može ograničiti kretanje jezika, orežati govor i činiti potreščiće pri živakanju (1, 2–7).

Među autorima istraživanja nema suglasja o učinkovitosti liječenja NGF-a (6), no svi se slažu da postoji rizik iz njega-
va ponovnog pojavljivanja što treba izbjegavati (7). Ovisno o rastu, liječenje uključuje izrezivanje povećane uobiča-
jenim kirurškim zahvatom, primjenjuju se i elektrokirurgija ili laser, apikalno se pomiče režanj vade se svi zubi i smanju-
je alveolaran kost kako bi se spriječilo ponavljanje bolesti (1, 2). Uspeh dentalne terapije ovisi o raspoređivanju preventiv-
nih kontrolnih pregleda kako se bolest ne bi ponovila (6, 7).

Ovom je radu svrha opisati način postavljanja dijagozne
tačne liječenja i praćenja nasljedne fibromatoze gingive na
temelju prikaza slučaja s povećanom gingivom kod pacijen-
tice na antikonvulzivnoj terapiji, što je povezano s upalnim
čimbenikom, tj. visokom razinom plaka. Uz to, ovaj prikaz
slučaja ističe način liječenja samo uz lokalnu kontrolu plaka
bez ikakvih promjena u primjeni sustavnih lijekova.

Prikaz slučaja

Nepokretna 20-godišnja pacijentica, koja je patila od mi-
krocefalije, epilepsijske i mentalne retardacije, upućena je u naš
centar u siječnju 2009. godine zbog, kako je rekla njezina
majka, boli u čeljusti. Tijekom anamneze istaknula je da je i
njezin pokojni sin patio od istih simptoma, tj. rasta gingive.
Pacijentica je bila na terapiji na 200 mg karbamazepina (3x/
dan), 160 mg fenobarbitala (1x/dan), uzmila je i 5 mg ci-
klobenzaprin-hidroklorida (1x/dan), 50 mg ranitidina (1x/
dan), željezo (40 kapi/ dan), kalij (10 ml 1x/dan) i minerale
(10 ml 1x/dan). Bila je u stanju od 50 bodova prema Kar-
nofskyjevoj ljestvici statusa izvedbe (8), jela je samo meku
hranu i težila 14 kilograma. Važno je napomenuti da je pacijen-
tica imala grand mal napadae od osam do deset puta na
dan, a kadkad je zbog njihove težine bila potrebna i hospitali-
zacija. Kad je došla u hitnu službu napadaji su bili u početnoj
fazi zato što je liječnik prilagodio dozu lijeka. Nakon nekoli-
ko promjena doza, konvulvizije su stabilizirane odgovarajućom
i redovitom uporabom antikonvulziva.

Intraoralni pregled pokazao je obimnu hiperplaziju gingi-
ve povezana s visokim indeksom plaka (3,16 prema indeksu
IHOS-a), zatim gingivalni kamenac, lažne parodontne dže-
ve asocijirane s visokim indeksom plaka (3,16 prema indeksu
1, 2). Uspjeh dentalne terapije ovisi o raspoređivanju preventive
nih kontrolnih pregleda kako se bolest ne bi ponovila (6, 7).

Case report

A bedridden, 20-year-old female patient, who suffered from
microcephaly, epilepsy, and mental retardation, was re-
ferred to our center in January 2009 with a chief complaint of
‘pain in the jaws’, according to her mother. In the course of tak-
ing medical history she reported another case of gum growth
in her brother, who had died. The patient was on carba-
mazepine 200 mg (3 times a day), phenobarbital 100 mg (1 time a
day), cyclobenzaprine hydrochloride 5 mg (1 time a day), ra-
nitidine 50 mg (1 time a day), iron (40 drops a day), calcium
(10 ml 1 time a day) and minerals (10 ml 1 time a day). At that
time, she had scored 50 points on the Karnofsky Performance
Status Scale (8), ate only a soft diet and weighed 14 kilograms.
It is important to note that the patient had grand mal seizures
about eight to ten times a day and sometimes there was need
for hospitalization due to the severity of her seizures. At the
time she went to our service, the seizures were in the initial
control phase, since the doctor was adjusting the medication
dose. After several alterations of dosage, the seizures stabilized
with appropriate and regular use of anticonvulsant medication.

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The intraoral examination revealed severe gingival hyper-
plasia associated with a high plaque index (3.16, according
to IHOS index) - gingival calculus, false periodontal pock-
ets, three decayed teeth and four teeth with active white spots
(Figures 1, 2 and 3). Dental treatment was performed under
general anesthesia because there were several dental proce-
dures to be carried out and the patient’s general health status
was classified as ASA II.

We performed the basic periodontal treatment and com-
posite resin restorations, sealing the active white spots using
modified, atrumatic, restorative treatments, and we surgically
removed the third molars. Regarding the gum, specifically,
after the marking the gingival pockets, we removed the
excessive tissue by means of internal bevel gingivectomy on
some sections of buccal and palatal-lingual surfaces. The inci-
tooth eruption, and facial disfigurement due to lip protru-
sion. In addition, in the most severe cases, gingival hyperpla-
sia can result in limited tongue movement, speech impedi-
ments and difficulty chewing (1, 2–7). There is no consensus
among authors about the efficacy of HGF treatment (6) and
the recurrence risk is real and needs to be avoided (7). De-
pending on the severity of the of the growth, treatment in-
volves the excision of the enlarged gingival tissues, using con-
ventional surgery, electrosurgery, an apically positioned flap,
or lasers through to the extraction of all teeth and reduction
of the alveolar bone to prevent recurrence (1, 2). The success
doing treatment depends on scheduling a return appoint-
ment as a preventative for recurring disease (6, 7).

This paper aims to review the diagnosis, treatment, and
follow-up of heredetary gingival fibromatosis by the presenta-
tion of a case report characterized by the exacerbated gingival
enlargement through the use of anticonvulsant medication,
associated with the inflammatory factor, i.e., high levels of
plaque. In addition, this case report presents the local treat-
ment plaque control as a differential without any interference
with systemic medication.
Slik 1. Intraoralni pogled na gornju desnu stranu NGF-a prije kirurškog liječenja u općoj anesteziji

Figure 1. Intraoral view of the maxillary right side, showing HGF before surgical dental treatment under general anesthesia.

Slik 2. Intraoralni pogled na gornju lijevu stranu NGF-a prije kirurškog liječenja u općoj anesteziji

Figure 2. Intraoral view of the maxillary left side, showing HGF before surgical dental treatment under general anesthesia.

Slik 3. Pogled sprijeda na mandibularne sjekutice prije kirurškog liječenja u općoj anesteziji

Figure 3. A front view of mandibular incisors before surgical dental treatment under general anesthesia.

Slik 4. A: Neposredno poslije operacije. B: Petnaest dana poslije operacije

Figure 4. A: Immediately after surgery. B: Fifteen days after surgery.

Slik 5. Kontrola nakon jedne godine

Figure 5. One-year follow-up.

Slik 6. Intraoralni prikaz gornje desne strane sedam godina poslije kirurškog liječenja u općoj anesteziji

Figure 6. An intraoral view of the maxillary right side seven years after dental treatment under general anesthesia.

Slik 7. Intraoralni pogled na gornju lijevu stranu sedam godina poslije kirurškog liječenja u općoj anesteziji

Figure 7. An intraoral view of the maxillary left side seven years after dental treatment under general anesthesia.

Slik 8. Pacijent koji je pod nadzorom nema karijes ili parodontnu bolest

Figure 8. The patient undergoing monitoring without evidence of caries or periodontal disease.
Hereditary Gingival Fibromatosis

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Discussion

Hereditary gingival fibromatosis is a rare condition of gingival tissues that can be associated with several other clinical manifestations and feature some syndromes (4, 5). This growth is slow and progressive and may be exacerbated by the use of drugs. It can occur in isolation or may be associated with syndromes, such as Zimmermann-Laband, Rutherford, and Ramon syndromes (3, 7). It is important to note that oral health of the patient, including the gums and teeth, was recovered without interfering with her medication, as the doctor took a long time to choose the drug and its dose to control convulsive seizures. For this reason, our team evaluated the three main causes of gingival hyperplasia, which were genetics, the use of anticonvulsant medication and the presence of plaque, i.e., the inflammatory factor. It is noteworthy that in this case, there was no intervention done concerning the genetic and medical factors. On the other hand, the control of the inflammatory factor, through periodic visits to the dentist and the family’s participation at home, has been responsible for the absence of the recurrence of gingival hyperplasia to date. This fact leads us to rule out the hypothesis of medical gingival hyperplasia. In this context, this situation underscores the importance of plaque control to prevent new and recurrent diseases, especially manifestations of rare and severe pathologies, such as HGF, in which the recurrence rate after surgery is relatively high (5). Therefore, despite the fact that HGF is not caused by plaque increase; it can be exacerbated by it (1).

It is worth noting that due to the patient’s neurological impairment and disabled protective reflexes, such as coughing, the diet recommended by her medical team had a “doughy consistency”. In addition, her oral health care had been relatively neglected in the context of general health framework that she presented. Such a situation is in accordance with Escribano-Hernández et al (9), who alleged that mentally disabled patients often have their oral health care neglected. They have further stated that the rate of dental carries and periodontal disease is substantially increased in this population because of a pureed, high carbohydrate diet and physiological limitations that prevent adequate teeth cleaning due to impaired muscle function.

Actually, the use of dental general anesthesia has been indicated for handicapped patients that do not have good comprehension as well as children infants and pre-school children who do not have good behavior and patients with excessive fear associated with several dental needs (9, 10). However, it is important to highlight that dental general anesthesia had good results only when associated with a postoperative prevention program (11). We endorse these results and believe that clinical success over seven years of follow-up has been achieved due to the correct diagnosis, precise treatment and family agreement with the professional team’s treatment proposal.

Rasprava

Nasljedna gingivalna fibromatoza rijetko je stanje gingivnih tkiva koje se može povezati s nekoliko drugih kliničkih manifestacija, ali je i obilježje nekoliko sindroma (4, 5). Raste sporo i progresivno, a može se pogoršati uporabom lijekova. Može se pojaviti izolirano ili se povezati sa sindromima kao što su Zimmermann-Labandov, Rutherfordov i Ramonovov (3, 7). Važno je istaknuti da je oralno zdravlje pacijentice, uključujući desni i zube, oporavljeno bez promjene i jednogov njezina lijeka, zato što je liječnik dugo odabirao lijek i prilagodavao njegovu dozu za kontrolu konvulzivnih napada. Zato je naš tim procijenio tri glavna uzroka za hiperplaziju gingive, a to su genetika, upotreba antikonvulzivnih lijekova i plak, tj. upalni čimbenici. Važno je istaknuti da u ovom slučaju nije bilo intervencije u vezi s genetskim i medicinskim čimbenicima. S druge strane, kontrola upalnih čimbenika tijekom povremenih posjeta doktoru dentalne medicine i obiteljska suradnja kod kuće, zaslužni su da se do danas hiperplazija gingive nije ponovno pojavila. Zbog toga možemo isključiti hipotezu da hiperplazija gingive nastaje samo zbog medicinskih razloga. U tom kontekstu treba istaknuti koliko je važna kontrola plaka kako bi se spriječile novi i pojavljujuće bolesti, posebno manifestacije rijetkih i teških patologija kao što je NGF, kod kojega je stopa recidiva nakon operacije razmjerno visoka (5). Stoga, unatoč tomu što NGF nije uzrokovao povećanim stvaranjem plaka, plak ga može pogoršati (1).

Važno je napomenuti da je zbog neuroloških poremećaja i nedostatka zaštitnih refleksa, kao što je kašljana, medicinski tim pacijentica preporučio kašastu hranu. Uz invaliditeta i nedostatka zaštitnih refleksa, kao što je kašljanja, medicinski tim pacijentica preporučio kašastu hranu, korisno je učinilo da se do danas hiperplazija gingive nije ponovno pojavila. Zbog toga možemo isključiti hipotezu da hiperplazija gingive nastaje samo zbog medicinskih razloga. U tom kontekstu treba istaknuti koliko je važna kontrola plaka kako bi se spriječili novi i pojavljujuće bolesti, posebno manifestacije rijetkih i teških patologija kao što je NGF, kod kojega je stopa recidiva nakon operacije razmjerno visoka (5). Stoga, unatoč tomu što NGF nije uzrokovao povećanjem stvaranja plaka, plak ga može pogoršati (1).

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Zapravo, upotreba stomatološke opće anestezije indicira je za pacijentice s poteškoćama u razvoju, one koji ne razumiju dobro, za malu djecu i predškolsku djecu koja se ne ponašaju odgovarajuće te za pacijentice s prekomjernim strahom povezanim s nekoliko stomatoloških zahvata (9, 10). No važno je istaknuti da je opća anestezija u dentalnoj medicini poštigla dobre rezultate samo kada je bila povezana s postoperativnim programom prevencije (11). Podupiremo te rezultate i vjeruju da je klinički uspjeh tijekom sedam godina praćen posljedica točne dijagnoze, preciznog liječenja i suradnje obitelji.
Nasljedna gingivna fibromatoza

Zaključak

NFG je rijetka bolest koja se mora oprezno liječiti, posebno kod pacijenata s potrebo za razvojem, kao što su oni s posebnim potrebama. Ovaj prikaz slučaja važan je za to što pokazuje da samo točna dijagnoza može rezultirati pravom terapijom. U ovom slučaju bilo je važno ne mijenjati antikonvulsivne lijekove. Povezano s tim, diferencijalnodijagnostički je bilo važno kontrolirati upalne čimbenike tijekom postoperativnog razdoblja. Zato je iznimno važno da takve bolesnike redovito pregledava doktor dentalne medicine i da se njihov indeks plaka drži pod kontrolom, jer bolest se uvi jek može vratiti.

Sukob interesa

Autori nisu bili u sukobu interesa.

Conflict of interest

The authors deny any conflicts of interest.

Abstract

Introduction: Hereditary gingival fibromatosis (HGF) is a rare disease characterized by gingival enlargement, normal color with benign and firm consistency. This growth may be exacerbated by use of drugs and plaque build-up. The treatment for this clinical condition is surgical excision of the enlarged gingival tissue or the extraction of all teeth. Case Report: A 20-year-old Brazilian female handicapped patient with a chief complaint of exaggerated gingival enlargement who had been prescribed Carbamazepine and Gardenal was referred to our center. According to the clinical presentation and family history, the final diagnosis of gingival enlargement was HGF. Full dental treatment was performed, including basic periodontal treatment, restorations, sealants, and gingivoplasty with internal bevel. Special care was taken to ensure that there was no change in patient’s anticonvulsant medication. The patient has been monitored for seven years without signs of recurrence of gingival hyperplasia due to constant professional and home control of plaque.

Conclusion

HFG is a rare disease that must be treated very carefully, especially in systemically compromised patients such as mentally disabled patients. This case report has relevance since the correct diagnosis may lead to proper treatment. In this case, it was of fundamental importance not to change the anticonvulsant medication. Associated with this, the differential was exactly the control of inflammatory factors during the post-operative period. Thus, it is extremely important that these patients are regularly seen by the dentist and their plaque index is kept under control because the disease can recur.

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