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PRIKAZ SLUČAJA
CASE REPORT

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Nasljedna gingivna fibromatoza: prikaz slučaja nakon sedmogodišnjeg praćenja

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

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Sažetak

Uvod: Nasljedna gingivna fibromatoza (NGF) rijetka je bolest koju obilježava povećanje gingive, no boja desni ostaje normalna, a konzistencija je dobra i čvrsta. Taj rast može potaknuti uporaba lijekova i nakupljanje plaka. Terapija izbora za to kliničko stanje jest kirurško uklanjanje povećanoga gingivnog tkiva ili vađenje svih zuba. **Prikaz slučaja:** U naš centar upućena je 20-godišnja Brazilka s posebnim potrebama zbog pretjeranog povećanja gingive. Od lijekova je uzimala Carbamazepin i Gardenal. Prema kliničkoj slici i obiteljskoj anamnezi, konačna dijagnoza zbog povećanja gingive bila je NGF. Odlučeno je da je potrebna stomatološka terapija, uključujući osnovno parodontno liječenje, restauracije, pećaćenja i gingivoplastiku s unutarnjim rezovima. Posebno se pazilo na to da se pritom ništa ne promijeni u primjeni antikonvulzivnih lijekova koje je uzimala. Pacijentica je nakon toga praćena sedam godina i hiperplazija gingive nije se ponovno pojavila zahvaljujući stalnom profesionalnom nadzoru i kućnoj kontroli zubnoga plaka.

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Ključne riječi
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Uvod

Nasljedna gingivna fibromatoza (NGF) rijetka je bolest autosomnih dominantnih i recesivnih tipova s učestalošću od 1 : 175.000 (1). Klinički je obilježava benigno proširenje gingive normalne boje i čvrste konzistencije te nehemoragična simptomatska bolest (2). Iako povećanje gingive ne utječe izravno na alveolarnu kost, oteklina može povećati akumulaciju bakterija te uzrokovati gingivitis, parodontitis, resorpciju kostiju i halitozu (1). Histološki, u vezivnom je tkivu povećan udjel kolagena i fibroblasta, a epitelno tkivo buja uz istaknute pupoljke. /istaknuta izbočenja/fistule. Unatoč tim obilježjima, histološke značajke NGF-a nisu točno određene, a konačna dijagnoza trebala bi biti postavljena na osnovi obiteljske anamneze i kliničkih nalaza (1). Neke kliničke komplikacije NGF-a mogu biti prekomjerni rast gingive, što rezultira pseudodžepovima i parodontnom bolešću zbog loše oralne higijene, diasteme, odgođenim nicanjem zuba i deformacijama lica zbog protruzije usana. Uz to, u najtežim slu-

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, otežati govor i činiti poteškoće pri žvakanju (1, 2 – 7).

Među autorima istraživanja nema suglasja o učinkovitosti liječenja NGF-a (6), no svi se slažu da postoji rizik od njegova ponovnog pojavljivanja što treba izbjegavati (7). Ovisno o rastu, liječenje uključuje izrezivanje povećane gingive uobičajenim kirurškim zahvatom, primjenjuju se i elektrokirurgija ili laser, apikalno se pomiče režanj vade se svi zubi i smanjuje alveolarna kost kako bi se spriječilo ponavljanje bolesti (1, 2). Uspjeh dentalne terapije ovisi o raspoređivanju preventivnih kontrolnih pregleda kako se bolest ne bi ponovila (6, 7).

Ovom je radu svrha opisati način postavljanja dijagnoze te način liječenja i praćenja nasljedne fibromatoze gingive na temelju prikaza slučaja s povećanom gingivom kod pacijentice 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 mikrocefalije, epilepsije 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 od 200 mg karbamazepina (3x/dan), 100 mg fenobarbitala (1x/dan), uzimala je i 5 mg ciklobenzaprin-hidroklorida (1x/dan), 50 mg ranitidina (1x/dan), željezo (40 kapi/ dan), kalcij (10 ml 1x/dan) i minerale (10 ml 1x/dan). Bila je u stanju od 50 bodova prema Karnofskyjevoj ljestvici statusa izvedbe (8), jela je samo mekanu hranu i težila 14 kilograma. Važno je napomenuti da je pacijentica imala *grand mal* napadaje od osam do deset puta na dan, a katkad je zbog njihove težine bila potrebna i hospitalizacija. Kad je došla u hitnu službu napadaji su bili u početnoj fazi zato što je liječnik prilagodio dozu lijeka. Nakon nekoliko promjena doza, konvulzije su stabilizirane odgovarajućom i redovitom uporabom antikonvulziva.

Intraoralni pregled pokazao je obimnu hiperplaziju gingive povezanu s visokim indeksom plaka (3,16 prema indeksu IHOS-a), zatim gingivalni kamenac, lažne parodontne džepove, tri karijesom zahvaćena zuba i četiri zuba s aktivnim bijelim točkama (slike 1., 2. i 3.). Dentalno liječenje obavljeno je u općoj anesteziji jer je bilo potrebno provesti nekoliko terapijskih postupaka, a opći zdravstveni status pacijentice bio je klasificiran kao ASA II.

Primijenili smo inicijalnu parodontnu terapiju i postavili kompozitne restauracije, zapečatili aktivne bijele točke korištenjem modificiranih, atraumatskih, restaurativnih tehnika te kirurški uklonili treće kutnjake. Kad je riječ o gingivi, nakon što smo obilježili dubinu džepova, uklonili smo višak tkiva s pomoću unutarnje kose gingivektomije s nekih dijelova bukalnih i palatalno-jezičnih površina. Rezovi s unutarnje strane bili su učinjeni tako da zadrže odgovarajuću količinu

tooth eruption, and facial disfigurement due to lip protrusion. In addition, in the most severe cases, gingival hyperplasia can result in limited tongue movement, speech impediments 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). Depending on the severity of the of the growth, treatment involves the excision of the enlarged gingival tissues, using conventional 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 of dental treatment depends on scheduling a return appointment as a preventative for recurring disease (6, 7).

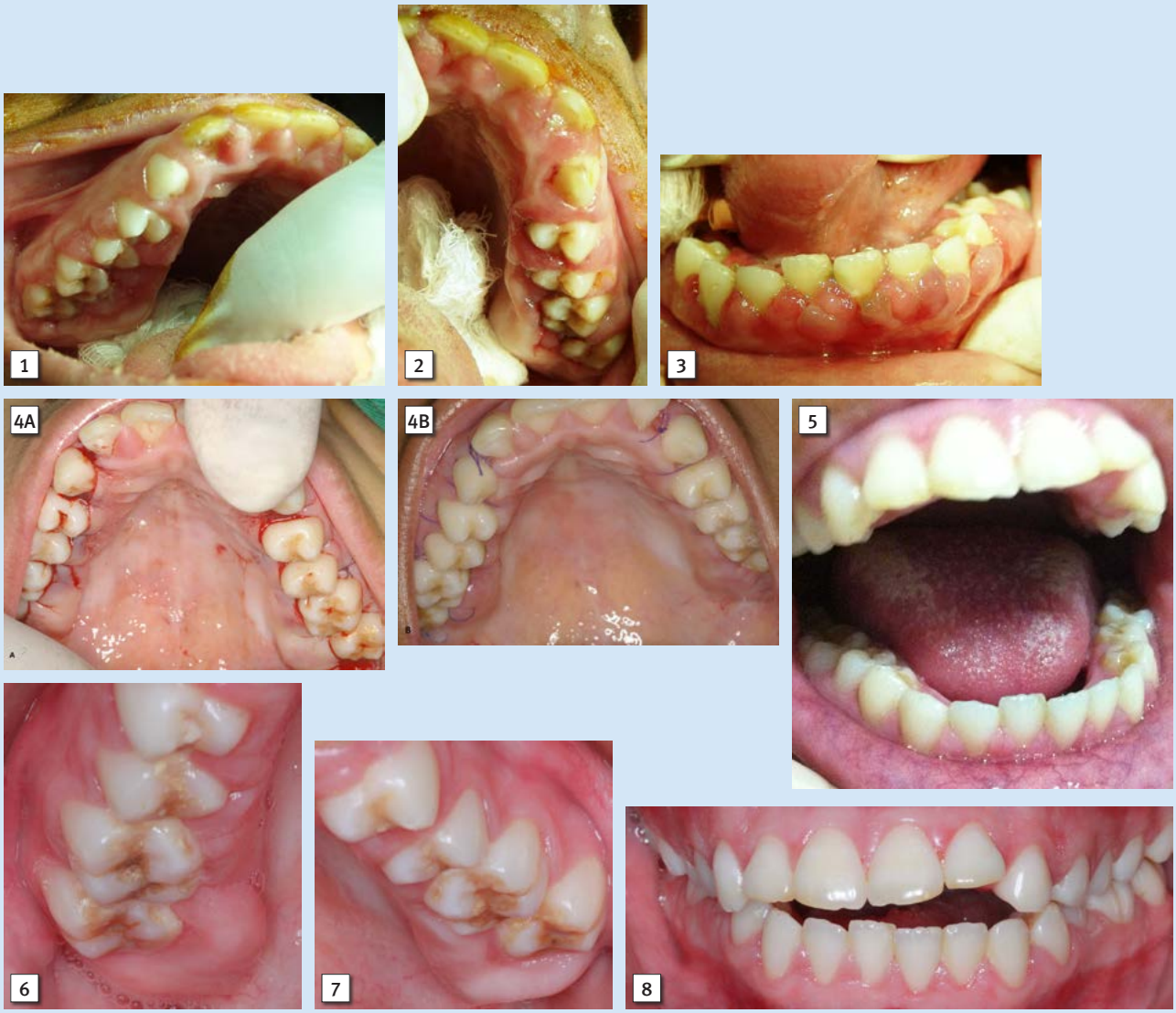
This paper aims to review the diagnosis, treatment, and follow-up of hereditary gingival fibromatosis by the presentation of a case report characterized by the exacerbated gingival enlargement through the use of anticonvulsant medication, associated with the inflammatory factor, i.e., high levels of plaque. In addition, this case report presents the local treatment plaque control as a differential without any interference with systemic medication.

Case report

A bedridden, 20-year-old female patient, who suffered from microcephaly, epilepsy, and mental retardation, was referred to our center in January 2009 with a chief complaint of 'pain in the jaws', according to her mother. In the course of taking medical history she reported another case of gum growth in her brother, who had died. The patient was on carbamazepine 200 mg (3 times a day), phenobarbital 100 mg (1 time a day), cyclobenzaprine hydrochloride 5 mg (1 time a day), ranitidine 50 mg (1 time a day), iron (40 drops a day), calcium (10 ml 1 time a day) and minerals (10 ml 1 time 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.

The intraoral examination revealed severe gingival hyperplasia associated with a high plaque index (3.16, according to IHOS index) - gingival calculus, false periodontal pockets, 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 procedures to be carried out and the patient's general health status was classified as ASA II.

We performed the basic periodontal treatment and composite resin restorations, sealing the active white spots using modified, atraumatic, restorative techniques, 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-



Slika 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.
Slika 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.
Slika 3. Pogled sprijeda na mandibularne sjekutiće prije kirurškog liječenja u općoj anesteziji
Figure 3 A front view of mandibular incisors before surgical dental treatment under general anesthesia.
Slika 4. A: Neposredno poslije operacije. B: Petnaest dana poslije operacije
Figure 4 A: Immediately after surgery. B: Fifteen days after surgery.
Slika 5. Kontrola nakon jedne godine
Figure 5 One-year follow-up.
Slika 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.
Slika 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.
Slika 8. Pacijent koji je pod nadzorom nema karijes ili parodontnu bolest
Figure 8 The patient undergoing monitoring without evidence of caries or periodontal disease.

tkiva gingive nakon kirurškog zahvata te omoguće pasivnu zamjenu kirurškog režnja, čime se postiže i brže zacjeljivanje rane. Odlučili smo se za tu tehniku zbog pacijentičinih posebnih potreba i velike količine tkiva koje se trebalo izrezati u jednom posjetu.

Pacijentica je dolazila u naš centar na kontrolne preglede svaka tri mjeseca tijekom sedam godina nakon kirurškog zahvata. Trenutačno joj je zdravstveno stanje pod nadzorom, uključujući i oralno zdravlje, te nema znakova hiperplazije

sions in the internal bezel were designed to preserve a proper amount of gum tissue after surgery as well as to allow the passive replacement of surgical flap, thus achieving healing all at once. We opted for this technique based on the patient's special care needs and the large amount of tissue to be excised in one session.

The patient is still undergoing follow-up every three months in our center, seven years after the surgical procedures. Currently, her health status is under control, includ-

gingive, lažnih parodontnih džepova ili karijesa (slike 4., 5., 6., 7. i 8.). Informirani pristanak potpisala je pacijentičina majka.

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 ijednoga njezina lijeka, zato što je liječnik dugo odabirao lijek i prilagođavao njegovu dozu za kontrolu konvulzivnih napadaja. Zbog toga 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 nove i ponavljajuće 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 uzrokovan povećanim stvaranjem plaka, plak ga može pogoršati (1).

Važno je napomenuti da je zbog neuroloških poremećaja te invaliditeta i nedostatka zaštitnih refleksa, kao što je kašljanje, medicinski tim pacijentici preporučio kašastu hranu.. Uz to, oralna zdravstvena zaštita razmjerno je zanemarena zbog općega zdravstvenog problema. Takva je situacija u skladu s tvrdnjom Escribano-Hernándeza i suradnika (9) koji napominju da se kod pacijenata s mentalnim poteškoćama često zanemaruje oralna zdravstvena zaštita. Oni također ističu da je stopa zubnog karijesa i parodontne bolesti znatno veća u toj populaciji zbog pasirane hrane, visokog udjela ugljikohidrata u prehrani i fizičkih ograničenja koja sprječavaju odgovarajuće čišćenje zuba zbog oštećene funkcije mišića.

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

ing her oral health, without any signs of gingival hyperplasia, false periodontal pockets, or caries (Figures 4, 5, 6, 7 and 8). Informed consent has been obtained from the patient's mother.

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 caries 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.

Zaključak

NFG je rijetka bolest koja se mora oprezno liječiti, posebno kod pacijenata s poteškoćama u razvoju, kao što su oni s posebnim potrebama. Ovaj prikaz slučaja važan je zato što pokazuje da samo točna dijagnoza može rezultirati pravom terapijom. U ovom slučaju bilo je važno ne mijenjati antikonvulzivne 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 uvijek može vratiti.

Sukob interesa

Autori nisu bili u sukobu interesa.

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 postoperative 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.

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.

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Key words
Gingival Fibromatosis; Gingival Hyperplasia

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