Long-term management of an idiopathic gingival fibromatosis patient with the primary dentition

Suttatip Kamolmatyakul, BSc, DDS, MSc, MPH, DScD  Suparp Kietthubthew, BSc, MSc
Orasa Anusaksathien, DDS, MScD, DMSc

Abstract

Gingival fibromatosis (GF) usually develops as an isolated disorder, but rarely as part of a syndrome. 1-4 It affects only one in 750,000 people. 5 Those clinical features commonly associated with syndromic form of GF are hypertrichosis, epilepsy and mental retardation. 1-3,6 Hereditary GF, also known as idiopathic GF, is usually identified as an autosomal dominant condition; although, recessive forms are described in literature. 5,7-10 Recently Hart and co-workers 11 demonstrated that at least two loci are responsible for autosomal dominant hereditary GF, one of which already known is 2p 21 – p22.12

Oral manifestations of GF can vary from localized to specific areas of the mouth, typically the maxillary tuberosities and the buccal gingiva around the lower molars, 13 to generalized involvement that can, as in the described cases, 5,7,14 inhibit the complete eruption of the teeth into the oral cavity and cause a delay in shedding primary teeth. 5,15 This enlargement can vary in severity, sometimes covering the entire crowns of the teeth and deforming the palate, thereby creating difficulties in speech and mastication. In extreme cases the lips are everted by the excess gingival bulk and are unable to close. 1 Diastema, malposition of teeth, and prolonged retention of primary teeth are the most common effects related to GF. 16 Other effects including cross-bite and open-bite have also been reported. 17

The overgrown gingival tissues appear normal at birth but begin to enlarge with the eruption of the primary teeth. The enlargement continues with the eruption of permanent teeth until the tissue essentially covers the clinical crowns of the teeth. The hyperplastic gingiva, resulting from an increase in the connective tissue elements of the submucosa, usually is of a normal color and has a firm consistency with abundant stippling on the adjacent gingiva. Buccal and lingual tissue may be involved in both the mandible and maxilla, and the degree of hyperplasia may vary between individuals. 18

Histologically, fibromatosis is described as a moderate hyperplasia of the epithelium with hyperkeratosis and elongation of the rete peg. The increase in tissue mass is primarily the result of an increase and thickening of the collagenous bundles in the connective tissue stroma. The lesions are relatively acellular and feature an increased amount of randomly arranged bundles of collagen. 19 The possibility of an association between growth hormone deficiency and gingival overgrowth when dental and skeletal alterations are present along with GF, has also been suggested. 20

The cause of gingival overgrowth in fibromatosis is unknown. Many drugs have been reported as inducing gingival overgrowth. Some examples of these drugs are cyclosporin, 21-27 an anti-rejection drug, verapamil 28 and nifedipine, 29,30 which are used for treating angina and cardiac arrhythmia; and phenytoin, a well-known anticonvulsant drug. 35,36

Case report

A one-year-and-six-month-old girl came with her mother to the dental hospital at Prince of Songkla University (PSU) with a chief complaint of gum swelling. At that time she was the only child of a non-consanguine marriage. There was no family history of gingival overgrowth. No prenatal history seemed contributory. The postnatal history revealed a convulsive disorder, which started when she was 10 months old due to high body temperatures resulting from a fever. Phenobarbital
Fig 2. Note a cupid bow mouth of the patient due to overgrowth of gingiva in both arches

Fig 3. A radiograph revealed all primary teeth and enamel formation of all first permanent molars

Fig 4. A dental orthopantomogram showed all primary teeth had erupted from the alveolar bone, except upper primary second molars. The upper primary canines were partially erupted from the alveolar bone. There were excessive spaces between the primary teeth. All permanent teeth were developed, except the third molars. There were radiolucent areas at the occlusal surfaces of both lower first primary molars adjacent to the pulp (arrows).

Grain I (60 mg/tab) was given to her starting then in the dosage of half a tablet twice a day. After that she became a regularly attended patient at the pediatric clinic at PSU hospital.

A physical examination demonstrated hepatomegaly, but no spleenomegaly. No evidence of delayed development, hypertrichosis or apparent hearing problems were detected. Other positive medical work up included strabismus and an abnormal sharp wave at bilateral posterior on an EEG. The extraoral examination revealed a coarse face with an incompetent lip (Fig 1) due to overgrowth of gingiva in both arches resembling a cupid bow mouth (Fig 2). Intraorally, there were pale-pink, firm, overgrown gingiva generalized in both arches with no significant degree of inflammation. The only teeth that could be seen clinically were the lower primary central incisors. A radiographic assessment revealed all primary teeth and enamel formation of all first permanent molars (Fig 3). On the basis of the medical history, family history and clinical finding, a diagnosis of idiopathic GF was made.

The child’s mother was advised that surgical removal of the hyperplastic tissue could be done to help improve her appearance, but the condition might recur within a few months. Since the patient was very young and had no problem with overgrown gingiva, apart from the aesthetic concern of her mother, it seemed inappropriate to perform a gingivectomy at that time. The mother was told to bring the child back for a check-up every 6 months. However they never returned for recall appointments until an episode of pain developed when the child was 5 years and 5 months old with a complaint of mild pain in her lower left jaw. Oral examination revealed a pale-pink, firm overgrowth of gingiva covering all primary teeth, except the lower primary central incisors.

A dental orthopantomogram (Fig 4) showed all primary teeth had erupted from alveolar bone except upper primary second molars. The upper primary canines were partially emerged from the alveolar bone. There were excessive spaces between the primary teeth. All permanent teeth were developed, except the third molars. There were radiolucent areas at the occlusal surfaces of both lower first primary molars adjacent to the pulp. Therefore, a gingivectomy of the lower arch was planned to be performed under general anesthesia to expose and restore the infected teeth. The overgrown gingival tissue in the lower arch was planned to be removed at the same time. A gingivectomy was planned to be performed only in the lower arch to monitor the tolerance of the young patient to this procedure.

Under general anesthesia (GA), overgrown gingival tissue was removed from the mandibular right primary incisor to the mandibular left primary second molar area by the conventional, external bevel gingivectomy technique. Removal of these thickened gingival tissue revealed the lower permanent central incisors were partially erupted lingual to the lower primary central incisors. Therefore, the lower primary central incisors were extracted which was followed by a periodontal pack placement for one week. No complications were experienced and the patient tolerated the procedure well. Instructions not to brush or to chew on the surgical site were given. A 0.2% chlorhexidine rinse after brushing twice a day for two weeks to reduce plaque formation was prescribed. The treatment of the lower first primary molars and gingivectomy of the rest of the lower arch had to be postponed.

This large-scale lesion was more difficult than expected; therefore, it took longer time than was planned for the gingivectomy. This lengthy operation resulted in excessive bleeding due to a lack of electrocautery or carbon dioxide laser to help stop the bleeding.
Histopathology

A histopathological examination of the excised gingiva demonstrated hyperplastic parakeratinized stratified squamous epithelium covered on fibro-collagenous tissue. It was a mass of dense collagenous bundles interacingly arranged and exhibited long slender rete pegs (Figs 5, 6). A pathological diagnosis of gingival fibromatosis was made. A histopathological examination of the mandibular primary incisors revealed normal structure of enamel and dentine.

A week later under N₂O/O₂ sedation, the mandibular left primary first molar was restored with amalgam after cervical pulpotomy with formocresol was performed. N₂O/O₂ sedation was used to reduce the anxiety of the patient. Fissure sealant was also placed on the occlusal surface of the lower left primary second molar. The permanent restoration with a stainless steel crown (SSC) was planned to be done later because of the patient’s behavior management problems, which include a speech delay and a learning disability.

Two weeks later, the patient returned for a check up and polishing of the amalgam restoration on the previously restored mandibular left primary first molar. The healing of gingiva on her lower left arch was good (Fig 7). The patient returned one and a half months later with a complaint of about 3 days of swelling and pain in the lower right posterior region. An intraoral examination revealed inflamed gingiva with an abscess discharge from the lower right first primary molar area. This tooth could not be seen clinically. Therefore, a gingivectomy of the lower right arch was performed before extraction of the lower right first primary molar could be done under GA.

The patient had an operation under GA for the third time at the age of 6 years and 2 months because she and her mother were very concerned about the unpleasant appearance of the overgrown gingiva. Although they understood that the nature of this disorder has a high recurrent rate, they agreed to have a gingivectomy of the upper arch performed under GA. The patient returned for follow-up visits within one, two and four weeks. The healing of gingiva on her upper arch was good (Fig 8). After that she began to be a regular patient at the pediatric dentistry clinic at PSU dental hospital.

At the age of 7 years and 1 month, a carious lesion in the mesial surface of the lower left second primary molar was detected. Amalgam restoration was then successfully performed with local anesthesia under N₂O/O₂ sedation. One month later the final restoration of the lower left first primary molar with an SSC was performed under N₂O/O₂ sedation and local anesthesia. On the following recall visit, at the age of 7 years and 7 months, a carious lesion on the occlusal surface of the lower right second primary molar was detected. Amalgam restoration was then successfully performed under local anesthesia without N₂O/O₂ sedation. The mother was encouraged to bring the child back for a check-up every 6 months.
As the patient grows older, some characteristics of Mucopolysaccharidoses could be more clearly noticed. These characteristics include an enlarged head, a short neck, a coarse face, a mid-face hypoplasia with a depressed nasal bridge, protrusion of the mandible (Fig 9), a short trunk, and clawed hands (not shown). A laboratory investigation of Mucopolysaccharidoses was conducted with a negative result. Therefore, the cause of gingival overgrowth in this case is still unknown.

Discussion

This case of GF may be idiopathic. There was no family history of gingival overgrowth and the prenatal history was non-contributory. There was no history of drug (related to gingival overgrowth) uses other than phenobarbital.

Most reports of hereditary GF have found that the gingival enlargement is noted at the time of tooth eruption, either primary or permanent, while others have reported gingival enlargement from birth. Recurrence of hereditary gingival overgrowth is unpredictable and each case should be considered individually.

A variety of factors can cause generalized gingival enlargement including inflammation, leukemic infiltration, and chemical induction as seen with phenytoin, verapamil, cyclosporin or nifedipine. Otherwise, GF may occur alone or as part of a syndrome, the least rare of which is a combination of GF, hypothyroidism, epilepsy and/or mental retardation. GF is also a feature of the Rutherford syndrome (GF and corneal dystrophy), the Laband syndrome (GF, ear, nose, bone, and nail defects with hepatosplenomegaly), the Cross syndrome (GF, microphthalmia, mental retardation, athetosis, and hypopigmented skin), the Murray-Puretic-Drescher syndrome (GF with multiple hyaline fibromas), the Jones syndrome (GF with sensorineural deafness) and the Byars-Jurkiewicz syndrome (GF, hypothyroidism and giant fibroadenomas of the breast).4,40 This patient had no clinical findings that fulfilled any of these possible syndromes.

As in this case, the hyperplastic tissue usually is of normal color. The degree of severity varies from mild involvement of one quadrant to severe involvement of all four quadrants. Gingival tissue enlargement usually begins with the eruption of teeth, either primary or permanent, while others have reported gingival enlargement from birth. Recurrence of hereditary gingival overgrowth is unpredictable and each case should be considered individually.

As the patient grows older, some characteristics of Mucopolysaccharidoses could be more clearly noticed. These characteristics include an enlarged head, a short neck, a coarse face, a mid-face hypoplasia with a depressed nasal bridge, protrusion of the mandible (Fig 9), a short trunk, and clawed hands (not shown). A laboratory investigation of Mucopolysaccharidoses was conducted with a negative result. Therefore, the cause of gingival overgrowth in this case is still unknown.

In this case, the post-natal history of a convulsive disorder developed at the age of 10 months. However, gingival overgrowth had been detected before phenobarbital had been given regularly to her. In addition, there has been no report of phenobarbital-induced gingival overgrowth.

Another interesting finding from this case was the delayed eruption of the primary teeth. At the age of 5 years and 5 months, her upper second primary molars still had not emerged from the alveolar bone and the upper primary canines had just partially emerged from alveolar bone (Fig 4) but were completely covered with overgrown gingiva. Further analysis needs to be done to investigate the cause of this delayed eruption which may be due to the abnormality of bone apart from the dense overgrown gingiva normally found in a GF patient. There was a report of delayed eruption in an 11-year-old Japanese girl in 1990 by Mega.45 The teeth involved in that case were permanent teeth and not primary teeth as in this case.

In the area of patient management, although the chief complaint on the patient’s first visit was gingival overgrowth at one year and six months old, a gingivectomy had not performed. This was due to the decision concerning the age of the child and the possibility of a high recurrent rate. On the other hand, a gingivectomy had to be performed at the age of 5 years and 5 months to expose the teeth which were causing the pain. Therefore, curative and restorative treatment could be carried out.

The teeth covered by overgrown gingiva would not likely be infected. However, this was not the case as the lower right first primary molar had already emerged from the alveolar bone; but was covered with overgrown gingiva, was decayed, and an abscess had developed. Therefore, the best course of action would be to perform a gingivectomy. This operation would prevent occlusal caries of the primary molars by fissure sealant placement at the age of 3 years instead of curative and restorative treatment after development of the abscess at 5 years of age. Then the same procedure to prevent occlusal caries of the permanent first and second molars, at the age of 6 and 12 years should be performed, respectively.

The preservation of primary teeth should be considered; even though, it may seem that there is excessive space in the primary dentition. The Orthopantomograph shows that all permanent teeth, except the third molars, are present and the sizes of the teeth are quite large. Therefore, there is a chance of crowding permanent dentition if there is a premature loss of primary teeth.

However, the preservation of the lower right second primary molar could not be accomplished because of a poor prognosis. On the other hand, a space maintainer at this area could cause irritation to the gingival tissues and worsen the condition of the gingival overgrowth. Therefore, a space maintainer should not be used for the prevention of space loss. Consequently, this might result in future crowding of the permanent dentition at this area.

Advance planning to prevent gingival inflammation and infection should be considered. Every effort should be made to give the most effective preventive procedure to the patient. This includes, but is not limited to, effective plaque removal, fluoride therapy, and fissure sealant. These procedures cannot be accomplished properly if the teeth are covered with overgrown gingiva. However, a GF patient may occasionally need a gingivectomy, as there is a possibility of recurrence of hyperplastic tissue following a gingivectomy which necessitates a repetition of the procedure. In addition, the psychological benefits resulting from cosmetic improvement far outweigh the risks of recurrence.41 Therefore, effective plaque removal, fluoride therapy, and fissure sealant can be occasionally done after a gingivectomy is performed. The question is: when is the suitable time to perform a gingivectomy for these patients who have a high recurrent rate of gingival overgrowth?

The appropriate time for the removal of overgrown gingiva varies. Emerson42 recommended that the best time is when all the permanent teeth have erupted. Rushton15 did not indicate an exact time, but suggested that the teeth be free of caries and gingivitis. In this case, the appropriate time for the removal of overgrown gingiva might have been at the age of 3, 6 and 12 years old to have effective plaque removal, to apply fissure sealant on the molars, and a topical fluoride application to prevent caries.
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References


ABSTRACT OF THE SCIENTIFIC LITERATURE

EFFECT OF COMBINATION THERAPY INCLUDING PROTEASE INHIBITORS ON MORTALITY AMONG CHILDREN AND ADOLESCENTS INFECTED WITH HIV-1

The combination of human immunodeficiency virus (HIV) specific protease inhibitors with nucleoside reverse-transcriptase inhibitors, nonnucleoside reverse-transcriptase inhibitors, or both has been demonstrated to slow the progression of HIV type 1 (HIV-1) disease dramatically and to lower mortality in adults. Recent studies provide some evidence of the efficacy and safety of these regimens in children and adolescents, but there is only limited evidence of reductions in mortality and morbidity.

The Pediatric AIDS Clinical Trials Group Protocol 219 (PACTG 219) study is a prospective cohort study designed to assess the long-term effects of prenatal and neonatal exposure to antiretroviral drugs in clinical trials and the late effects of antiretroviral treatment in children infected with HIV-1. A cohort of 1028 HIV-1 infected children and adolescents, from birth through 20 years of age, who were enrolled in research clinics in the U.S. before 1996 was followed prospectively through 1999. The medical history was obtained, a physical examination was performed, the height and weight were measured, and data on lymphocyte subpopulations was collected at base line and every 6 months for children less than 24 months of age and yearly for children 24 months old or older. The effects of combination therapy including protease inhibitors on mortality among children and adolescents infected with HIV-1 were investigated. Differences in the time of initiation of this therapy according to age, sex, socioeconomic status, or ethnic background were also identified.

Seven percent of the subjects were receiving combination therapy including protease inhibitors in 1996. By 1999, 73% were receiving such therapy. After adjustment for covariates, the differences among racial and ethnic groups to initiation of combination therapy were not statistically significant. Mortality declined from 5.3% in 1996 to 2.1% in 1997, 0.9% in 1998, and 0.7% in 1999 (P for trend <0.001). There were reductions in mortality in all subgroups defined according to age, sex, percentage of CD4+ lymphocytes, educational level of the parent or guardian, and race or ethnic background. In adjusted analyses, the initiation of combination therapy, including protease inhibitors, was independently associated with reduced mortality (hazard ratio for death, 0.33; 95% confidence interval, 0.19 to 0.58; P<0.001). Therefore, it is concluded that the use of combination therapy including protease inhibitors has markedly reduced mortality among children and adolescents infected with HIV-1.

Address correspondence to the Center for Biostatistics in AIDS Research, Harvard School of Public Health, Boston, MA 02115.


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