Gingival fibromatosis and significant tooth eruption delay in an 11-year-old male: a 30-month follow-up

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Summary. This case report describes the dental management of an unusual case of idiopathic gingival fibromatosis with multiple impacted primary teeth, and the absence of eruption of permanent teeth, in an 11-year-old boy and at the 30-month follow-up. The patient presented with severely enlarged gingival tissues affecting both arches and multiple retained and nonerupted primary teeth. He had already been subjected to localized gingivectomies at the ages of 7 and 9 years. He had no known syndrome and there was no family history of any similar disorder. The patient was treated under general anesthesia to remove the excessive gingival tissues using apically positioned flaps. During the surgical procedure, over-retained and unerupted impacted primary teeth were extracted in order to facilitate the eruption of the permanent successors. Two years postoperatively, there was no recurrence of the gingival enlargement. Overdentures were then constructed because none of the permanent teeth had yet erupted. Furthermore, preeruptive coronal resorption was detected radiographically affecting the crown of the unerupted 36. Thirty months postoperatively, no recurrence of gingival enlargement was seen, but the permanent teeth had still not erupted.

Introduction

Gingival fibromatosis is a rare disease that is characterized by a slowly progressive enlargement of the maxillary and mandibular gingiva [1,2]. The prevalence is one per 175,000 population, and it affects men and women equally [3]. The condition occurs either as an isolated disorder [1], or as a part of distinct genetic syndromes such as Zimmermann-Laband syndrome, Murray-Ramond syndrome, Rutherford syndrome and Cross syndrome [4]. Associations between hereditary gingival fibromatosis and deficiencies in growth hormone and growth hormone releasing factor have also been reported [5]. Gingival fibromatosis may occur concomitantly with other conditions, such as hypertrichosis, epilepsy, learning difficulties and sensorineural hearing loss [4]. The disease may be hereditary or idiopathic. Most hereditary gingival fibromatosis cases are autosomal dominant conditions; however, there have been reports of an autosomal recessive pattern of inheritance [6,7].

The exact pathogenesis of the disease is unknown. Hereditary gingival fibromatosis is associated with increased extracellular matrix production by fibroblasts and decreased collagen degradation [8–10]. Transforming growth factor beta (TGF-β) stimulates fibroblasts to produce extracellular matrix materials such as procollagen polypeptides, resulting in overproduction of collagen, mainly type I [11]. It has recently been found that there are qualitative and quantitative differences in TGF-β isoform and receptor expression by fibroblasts in gingival overgrowth, and this may contribute to the pathogenesis of the disease...
It has also been reported that fibroblasts from hereditary gingival fibromatosis proliferate in vitro more rapidly and produce lower amounts of matrix metalloproteinase-1 and -2, which are mainly associated with extracellular matrix degradation, thus leading to decreased collagen degradation [9].

Clinically, the enlarged gingivae are usually pink and not erythematous, with firm consistency and heavy stippling [6,9,12]. The condition may be generalized or occur on localized portions of the gingiva [13-15]. Both types vary in extent and severity. The gingival enlargement may become so severe that it may cover the teeth up to their occlusal surfaces [1,14,15]. The most common side-effects related to the gingival lesions are diastemata, malpositioning of the teeth and prolonged retention of primary teeth [12], as well as delayed eruption of permanent teeth [5,8,16,17].

Gingival tissue enlargement usually begins with the eruption of the permanent dentition, although it may also develop with the eruption of primary teeth but it is rarely present at birth. Enlargement seems to progress suddenly during the eruption of both primary and permanent teeth, and decrease upon completion of eruption [12].

Histological findings consist of fibrous connective tissue with bundles of coarse collagenous fibres, young fibroblasts, scarce blood vessels, and epithelium with elongated rete pegs and hyperkeratosis; usually, no subepithelial inflammation is present [5,12]. Electron microscopic observations have shown flocculent material in the extracellular matrix, collagen fibrils with structural abnormalities and variations in diameter [18]. In general, the histological features are relatively nonspecific, and therefore, the definitive diagnosis of gingival fibromatosis is mainly based on family, medical and dental history, and on clinical findings [12,15,19].

This case report describes the dental management of an unusual case of idiopathic gingival fibromatosis, with multiple impacted primary teeth, absence of the eruption of the permanent teeth and microcephaly, in an 11-year-old boy, and the 30-month follow-up.

Case report

An 11-year-old Caucasian boy was referred to the Department of Paediatric Dentistry, University of Athens, Athens, Greece, because of extensive gingival swelling that had caused him psychological, aesthetic and functional problems.

The boy was the eldest of three children. He had been born after an uncomplicated pregnancy at 38 weeks of gestation. His medical history was noncontributory. The subject was not taking any medication. According to national standards, he was of short stature (135 cm) and low weight (26 kg), below the fifth percentile for his age, and had microcephaly (head circumference = 48 cm). His growth hormone levels had been checked at age 8 years of age and were within normal limits. The subject also had dark skin, and skin pigmentation on the nose and cheeks. He had no hearing impairment, no hypertrichosis and no learning difficulties. The patient’s main complaint was difficulty in mastication and poor aesthetics.

There was no family history of such a disorder. His mother could not remember exactly when the problem had started, but she reported that he had been subjected to localized gingivectomies at the ages of 7 and 9 years. This indicated that the gingival enlargement had been present for at least 4 years.

Extraorally, the subject had no facial deformities. He had difficulty in closing his lips because of the severely enlarged gingival tissues, especially in the mandibular arch, which showed through his lips.

The intraoral clinical examination revealed generalized and severe gingival enlargement that affected both the maxillary and the mandibular arches. Enlarged gingival tissues, which were pink in colour and of a firm consistency, covered the largest part of the crowns of the teeth. Many primary teeth were retained with increased spacing and no permanent teeth had erupted. The only teeth present in the oral cavity at the time of the clinical examination were 51–54, 61–64 and 71–73 (Fig. 1). The oral hygiene was moderate and the initial plaque index was 1.25 [20]. There was no bleeding on probing with the periodontal probe. No clinical attachment loss or increased probing depth could be identified around the existing teeth. The depth of the gingival crevice ranged from 0 to 2 mm.

The radiographic examination included panoramic and cephalometric views, and periapical views of the upper and lower anterior teeth. Unerupted primary (55, 65, 75 and 85) and permanent (16, 26, 36 and 46) teeth were visible on the radiographs (Fig. 2). The occlusal surfaces of the impacted permanent first molars were covered by a thin layer of bone, although their roots were almost fully developed. Radiographs suggested that the second premolars were likely to erupt ectopically as a result of retention of the primary predecessors. The alveolar bone was

intact and no loss was evident around the existing teeth. Cephalometric analysis did not reveal any skeletal discrepancies.

The subject was then referred for chromosomal analysis. This proved to be normal and no known syndrome could be identified. At the genetic consultation, it was suggested that the affected subject might have inherited an autosomal recessive gene from each parent or presented a new mutation.

Based on the noncontributory medical and family history, the absence of medication and the clinical findings, the patient was diagnosed as having a symmetrical form of idiopathic gingival fibromatosis that was not associated with any known syndrome.

The treatment plan included oral hygiene instructions, surgical removal of the hyperplastic tissues and restoration of the morphology of both arches, extraction of the over-retained and nonerupted primary teeth, removal of the bone layers covering the nonerupted permanent teeth, and long-term monitoring of the eruption process.

The patient lived in a remote rural area and it was very difficult for the family to bring him to the authors department for repeated procedures. Because of the difficulty in attending and to avoid multiple surgical procedures, which were likely to be too stressful for a child of his age, it was decided to perform surgery under general anaesthesia in a single session.

During surgery for both upper and lower arches, an initial (buccal) internal bevel incision was made along the alveolar crest at least 3 mm coronal to the mucogingival junction using a 15 Bard-Parker blade.
A second (lingual) internal bevel incision was made, using the same blade, along the alveolar crest at a distance at least 4 mm from the initial incision. Full-thickness flaps were elevated and were then internally thinned, especially in the lingual incision. A horizontal incision at the level of the alveolar crest was made to facilitate the removal of soft tissue between the facial and lingual incisions. Over-retained and impacted primary teeth, i.e. 55, 54, 52, 51, 61, 64, 65, 71, 72, 75 and 85, were extracted. The bone layers covering the occlusal surfaces of the nonerupted teeth, i.e. 16, 26 and 36, were removed. The thinned flaps were finally replaced and sutured with continuous mattress technique using a 4-0 silk suture. During the surgical procedure, there was excessive bleeding. The treatment plan was modified in order to avoid excessive blood loss, and in the least affected sextant, the mandibular right, the impacted 85, was extracted, but the enlarged gingival tissues were not excised. Surgical excision in this area could be performed under local anaesthesia on another occasion. Following surgery, a 0.2% chlorhexidine rinse, twice a day after brushing for 2 weeks, was prescribed to reduce plaque formation [21].

The sutures were removed one week after the procedure. The postoperative healing was uneventful. Four weeks after suture removal, healing was satisfactory, oral hygiene had improved and the recorded plaque index was 0.65 [20].

The excised tissues were fixed in 10% formalin solution. The histological examination revealed proliferating dense fibrous connective tissue that was characterized by high cellularity, and numerous stellate and fusiform fibroblasts (Fig. 3). The epithelium showed hyperkeratosis and acanthosis with elongated papillae. There was little inflammatory infiltration.

The patient could not keep his scheduled recall appointments, although he presented for 6-month recall, he was lost to follow-up for the next 1-5 years. At the 6-month recall, no new teeth had erupted, and only teeth 53, 63, 73 and 74 were present. A panoramic radiograph revealed an improvement in the eruption direction on the previously ectopically erupting lower second premolars.

Two years postoperatively, there was no recurrence of the gingival enlargement, but none of the permanent teeth had erupted. The panoramic radiograph taken at that time (Fig. 4) showed very little progression in the eruption process of the permanent teeth; however, their roots showed slight development. The subject's mother refused a second-stage surgery to remove the enlarged tissues and the bony layers covering the permanent teeth in the mandibular right quadrant. A radiolucency in the crown of the unerupted 36 was indicative of preeruptive coronal resorption [22]. Careful inspection of the previous panoramic radiograph, taken 18 months earlier, showed that there had been a very small intracoronal radiolucency in 36 that had not been detected initially.

The subject, although pleased by the removal of the excessive gingival tissues, still had difficulty in mastication and was anxious to have "teeth". Because of the significant delay in the eruption of permanent teeth, overdentures were constructed for both arches, which were to be used until the eruption of the teeth (Fig. 5). Six months later, no recurrence of the gingival enlargement was noticed, but the permanent teeth had still not erupted (Fig. 5). The subject had grown much taller (158 cm, above
the fiftieth percentile for his age) and heavier (40 kg, above the twenty-fifth percentile), but still had microcephaly (head circumference = 49.3 cm).

Discussion

Several reports of gingival fibromatosis in childhood [5,8,9,12,22] and adolescence [9,17,23,24] have been published. This is an unusual case of an 11-year-old male, with gingival fibromatosis, microcephaly, impacted primary teeth and a significant delay in eruption of the permanent teeth. Microcephaly has not been previously associated with gingival fibromatosis. In a review of the literature, no other case of gingival fibromatosis was encountered with so many impacted primary and permanent teeth, and the complete absence of the eruption of the permanent teeth until the age of 13 years. A delay in the eruption of the teeth has been reported in individuals with this disorder who suffer from Rutherford syndrome and also have corneal dystrophy [4], but this subject did not have any sign of corneal dystrophy. Chromosome analysis proved to be normal and he did not suffer from any other recognizable syndrome. His case may be idiopathic since the patient has no family history for such disorder. Difficulties were encountered in the oral management of the severe gingival enlargement, the impacted primary and permanent teeth, the delayed eruption of both the primary and permanent teeth, the functional and aesthetic considerations, the partial anodontia and the subject's lack of compliance with suggested recalls. If such a severe case of gingival fibromatosis is left untreated, quite apart from the aesthetic concerns so important for an adolescent, it may also lead to severe impairment in mastication and speech [25].

Gingival fibromatosis is a disease that can be controlled with varying degrees of success [9]. When gingival enlargement is minimal, debridement of the
tooth surfaces and good oral hygiene may be sufficient to control the disorder [9]. However, in severe cases like this one, surgical excision may be necessary, along with restoration of the gingival contours [25]. For patients with large-scale lesions such as this subject, treatment should be ideally performed in two or three stages at intervals of 4–5 months [9,11,23,25]. The lower right sextant was left to be treated at another time on this basis.

Many techniques have been used for the excision of the enlarged gingival tissues, including: an external or internal bevel gingivectomy [12,23,26,27]; an apically positioned flap [15]; electrocautery [28]; and a carbon dioxide laser [14,29]. In the present case, an apically positioned flap was selected over gingivectomy to facilitate access to the alveolar bone in order to extract teeth and remove the bone layers covering the occlusal surfaces of the impacted teeth.

There is no consensus among authors regarding the timing for surgery [12]. Some clinicians have suggested that the best time to perform surgery is when all the permanent teeth have erupted [25,30]. In the present case, the enlarged gingival tissues were excised prior to the eruption of the permanent teeth because of the compromised aesthetics and in order to facilitate eruption. The eruption of the teeth could be impaired by the dense fibrous tissue [9].

Recurrence of the gingival enlargement is common over varying periods [23,25], and is most often seen in children and adolescents rather than older patients [24]. However, whether and when it will occur is not predictable [8,25]. Recurrence has been associated with dental plaque accumulation, and has been reported to occur from as soon as 3 months [16] to as late as 14 years postoperatively [26], but is expected to be minimal if surgical excision is performed after the eruption of the permanent dentition [8]. Its extent in this patient is not known because localized gingivectomies had been performed twice in the past elsewhere. Since the last surgical intervention, no recurrence of the gingival enlargement has been noticed for 30 months postoperatively. Although this could still occur, the absence of recurrence in the present study is in accordance with some previous reports. No recurrence was reported in a 28-year-old woman 2 years postoperatively [23], and in one 5-year-old girl [13], this was the case up to 36 months postoperatively. The longest follow-up with no recurrence was 14 years [7].

Delay in the eruption of primary teeth has rarely been associated with gingival fibromatosis in previous reports [21,22]. However, a delay of several months in the eruption of permanent teeth and in the exfoliation of primary teeth has been reported in some patients with gingival fibromatosis [5,8,16,17]. In this case, not only were there impacted primary molars at the age of 11 years, but there was severe delay both in the exfoliation of primary teeth and the eruption of permanent teeth, which continued up to the age of 13.5 years, when none of the permanent teeth had yet appeared in the oral cavity. The eruption might have been inhibited by the dense fibrous gingival tissues covering the teeth after treatment, and could have been facilitated if a surgical dressing had been put in place after the primary tooth extractions, thereby keeping the passage free for the eruption of the permanent teeth [31–33]. However, no surgical dressing was placed in the present case since there were no teeth to stabilize one. Severe delays in both the exfoliation and in the eruption processes have previously been reported in an 11-year-old adolescent with gingival fibromatosis [17]; by the age of 15 years and 10 months, all this subject’s permanent teeth had erupted, except the second and third permanent molars, after multiple extractions of primary teeth.

Because of a significant delay in tooth eruption and the existence of only four primary teeth in the oral cavity (53, 63, 73 and 74) at the age of 13 years, overdentures were constructed in order to improve aesthetics and function. There are no other reports in the literature regarding the prosthetic rehabilitation of individuals with gingival fibromatosis, but in most other cases, when the primary teeth had exfoliated, the permanent teeth erupted soon after and the child was not left edentulous for a long period.

The occurrence of preeruptive coronal resorption [34] in the crown of the impacted 36 may be an incidental finding since there have been no other reports associating this entity with gingival fibromatosis. However, preeruptive coronal resorption is a progressive lesion, and early intervention is essential to prevent crown destruction and pulpal involvement [21]. In this case, tooth 36 is being closely monitored since endodontic involvement may occur, and even upon partial eruption, there should be rapid intervention to either restore or carry out endodontic treatment, depending on the extent of the lesion.

The future treatment plan includes monitoring the eruption process of the teeth and adjustment of the overdentures, restorative or endodontic treatment of
36, surgical excision of the enlarged soft tissues in the mandibular right quadrant, and if needed, removal of bone overlying the teeth in the same quadrant and in the mandibular anterior. Extrusion of impacted teeth may be further facilitated by orthodontic treatment, after the eruption of a few teeth which may be used as anchorage. If the teeth do not erupt, the use of implants may be considered, after the extraction of the impacted teeth.

Early diagnosis of gingival fibromatosis is imperative for maintaining optimum gingival health, monitoring the eruption of permanent teeth, improving oral function and aesthetics, and reducing psychological effects. Long-term follow up is necessary in patients with hereditary gingival fibromatosis because of the risk of recurrence, which is not predictable. Treatment of such cases involves close collaboration between the practitioners of several dental specialties.


Der Patient wurde vorgestellt mit stark vergrößertem Gingivawebewebe in beiden Kiefern und multiplen retinierten bzw. nicht durchgebrochenen Milchzähnen. Lokalisierte Gingivektomien waren bereits im Alter von 7 und 9 Jahren vorgenommen worden. Es lag kein bekanntes Syndrom vor, die Familienanamnese war unauffällig.


Dreißig Monate nach Therapie war kein Rezidiv der Gingivavergrößerung erkennbar, es war aber auch keine Eruption eines bleibenden Zahnes erfolgt.

Resumen. El objetivo de este caso clínico fue presentar el tratamiento dental de un caso inusual de fibromatosis gingival idiopática con múltiples dientes primarios impactados y ausencia de la erupción de dientes permanentes en un niño de 11 años y a los 30 meses de seguimiento.
El paciente presentaba tejidos gingivales severamente agrandados afectando a ambas arcaras y múltiples dientes primarios retenidos y no erupcionados. Ya había estado sujeto a ginvectomías localizadas a los 7 y 9 años. No tenía síndrome conocido y no había una historia familiar de una alteración similar. El paciente se trató con anestesia general para eliminar el exceso de tejido gingival usando colgajos posicionados apicalmente.

Durante el procedimiento quirúrgico se extrajeron los dientes primarios retenidos e impactados para facilitar la erupción de los sucesores permanentes. A los dos años del postoperatorio no hubo recidiva del agravamiento gingival, se construyeron entonces dentaduras porque ninguno de los dientes permanentes había erupcionado todavía.

Además, se detectó radiográficamente reabsorción coronal pre-erupción afectando a la corona del #36 no erupcionado. En los 30 meses de postoperatorio no se vio recidiva del agravamiento gingival pero todavía no había erupcionado ningún diente permanente.

References
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