Hereditary gingival fibromatosis with distinct dental, skeletal and developmental abnormalities

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Abstract

A case of a 9-year-old child with hereditary gingival fibromatosis, supernumerary tooth, chest deformities, auricular cartilage deformation, joint laxity and undescended testes is described. The exact mode of inheritance is unclear; a new mutation pattern is possible. These features resemble but differ from the previously reported Laband syndrome. The dental treatment consisted of surgical removal of the fibrous tissue and conservative restorative treatment under general anesthesia. The dental practitioner should be alert for developmental abnormalities such as supernumerary teeth and delayed tooth eruption. A comprehensive medical history and physical systemic evaluation is essential to rule out other systemic abnormalities. Genetic consultation is mandatory for future family planning. (Pediatr Dent 24:253-256, 2002)

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idiopathic or hereditary gingival fibromatosis is a rare condition of the gingival tissues, characterized by enlargement of the free and attached gingiva. The clinical presentation is generalized firm nodular enlargements with pink to red and inflamed, smooth to stippled surfaces, with little tendency to bleed. In some cases the gingiva can become so firm and dense as to feel like bone on palpation. The enlargement is painless and may extend up to the mucogingival junction but does not affect the alveolar mucosa.1-4

The histological appearance shows hyperplasia of fibrous tissue of the corneum. The tissues are composed mainly of dense connective tissue, which is rich in collagen fibrils, but contains only a few fibroblasts. The overlying epithelium is normal, but is slightly hyperplastic in some areas, with rete pegs into the corneum. 5 Gingival enlargement may occur during the eruption of primary teeth and affect both dentitions, but does not occur once growth of the patient has ceased.1-4 Nevertheless, there are few persons who continue to experience slow enlargement into adult life.1

The fibrous enlargement is usually symmetrical but may be unilateral and generalized or localized. The localized form usually affects the maxillary molar and tuberosity area, particularly on the palatal surface. When there is severe involvement, teeth are almost completely covered3,6 and delayed eruption and displacement of teeth can occur.1-3 In a study of few families affected with hereditary gingival fibromatosis, no linkage with HLA, antigen was observed.7

The condition is most frequently reported to be transmitted as an autosomal dominant trait, and recently at least two gene loci on the short arm of chromosome 2 that are responsible for gingival fibromatosis were identified in a Brazilian family. One locus was located in 2p21-2p22 and the other was located more proximally in the region of 2p13-2p16.5,9 Xiao et al identified a new locus (GINGF2) located on chromosome 5q13-q22.10

Gingival fibromatosis can be caused by number of factors, including inflammation, leukemic infiltration and use of medications such as phenytoin, cyclosporine or nifedipine4 and vigabatrin.11 Gingival enlargement can be associated with other features as part of a syndrome such as the Laband, Rutherford, Ramon or Cross syndrome.12 Association with hearing loss and supernumerary teeth, cherubism and psychomotor retardation and with prune-belly syndrome was also reported.13,14 Other features include abnormal fingers, nose and ears, splenomegaly, aspartylglucosaminuria and gangliosidosis.15 The treatment recommended is gingivectomy and good oral hygiene, but recurrences are common.4
A 9-year-old boy of Ashkenazi origin (Eastern-European) was referred by his dentist to The Oral Medicine Clinical Center at The Maccabi Health Organization in Tel Aviv, due to dense gingival tissue that was preventing tooth eruption. On examination, his weight was 31 kg and his height 136 cm, matching his chronological age (75th percentile). The physical examination revealed absence of cartilage in the auricular region, an asymmetrical chest in the form of right “pigeon chest “ (prominence of the sternum), undescended right testes and extended joint laxity. The medical history was non-contributory for other systemic diseases or use of medications. Mental development was normal.

The oral examination revealed a mixed dentition with poor oral hygiene, a prominent anterior open bite and dense coarse gingival tissue (Figs 1 and 2). Teeth #D and #E were still present with marginal gingivitis. Tooth #F had exfoliated and #G was present with a large carious lesion. The roots of both maxillary lateral incisors showed no advanced resorption. In the mandible, partial eruption of both permanent central incisors and #26 was noted, while #23 had not erupted yet. The roots of the first permanent molars were almost fully developed, but the teeth were totally entrapped in a gingival mass of tissue, thus impairing their eruption.

An unerupted supernumerary tooth was found between the roots of the mandibular right canine and lateral incisor. The clinical and radiographic findings indicated a gross delay in tooth eruption (Fig 3). The permanent maxillary central incisors and first permanent molars were still unerupted. Under general anesthesia, using CO₂ laser, surgical removal of dense fibrous tissue was performed exposing the teeth crowns. Conservative restorative treatment was performed as well. The patient tolerated the procedure well with no postoperative complications. After recovery, oral hygiene instructions were reinforced and complete tissue recovery was evident after 3 weeks. Partial recurrence of the gingival overgrowth was noted after 3 months. At this stage, professional prophylaxis was performed by a dental hygienist. At the six months evaluation, further eruption of all first permanent molars to the level of the second primary molars was noted (Fig 4). The need for a second surgical intervention was discussed with the parents.

Genetic consultation suggested an isolated case with new mutation but not autosomal recessive mode, since no other members of the family were involved. The skeletal abnormalities, together with the retarded eruption of the permanent teeth, might point to a defect in collagen breakdown and/or bone resorption. However, no signs of osteopetrosis or pycnodysostosis were noted. The family refused chromosomal study, so the exact mode of heredity remained undetermined.
Delay in tooth eruption is a common finding related to gingivofibromatosis and Laband Syndrome. Joint hypermobility, absence of cartilage in the auricular area, resembles Laband Syndrome, which manifests gingival fibromatosis with defects of the ears, nose, bones, nails and terminal phalanges, hypervensitive joints and characteristic facies. The presence of supernumerary teeth associated with gingival fibromatosis was also reported, however, not in Laband syndrome. Asymmetrical chest and undescended testis are findings that were not described in the literature in association with gingival fibromatosis.

Hereditary gingival fibromatosis is associated with either autosomal dominant such as Rutherford and Laband syndromes, or autosomal recessive inheritance as in Murray-Puretic-Drescher, Cross, Ramon and lysosomal storage disease. Rutherford syndrome consists of mental retardation, aggressive behavior, dentigerous cysts associated with congenitally enlarged gingivae and delayed tooth eruption. Murray-Puretic-Drescher syndrome is characterized by gingival fibromatosis with multiple juvenile PAS-positive hyaline fibromas of the head, flexion contractures, mental retardation and elevated urinary hyaluronic acid.

Cross described gingival and alveolar enlargement, microphthalmia, cloudy corneas, hypopigmentation and atheosis. In Ramon syndrome, gingival fibromatosis is associated with cherubism, mental retardation, epilepsy and juvenile rheumatoid arthritis. In his comprehensive review study, Hart et al presents a table with more than 18 different genetic forms of gingival fibromatosis with various systemic manifestations.

In the present case, some features of Zimmerman-Laband syndrome were present such as joint laxity and auricular cartilage defects. However, new findings such as chest deformity and undescended testes and supernumery tooth were also found. Splenomegaly and hepatomegaly were not present in this patient. Since no chromosomal study was done, we could not conclude if the present case is truly a variation of Laband Syndrome or if it represents a new one.

The surgical technique used for treatment in this patient was a CO₂ laser. The advantages of using the CO₂ laser rather than the scalpel in the surgery for gingival lesions are the ability of the laser to coagulate and seal blood vessels, vaporize the tissue, make an accurate incision and improved the healing effect due to its antimicrobial properties.

The effect of oral hygiene and the superimposition of plaque accumulation on the prognosis of gingivofibromatosis are crucial. Partial compliance of the patient may be in part responsible for the recurrence of the overgrowth.

Substantial tooth eruption was noted in the molar area after 6 months post-treatment. Excision of the dense fibrous tissue facilitated the further eruption.

The dental professional should be alert for the possibility of gingival fibromatosis being part of a genetic syndrome especially in cases where genetic family consultation might be mandatory for future family planing.