題名:Familial gingival enlargement- a case report.

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上傳時間:2009-11-23T02:57:57Z

摘要:BACKGROUND: Gingival fibromatosis (GF) is characterized by fibrotic enlargement of the gingiva that can be inherited as an isolated trait (named hereditary gingival fibromatosis) or as a component of a syndrome. This article reports one kindred affected by a syndrome characterized by GF associated with dental abnormalities (DA) including generalized thin hypoplastic amelogenesis imperfecta (AI). METHODS: To characterize the pattern of inheritance and the clinical features, 70 family members were examined. Hematoxylin and eosin staining, immunohistochemistry, and scanning electronic microscopy (SEM) were performed to identify the alterations on gingiva, teeth, and dental follicles. RESULTS: Examination of the family pedigree demonstrated multiple consanguineous first-cousin marriages and an autosomal recessive trait of inheritance. Four members demonstrated mild GF in association with DA, including generalized thin hypoplastic AI, intrapulpal calcifications, delay of tooth eruption, and pericoronal radiolucencies involving unerupted teeth. One of those four patients also had mental retardation (MR). MR as an isolated feature was observed in six members, whereas isolated GF was found in one individual. A combination of gingivectomy and gingivoplasty followed by regular dental procedures were performed in these patients. Histologic examination of the gingival enlargement revealed a dense connective tissue containing myofibroblasts, islands of odontogenic epithelium, and calcified psammomatous deposits, which resembled cementicle-like structures by SEM. Pericoronal lesions also showed calcified psammomatous deposits in

association with islands of odontogenic epithelium. Enamel ultrastructure analysis revealed normal surface alternating with irregular and porous areas. CONCLUSION: To the best of our knowledge, these cases represent a new syndrome within the spectrum of those including GF.