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Hereditary Gingivo-Alveolar Hyperplasia: A Report of Two Siblings

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Gingival hyperplasia is characterized by fibrotic gingival overgrowth. The lesion may bury all the crown of the teeth and lead to impairment in masticatory functions and aesthetic disfigurement. This inherited disease is considered rare. We presented two cases of gingival hyperplasia in two siblings: an 11-year-old girl and an 8-year-old boy, whose mother had also suffered from the disease. The two siblings presented with generalized gingival overgrowth involving the maxillary and mandibular arches and covering almost all of the teeth. We performed surgery to reduce the excessive gingivo-alveolar tissue and disclosed most of the teeth. The patients showed functional and aesthetic improvement. The last follow-up through a phone call, which was conducted 12 months after the surgery, revealed no recurrent hyperplasia.

Key words: Gingiva – Gingival hyperplasia – Gingivectomy – Mastication

Gingival hyperplasia is a rare disease which affects one in 750,000 people.[1] It is characterized by localized or generalized enlargement of the gingiva[1] and causes impairment of speech function, mastication, occlusion, and aesthetics.[1–4] Gap between two teeth, malposition of the teeth, prolonged retention of primary dentition, and delayed eruption of permanent dentition have also been reported.[1,5]

We conducted a literature search in PubMed on March 15, 2013, using Boolean operator OR, the field of title, under the keywords "gingival hyperplasia," "drug induced gingival hyperplasia," "gingival overgrowth," "gingival fibromatosis," "hereditary gingival fibromatosis," and "idiopathic gingival fibromatosis." Only publications in English were retrieved to have the number of cases that have been reported. Our unpublished study showed there were 1096 cases reported. This number was very low compared to the total world population, which was 6.8 billion people in 2010.[6] When we compared it to other rare craniofacial syndromes such as Crouzon syndrome (1:65,000),[7] that number was still 10 times lower. Considering that fact, we were keen

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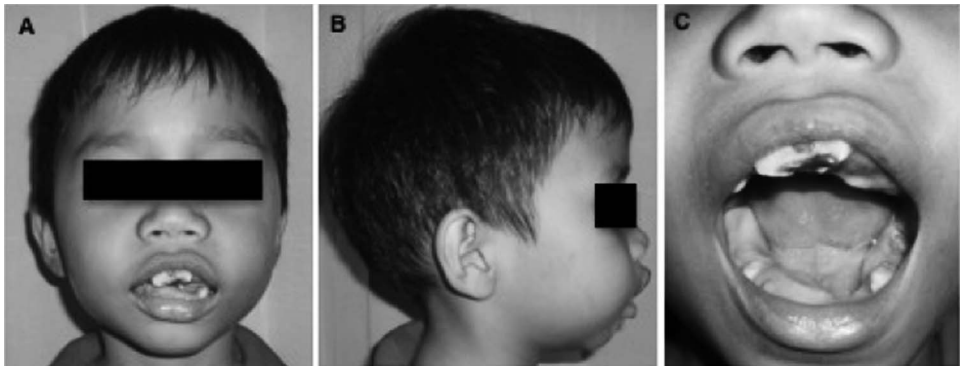

Fig. 1 Preoperative view of an 8-year-old boy with gingival tissue enlargement of both upper and lower arches (A). Profile photo showed maxillary protrusion and mandibular retrusion (B). There were crusts on central incisor, presumably resulted from the previous surgery (C).

to report our cases of gingival hyperplasia in two siblings whose mother had also suffered from the disease. This was intended to enrich the cases reported in the literature.

Case Series

Case 1

An 8-year-old boy presented with enlargement of both upper and lower arches of gingival tissue. The early sign of enlargement occurred during the eruption of his first incisor at age 1 year. The swelling progressed involving both gingival arches. Despite undergoing normal physical growth, the boy suffered from masticatory functional impairment, which allowed him to eat soft diet only. The patient had had a surgery by another surgeon 3 weeks before his presentation. Apparently, the surgery only uncovered part of the upper central incisors (Fig. 1). There was no history of drugs, systemic diseases, and syndromes associated with the enlargement. His profile showed maxillary protrusion and mandibular regression. It is likely that his sister and mother presented with the same profile, despite the fact that the mother had already underwent surgery 20 years in advance without any sign of recurrence.

In the upper jaw, the gingival tissue covered both the canines and the lateral incisors. Part of the central incisors, first molars, and second molars were visible. In the lower jaw, the crowns of the left premolar, the first molar, and the right premolar were partially visible. The gingival tissue was firm and dense.

Case 2

The boy's sister, an 11-year-old girl, also had the same gingival condition. The first enlargement appeared 5 years before presentation when her permanent teeth erupted. Nevertheless, it progressed slowly. She had low self-esteem. Her obvious gum made her feel embarrassed, and it was even worse when her peers found her eating only soft diet. Intraoral examination revealed generalized enlargement of the gingiva of both

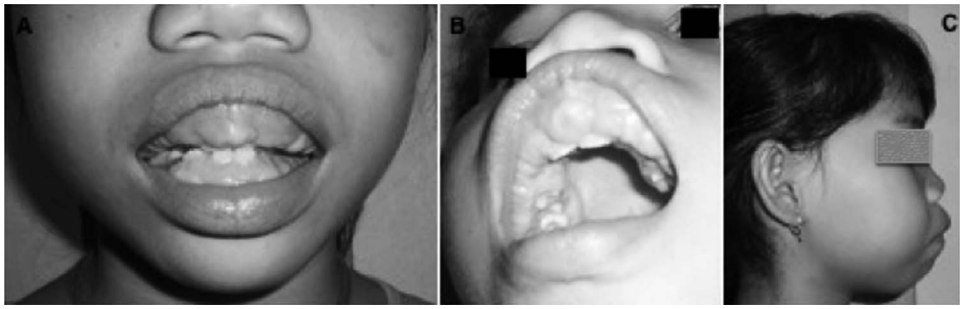

Fig. 2 Preoperative view of an 11-year-old girl with gingival tissue enlargement of both upper and lower arches (A+B). Profile photo showed maxillary protrusion and mandibular retrusion (C).

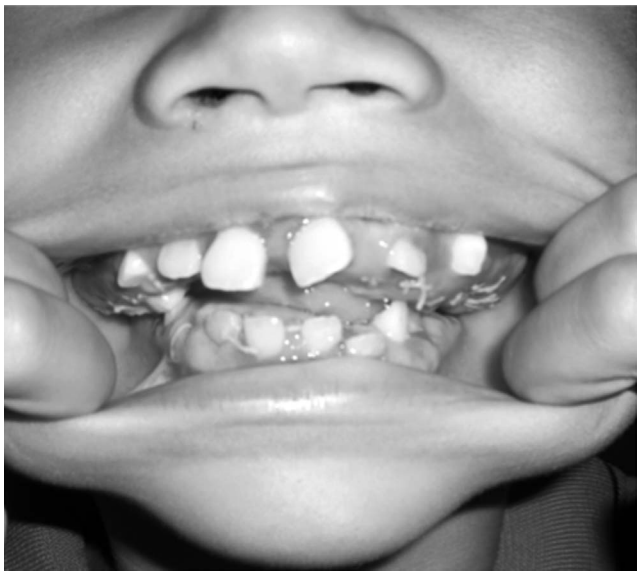

Fig. 3 The boy presented 9 days after surgery. Most of the anterior teeth were uncovered with good oral hygiene. Some stitches are still visible at the molar areas.

arches without any inflammatory signs. In the upper jaw, the gingiva covered almost all of the teeth except the hidden first incisors. In the lower jaw, the first premolar and the right canine were buried, while the same situation applied for the lateral incisor, the canine, the first premolar, and the second molar on the left side (Fig. 2).

The senior author performed gingivectomy on both siblings under general anesthesia. There was no particular design for the procedure. The "cut as you go" principle was the way to reduce the excessive gingivo-alveolar tissue to uncover the teeth. In some difficult areas, which could not be closed primarily, the gingival surface was left for secondary healing by applying polyvinylpyrrolidone gel 3 times a day. Additionally, the patients were also instructed to gargle polyvinylpyrrolidone 3 times a day. Patients showed uneventful healing and, at the utmost, functional and aesthetic improvement. The last follow-up through a phone call was conducted 12 months after the surgery, and the parents did not report any recurrent hyperplasia. They were satisfied with the current outcome. The parents also reported briefly about the girl's increasing self-confidence.

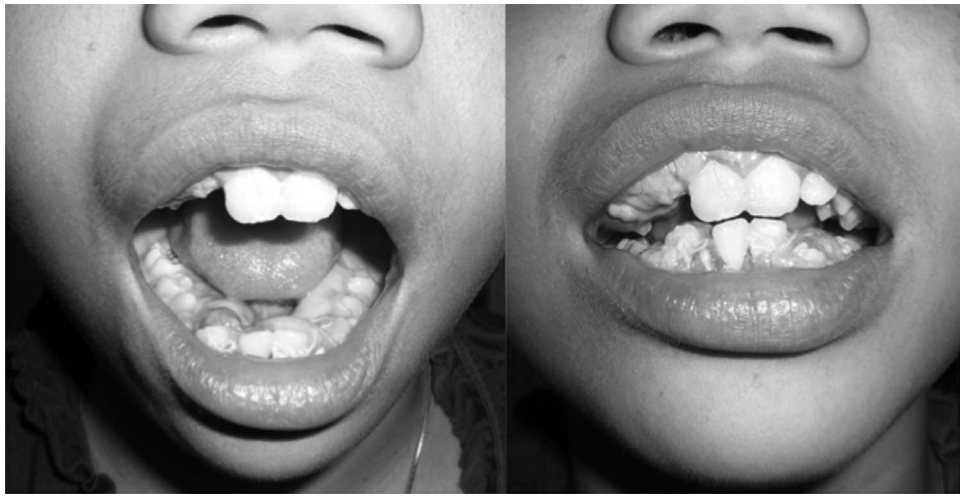

Fig. 4 The girl presented 3 weeks after surgery.

Significant improvement was shown. Although some molar crowns at the upper right jaw were not visible yet, the gum has been significantly reduced.

Histopathologic examination of the gingival tissue revealed hyperplastic squamous epithelium with chronic inflammatory cells on the fibrotic lamina propria. The alveolar bone showed fibrotic tissue with chronic inflammatory cells. The final diagnosis was fibro-inflammatory hyperplasia of both gingival and alveolar tissue.

Discussion

Early signs of gingival enlargement had been observed during the eruption of the deciduous teeth in the boy and permanent teeth in the girl. The enlargement was slow, progressive, and involved both jaws. This is in line with previous publications where hereditary gingival hyperplasia is characterized by slow, progressive enlargement of the gingiva, which usually begins with the eruption of deciduous or permanent teeth.[1,4,5,8]

The gingiva is usually pink, firm, and does not bleed easily.[1,4,5,8] The cases presented in this report could be categorized as severe since almost all of the crowns of the teeth were buried in. However, it did not bleed profusely during the surgery although it was not easy to inject vasoconstrictor. Pseudo-pocketing was found in the upper incisors of the girl as shown in Fig. 2, but there was no periodontal problems as expected. Usually, periodontal problems does not involve the alveolar bone.[4]

In these cases, gingivectomy was done after the eruption of permanent dentition. There was no recurrence reported at 1 year following the surgery. Although a longer follow-up is needed to detect any recurrence, it does not seem to be a serious problem as it is reported in the literature that the risk of recurrence is higher if the surgery was done before eruption of all permanent dentition.[1] Besides, the mother also did not have recurrence 20 years after her surgery. The patients showed significant improvement of mastication function and aesthetics.

Several etiologies of gingival hyperplasia are known, such as drugs (phenytoin, cyclosporine, nifedipine, amlodipine), genetics, inflammation, systemic disease, hormones, (in pregnancy), and idiopathic.[1–5,8–15] Gingival hyperplasia can also be an isolated disease affecting only the gingiva or a part of a syndrome, such as Murray-Puretic-Drescher syndrome, Zimmermann-Laband syndrome, Rutherford syndrome, Jones syndrome, Cross syndrome, and Ramon syndrome.[1,3–5,13–18] This report does not seem to be related to any syndrome without clear information in regards to etiology except hereditary involvement. Unfortunately, we did not get any data to confirm the genetic inheritance beside familial relationship. The pathogenesis of the gingival hyperplasia remains unclear. A recent study identified mutation in the son-of-sevenless (SOS-1) gene as a potential pathogenesis mechanism.[4,8] Gingival hyperplasia can be inherited in autosomal recessive genes, but mainly in autosomal dominant.[1,3–5]

The current report lacks information on the dentition, which was still covered after the surgery as well as the status of possible remaining primary teeth. No X-ray data is available. It is also unfortunate that the patients live far away in a remote province without the funds to travel and to get further examination. The patients were supported with limited funds granted by a solo donor when they underwent surgery at our unit.

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