Case Report

Multiple Unerupted Teeth with Amelogenesis Imperfecta in Siblings

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Abstract

Amelogenesis imperfecta encompasses a group of inherited abnormalities that are generally considered to primarily affect the formation and/or calcification of enamel. This case report describes the unusual presentation of amelogenesis imperfecta in siblings as multiple unerupted teeth, multiple pulpal calcifications, and multiple dilacerations of roots along with the defect in the enamel. The intent of our report is to highlight a rare co-occurrence of amelogenesis imperfecta with multiple morphologic alterations in siblings.

Keywords: Amelogenesis imperfecta, impacted teeth, unerupted teeth

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Introduction

Amelogenesis imperfecta (AI) encompasses a group of hereditary diseases that involve the defective formation and/or calcification of enamel.[1] Presence of enamel defects without any evidence of generalized or systemic defects is pathognomic feature of AI.[2] Generally, both the primary and permanent dentitions are involved.[3]

Nonenamel anomalies such as delayed eruption, crown resorption, pulpal calcifications, dental follicular hamartomas, and gingival hyperplasia were found to be associated with AI.[4,5] Only few cases reported in the literature have discussed about the association of AI and delayed eruption.[3,6] The aim of this report is to highlight the rare presentations of AI with multiple morphologic dental alterations and periodontal changes in siblings.

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Case Reports

Case 1

A 20-year-old female patient reported to the department with the complaint of missing teeth. The patient gave a history of discolored primary teeth which exfoliated 8–10 years before the date of reporting. Medical history was non-contributory. She did not report of any major illness or prolonged hospitalization and medication. She had earlier visited a dentist for the root canal treatment of the mandibular right first molar and restoration of the maxillary left first molar. A detailed family history revealed that the patient's younger brother also had similar dental abnormalities. There is no history of consanguineous marriage between the parents.

Intraoral examination revealed erupted teeth with respect to 17 (partially erupted), 16, 12, 11, 21, 26, 27 (partially erupted), 36, 31, 41, and 46. Generalized gingival hyperplasia was observed. There was generalized attrition of the cusp tips and incisal edges leading to shortened cervico occlusal length and yellowish white smooth occlusal surface of the teeth [Figure 1a, b]. Intraoral periapical radiograph of the maxillary first molar region revealed the loss of enamel cap [Figure 1c]. Owing to multiple unerupted teeth, a panoramic radiograph was made and multiple impacted teeth with respect to 18, 15, 14, 13, 23, 24, 25, 28, 37, 35, 34, 33, 32,

42, 43, 44, 45 and 47 were noticed. Panoramic radiograph also showed other abnormalities such as generalized loss of enamel cap, multiple pulp calcifications, and multiple dilacerations in the apical third of impacted and erupted teeth [Figure 1d].

Clinical and radiological findings led to the diagnosis of hypoplastic type of AI. Treatment included thorough oral prophylaxis, rehabilitation with over denture prosthesis.

Case 2

Patient's younger brother was a 17-year-old male, who reported to the department with the complaint of missing teeth. Patient's medical history was unremarkable. He had visited dentist for restoration of the maxillary right first molar and extraction of retained deciduous tooth in the maxillary anterior region. Extraoral examination did not reveal any relevant findings.

Intraoral examination revealed erupted teeth with respect to 16, 12, 11, 21, 22, 24, 26, 36, 35, 34, 32, 31, 41, 42, 44, 45, and 46. Retained deciduous teeth with respect to 54, 55, 63, 65 and 83 were noticed. There was generalized attrition of the crown with yellowish white smooth surface of the teeth [Figure 2a, b]. Panoramic radiograph revealed generalized loss of enamel cap, multiple unerupted teeth, and pulp calcification with 17 and 26 [Figure 2c].

The above findings led to the diagnosis of hypoplastic type of AI. Interdisciplinary approach was suggested for the treatment of the patient. Unfortunately, due to certain logistical reasons patient could not keep up with appointments.

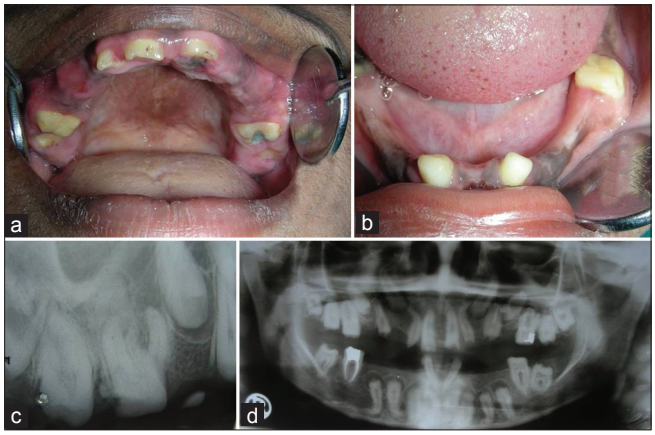

Figure 1: (Case 1) (a and b) Clinical image showing hypodontia in the maxillary and mandibular arch; also observe yellowish white smooth occlusal surface of the erupted teeth. (c) Intraoral periapical radiograph showing loss of enamel cap and lack of contrast between enamel and dentine. (d) Panoramic radiograph showing multiple impacted teeth, multiple pulp calcifications, and multiple acute dilacerations in the apical third of impacted and erupted teeth

Discussion

AI is a developmental condition of the dental enamel that shows autosomal dominant, autosomal recessive, sex linked inheritance patterns and sporadic cases.[7] Based on enamel appearance and hypothesized developmental defects, AI is classified into four patterns: hypoplastic, hypomaturation, hypocalcified, and hypomaturation-hypoplastic.[3]

Dental features associated with AI include qualitatively and quantitatively deficient enamel. It also associated as a spectrum of other features such as progressive root and crown resorption, pulpal calcification, taurodontism, malformation of root, impaction of permanent teeth, and congenitally missing teeth.[6] In our cases, features such as tooth impaction, delayed eruption, pulp stones, and dilacerations were noticed. Few authors have reported about the association of AI with gingival hyperplasia and follicular lesions.[1,5] Very sparse literature is available associating unerupted teeth with AI.[1,3,6] In our case, unerupted teeth also involved premolars, which have been not reported in the literature till date. Unerupted premolars were associated with enlarged follicular space in case 1.

Numerous etiologies have so far been suggested for the lack of eruption of teeth in AI patients. These include abnormal molecular control of the eruption process, lack of space, and concurrent follicular enlargement.[6] Both of our cases had multiple unerupted teeth in either arches. Sixteen unerupted permanent teeth were observed in case 1, and 13 unerupted permanent teeth were observed in case 2. AI patients have six times more tendency

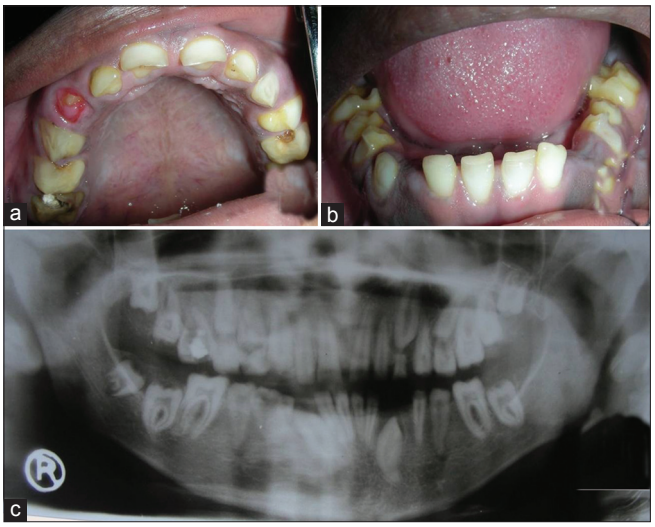

Figure 2: (Case 2) (a and b) Clinical image showing erupted teeth in the maxillary and mandibular arch with smooth surface and yellowish color. Also note the presence of multiple retained deciduous teeth. (c) Panoramic radiograph showing multiple unerupted teeth, generalized loss of enamel cap, retained deciduous teeth, and pulpal calcifications

than unaffected people to have impaction of permanent teeth.[3,6] In the literature, only 15 cases of delayed eruption, 14 cases of pulp stones, and 9 cases of tooth agenesis have been reported.[3] It was reported that pulp stones are formed as a result of the external local irritation because of the thin enamel layer and attrition. In our report, five teeth with pulp stones were observed in Case 1, and two teeth with pulp stones were observed in Case 2. Dentinal hypersentisity, vertical dimension, and poor esthetics are frequently encountered clinical problems in AI.[8] Loss of vertical dimension and poor esthetics were also observed in our cases. Dental radiographs contribute significantly to the diagnosis of AI. Lack of contrast in the radiopacity between enamel and dentine is a characteristic radiographic observation in AI.[4] Presenting cases showed lack of radiographic contrast between enamel and dentine and generalized loss of enamel cap.

Since the treatment consideration for AI requires multidimensional outlook of patient's socioeconomic status and severity of the disease, a combined multidisciplinary approach has been advised by many authors.[8] The management of esthetics and function is with prosthetic approach in a patient with AI and tooth impaction.[3]

The aim of our report is to highlight clinical and radiological features of an atypical presentation of AI in siblings. Rare features such as of unerupted premolars, hyperplastic follicular space, and gingival hyperplasia have been described in our report. We also describe a multidisciplinary approach of this rare condition.

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