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Research Article

FAMILIAL AMELOGENESIS IMPERFECT IN SIBLINGS- A REPORT OF TWO CASES

Navya M K*., Sujatha G P., Poornima R and Ashok L

Department of Oral Medicine & Radiology, Bapuji Dental College & Hospital Davangere

A R T I C L E I N F O

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A B S T R A C T

Developmental anomalies are frequently encountered during our clinical examinations. Among these anomalies is Amelogenesis Imperfecta (AI) which though is rare among the dental anomalies yet causes great physical and psychological discomfort to the patients. Here we are reporting a case of AI in siblings. The diagnosis of the disease was carried out based on family history, clinical and radiographic examination.

INTRODUCTION

Amelogenesis Imperfecta (AI) is a hereditary disorder causing developmental alteration in the enamel of the teeth. The defect could be in the formative stage or maturation and calcification stage of the enamel. This disease usually affects both the primary and permanent dentition. The frequency of AI in population varies between 1:718 and 1:14000 in western population.[1] The prevalence of AI in Indian population is about 0.27% which is very rare among the other developmental dental anomalies. [2] The defect in enamel leads to hypersensitivity, loss of vertical height in occlusion, malocclusion, delayed eruption, aesthetic problems etc. Patients are usually more concerned about the aesthetics and so a proper treatment planning is necessary for this disorder.

Case report

Case-1

A 19 year old female patient came with complaint of pain in her lower right back tooth since 1 week. Pain was sudden in onset, intermittent, moderate, aching type, localised, aggravated on chewing food and relieved by taking medication. Patient also revealed that she has yellowish discoloration of teeth since childhood. Her medical history was non-contributory and past dental history revealed that she had consulted a dentist 1 year back for pain in right lower back tooth and got it restored. A detailed family history revealed that the patient's younger brother also had similar dental abnormalities. There was no history of consanguineous marriage between the parents. On intraoral examination, complete compliment of permanent teeth were present except for 18 and 28. Generalized yellowish discolouration of the teeth was evident with rough enamel surface appearance. Pitting was evident on the surface of the crown. Retained deciduous tooth portion of 75 was evident buccal to 35. Partially erupted 38 & 48. Class two cavity was evident in mesio-occlusal aspect of 46. Severe attrition of the buccal aspect of crown was evident wrt 34,36,37,44,46 & 47. (fig-1)

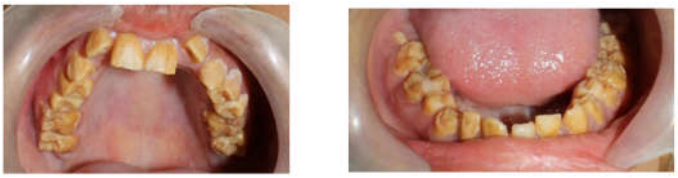

Figure 1 complete compliment of teeth showing yellowish discoloration and roughness of the surface of enamel, severe attrition of posteriors, retained deciduous 75.

Her occlusion was Angle's class II bilaterally with anterior deep bite. Overjet and overbite was reduced. (fig-2)

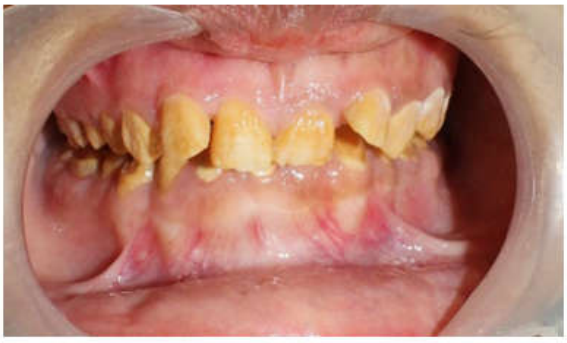

Figure 2 Anterior deep bite evident with reduced overjet.

Corresponding author:* **Navya M K
Department of Oral Medicine & Radiology, Bapuji Dental College & Hospital Davangere

On palpation of the teeth, generalized surface roughness was evident. No chipping of the enamel was evident on probing. 46 was tender on percussion. Based on the history and clinical findings a provisional diagnosis of acute apical periodontitis wrt 46 and amelogenesis imperfecta hypoplastic type and Angle's class II malocclusion was given.

Intraoral periapical radiographs wrt upper and lower incisors as well as 46 and bitewing radiographs wrt upper and lower posteriors were advised. Radiographs revealed, altered morphology of crown giving it a square shape due to loss of contact between the teeth with decreased density of enamel. Disto-occlusal and mesio-occlusal radiolucency was evident involving enamel, dentin and pulp wrt 46 and mesio-occlusal radiolucency involving enamel and dentin was evident wrt 47. Loss of lamina dura and PDL space widening was evident at apical third of the roots with diffuse periapical radiolucency on the mesial root wrt 46. (fig-3)

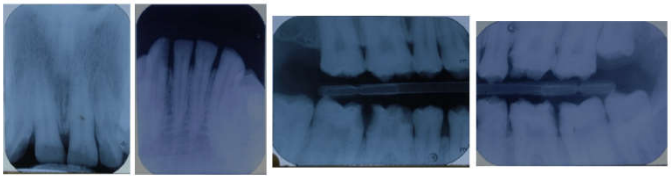

Figure 3 IOPA and bitewing radiographs showing irregular enamel surface, thin peripheral enamel, loss of contact between teeth, square shaped crowns, proximal radiolucency involving enamel. Dentin & pulp wrt 46 and periapical radiolucency wrt 46.

Digital OPG revealed, similar features of generalized reduced density of enamel, thining of enamel and irregular surface of the occlusal aspect of teeth. (fig-4) Based on the radiographic findings, a final diagnosis of chronicperiapical abscess wrt 46 and Amelogenesis Imperfecta hypoplastic type was given.

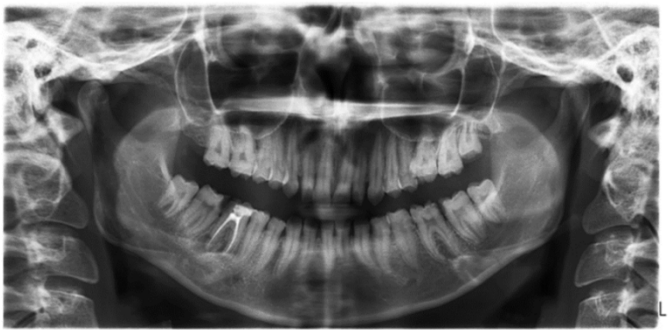

Figure 4 Digital OPG showing reduced enamel thickness and density with altered crown morphology

Case-2

A 17 year old male patient reported with complaints of yellowish discoloration of teeth. History of present illness revealed that his teeth were this colour since it erupted in the oral cavity. His past dental history revealed that he had undergone restoration of teeth 3 months back from a clinic in Gangavathi. His past medical history was non-contributory. On extraoral examination, no gross facial asymmetry was evident. On intra oral examination, yellowish discoloration of the teeth was evident with generalized altered morphology of crown and pitting on the surface. No chipping of the enamel was evident on probing. (fig-5) Clinically missing 26 & 27 was evident, deep class I caries was evident w.r.t 36, 46 & 47. 26 was restored and deep class II caries was evident wrt 17. Angles class I molar relation bilaterally and anterior edge to edge bite was evident. Based on the history and clinical findings a provisional diagnosis of Kennedy's class 3 edentulous space wrt 26 & 27, Angle's class I malocclusion and Amelogenesis imperfecta hypoplastic type was given.

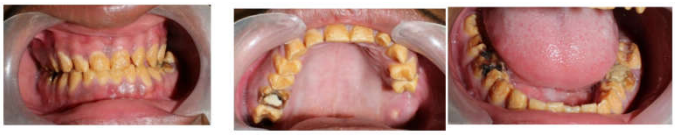

Figure 5 Yellowish discoloration and pitting of the enamel surface.

Intraoral periapical radiographs wrt upper and lower incisors and bitewing radiographs wrt upper and lower posteriors (fig-6) and digital OPG (fig-7) were advised. Radiographs revealed, altered morphology of crown giving it a square shape due to loss of contact between the teeth with decreased density of enamel.

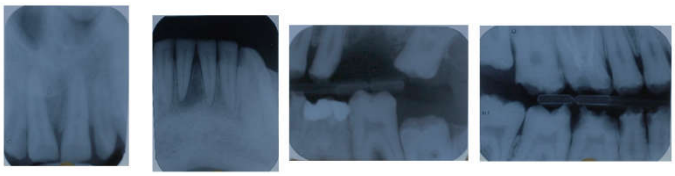

Figure 6 thin and reduced density of enamel evident with irregular surface of crown.

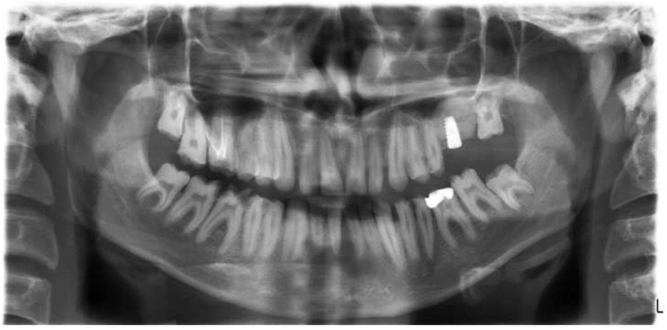

Figure 7 Digital OPG revealing reduced density of enamel and alteration in the morphology of the teeth.

Complete loss of enamel was evident wrt 31, 32, 41, 42. Loss of lamina dura was evident at apical 3rd of root wrt 41, pdl space widening all along the root was evident wrt 41 along with diffuse periapical radiolucency extending mesiolateraaly till middle 3rd of root wrt 41. Proximal radiolucencies involving enamel and dentin were evident wrt 25, 44, 45, 46, 47. Coronal radiolucency involving enamel and dentin & radiopacity involving pulp canals wrt 16. Digital OPG revealed, generalized thin cap of enamel over the crown, reduced density of the enamel, irregular surfaces of the crown and square shaped crowns. Based on clinical and radiographic examination the diagnosis of amelogenesis imperfect a hypoplastic type was confirmed.

DISCUSSION

Amelogenesis imperfecta (AI) is a developmental disorder which affects the amelogenesis leading to defects in enamel formation, maturation or mineralization. This group of disorder can be autosomal dominant, autosomal recessive and X linked. There are various classification for AI, the most commonly used classification of AI is given by Witkop & Rao (1971), based on its phenotype and inheritance as hypoplastic, hypocalcified and hypomaturation type. [3] The clinical appearance of the disorder is based on its type. In case of hypoplastic type of AI, the enamel will be well mineralized but

the amount of enamel will be reduced. [4] There will be pitting and rough surface appearance and also loss of occlusal contact due to tapering of the proximal surface of the teeth occlusaly. Radiographically there will be thin periphery of enamel with loss or absent cusps, similar to our case. In hypomaturation type, the teeth will show mottled appearance with whitish yellow discoloration. Enamel will be soft on probing. Radiographically, the thickness of enamel will be normal but its density will be same as dentin. While in hypocalcified type of AI, the enamel will be discoloured, soft and easily chip off on probing. Radigraphically thickness will be normal but density will be less than that of dentin. The differential diagnosis for this disorder is dental fluorosis, enamel hypoplasia, dentinogenesis imperfecta. Dental fluorosis may present with areas of horizontal white bands in the enamel and may spare premolars. History will reveal excessive fluoride intake in these patients. Enamel hypoplasia also appears similar to dental fluorosis, it may be caused due to prolonged GI disturbances, Coeliac disease, anti-leukemia therapy etc which will be revealed in history. [6] Dentinogenesis imperfecta is an autosomal dominant hereditary disorder in which shape and size of tooth will be normal with normal density of enamel and there will be obliteration of pulp chamber and root canals. [7] The diagnosis involves exclusion of extrinsic environmental factors or other intrinsic factors, inheritance pattern & phenotype.

The problems associated with this disorder affects both function as well as aesthetics of the patient. Patients are usually more concerned about the aesthetics rather than the function. Various problems associated with AI are hypersensitivity, loss of vertical height in occlusion, delayed eruption of teeth, malocclusion, taurodontism, dens in dente, dental caries etc. [5] But in our case, patient did not have any hypersensitivity but had caries in both the siblings. Attrition of the teeth and malocclusion was evident in our patient.Certain clinical reports have done the restorations of individuals affected by amelogenesis imperfecta with adhesive restorative techniques, overdentures, fixed partial dentures, full porcelain crowns, porcelain fused-to metal crowns and inlay/onlay restorations constitute the contemporary treatment modalities. [8,9,10]

CONCLUSION

AI is a genetic disorder which causes functional as well as aesthetic problems to the patient. Though this disorder is least common among the developmental anomalies affecting the teeth yet its diagnosis is of great importance. The history and clinical features of the disorder directs us to the diagnosis. Mostly the diagnosis of this disease is based on the exclusion of other disorders which cause defects in the tooth structure. These patients are more prone to dental problems and so need extensive treatments. Maintenance of oral health, correction of malocclusion and prosthetic rehabilitation are the commonly carried out treatment for such patients.

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