A Case Report

Amelogenesis Imperfecta : A Case Report

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INTRODUCTION

Amelogenesis imperfecta encompasses a complicated group of conditions that demonstrate developmental alterations in the structure of the enamel in the absence of a systemic disorder.

Amelogenesis imperfecta (AI) is a heterogeneous group of genetic disorders characterized by defects in tooth enamel formation in the absence of any generalized or systemic diseases. AI is currently classified into 14 distinct subtypes based on the clinical phenotype and mode of inheritance. Major enamel matrix proteins (amelogenin, enamelin, and ameloblastin) are suggested to contribute to the enamel formation of teeth. During the secretory stage of enamel formation, these proteins are secreted by ameloblasts and play key roles in the growth of enamel crystal. Several reports have shown that mutations in the amelogenin gene located at Xp22.1-p22.3 cause X-linked AI . The most common type of AI is the autosomal-dominant form .

ETIOLOGY

It is an inherited disorder related to the alteration of the gene involved in the formation & maturation of the enamel.

Different inherited patterns are:-

Autosomal dominant., Autosomal recessive, X-linked. Most common type is autosomal dominant type. The main types are: hypoplastic (type I); hypomaturation (type II); hypocalcified (type III); and hypomaturation/ hypoplasia/ taurodontism (type IV).

Clinical Features: Amelogenesis imperfecta affects the enamel of all of the teeth of the affected individuals within a kindred, in a more or less equal manner, without reference to chronology, occasionally in association with other generalised conditions. The enamel may be hypoplastic, hypomineralised or both, and teeth affected may be discoloured, sensitive or prone to disintegration either post eruption (post-eruptive breakdown) or pre-eruption (idiopathic resorption), usually they are smaller than normal teeth.

Radiographic Features: Extra oral radiographs may reveal the presence of unerupted and sometimes spontaneously resorbing teeth, absence of cement enamel junction. Intra-oral radiographs will reveal the relative contrast between enamel and dentine in cases where mineralisation may have been affected. Features include a square crown, low or absent cusps, multiple open contacts between the teeth, a relatively thin radioopaque layer of enamel.

Histopathological Features: predentin areas with irregular canaliculi between normal dentin and internal resorption areas in the pulp tissue. amelogenesis imperfecta, enamel tissue is mostly affected besides minor defects in dentinal and pulpal tissue.

Differential Diagnosis

Dental fluorosis, Extrinsic disorders of tooth formation, chronological disorders of tooth formation and localised disorders of tooth formation should be considered in the differential diagnosis.

CASE REPORT

A 23 year old male patient reported with chief complaint of missing teeth and discolored teeth on both upper and lower jaw region. Teeth were not normal since childhood. Initially teeth were loosened and gradually lost most of his teeth for last 10-12 years and some teeth were removed by a local practitioner (fig 1,2,3). Family history was revealed that his sister also had similar type of discolored teeth since her childhood with multiple missing teeth.

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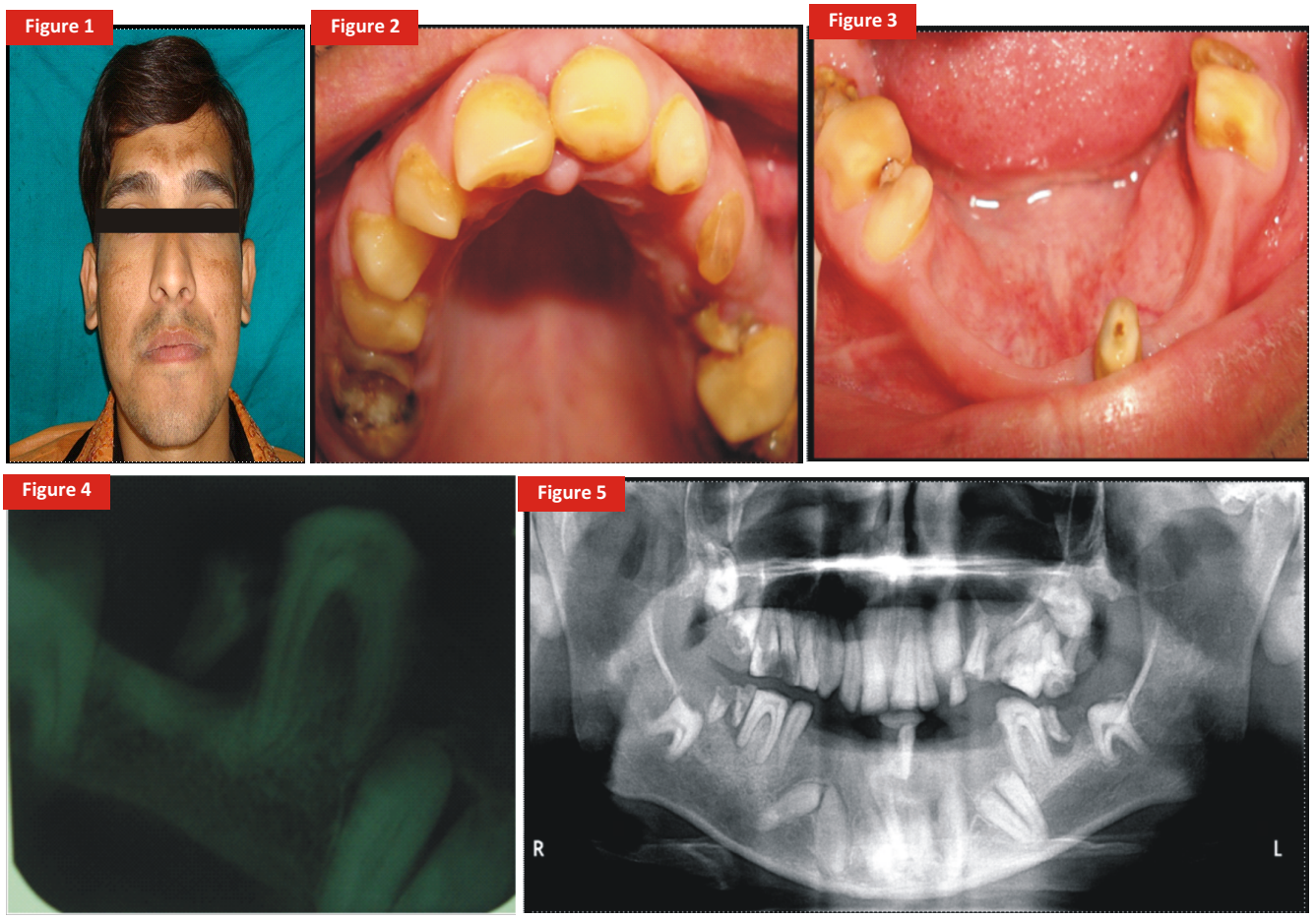

On Examination- On general physical examination right submandibular lymph node was palpable, single in number, mobile, soft in consistency & non tender.-Intra oral examination revealed that discoloration was present all over the teeth which were yellowish white in color with conical shaped crown. There were multiple missing teeth in relation to 32,33,34,35,41,42,43, and 44, and root stumps were present i.r.t-15, 25, 37, and 47. Dental caries was seen in relation to 16, 46 and 31 was mobile.

Provisional Diagnosis

Considering the history and clinical examination, the provisional diagnosis of Amelogenesis Imperfecta was given.

Investigations

Full mouth Intra-oral peri-apical radiograph and OPG was advised to the patient. Radiographs revealed that enamel density was not appreciable and open contact between the teeth was seen. Cemento-enamel junction was completely absent. Radiolucency on the coronal portion confirmed generalized attrition of the tooth. Radiopacity in the pulp chambers of the tooth was suggestive of generalized pulp stone.(fig.4 &5) Multiple impacted teeth were present without enamel capping. Extraction of teeth with poor prognosis i.e. 31, 25 and 37 was done and sent for the ground sectioning and histological examination,

Final diagnosis- Hypoplastic Amelogenesis Imperfecta

Treatment

Primary treatment includes cosmetic improvement of the patient with the help of placement of crown or facial veneer on the teeth. Desensitizing agents are advised to treat the hypersensitivity in the patients due to dentine exposure of the teeth. Over dentures are also advised in case of patient who doesn't have sufficient crown length for the restoration.

DISCUSSION

Amelogenesis imperfecta is a disorder of tooth development . This condition causes to be usually small, pitted, discoloured, grooved and prone to rapid wear and breakage . Other dental abnormalities are also possible.

CONCLUSION

Amelogenesis Imperfecta is an uncommon disorder; however, its clinical and radiological manifestations are characteristic. Management of patient with amelogenesis Imperfecta is important because it provides good aesthetics and maintains healthy supporting tissues. It helps the patient to develop a good psychologic self image. New alternatives for rehabilitation for such patient must be carefully considered, taking into account the presence of exposed dentin.

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