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CASE STUDY

AMELOGENESIS IMPERFECTA WITH TAURODONTISM & CONGENITALLY MISSING TEETH IN SIBLINGS – A RARE COMBO

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ABSTRACT

Amelogenesis imperfecta is a group of hereditary developmental defect in enamel formation either in quality and/or quantity which affects both deciduous and permanent teeth. It may be hypoplastic, hypomaturation, hypocalcified type. Hypomaturation-hypoplastic AI with taurodontism is a rare variant. AI has been associated with different dental anomalies including discoloured teeth, taurodontism, congenitally missing teeth, microdontia, pulpal calcification, hypercementosis, crown and root resorption. This is a case report of Hypomaturation AI with taurodontism in siblings born to parents with consanguineous marriage and congenitally missing tooth in one of the sibling.

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INTRODUCTION

Appearance of dentition is of great concern and in particular colour has cosmetic importance. Tooth discolouration can be either intrinsic or extrinsic. Change in structural composition or thickness of dental hard tissues causes intrinsic discolouration and Amelogenesis imperfecta (AI) is one such condition. AI, a group of hereditary diseases affecting the tooth enamel in either quality or quantity, is associated with crown malformation and abnormal enamel density (Chamarthi *et al*., 2012) which may be inherited as an autosomal dominant trait or autosomal recessive disorder or as an X-linked condition. The prevalence varies from 1:700 to 1:14,000 according to the populations studied (Crawford *et al*., 2007) affecting both primary and permanent teeth causing unusually small, pitted, grooved, discoloured, and prone to rapid wear and breakage. Witkop classified AI into 4 major categories based primarily on phenotype: hypoplastic [HP], hypomaturation [HM], hypocalcified [HC], HM□HP with taurodontism; subdivided into 15 subtypes based on clinical phenotype and mode of inheritance. (Witkop, 1988) AI regardless of subtype have similar oral complications: teeth sensitivity, decreased vertical dimension, poor esthetics and reduced self-confidence when anterior teeth are affected. (Fonda *et al*., 2006; Visram Salima and Mckaig Sarah, 2006) It has been associated with different dental anomalies including discoloured teeth, taurodontism, congenitally missing teeth, microdontia, pulpal calcification, hypercementosis, crown and root resorption.

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Case report 1

A 8yr female reported to dept. of pedodontics with parents complaining of discoloured tooth since the time tooth erupted expressing the concern that child feels shy to smile socially and sensitivity in teeth. Family history revealed child borne from consanguineous marriage, dentition was normal in them but younger sibling showed similar condition and none of the other family members had such condition. Medical history was non-contributory and general examination suggested good state of health. Intra oral hard tissue examination revealed mixed dentition status. Generalised yellowish discolouration and attrition of teeth exposing the dentin on certain teeth. Distal marginal ridge of 54, 64 was broken (Figure 1); 51,61,71 showed preshedding mobility. A Provisional diagnosis of Amelogenesis imperfecta was given. Panoramic radiograph showed permanent 1st molars with enlarged body, pulp chamber as well as apical displacement of pulpal floor representing Taurodontism. (Figure 2) Histopathological examination of

ground section of extracted tooth showed indistinct enamel rods with less maturation, variable thickness of enamel and lamellae running across the enamel rods. Exposed dentin shows dead tracts with sclerotic dentin at the pulpal border and smooth DEJ. Newly formed dentin also shows defects in mineralization. (Figure 3) Based on all the findings final diagnosis of Hypomaturation type of AI with taurodontism (Type IV) was given.

Case report 2

A 6yr female, younger sibling of case 1 reported with similar condition. Intra oral hard tissue examination revealed mixed dentition status with congenitally missing deciduous teeth 52. Generalised yellowish discolouration and attrition of teeth exposing the dentine on certain teeth was seen. Mesial marginal ridge of 55, 65 was broken; 71 showed preshedding mobility (Figure 4). Condition was provisionally diagnosed as AI. Panoramic radiograph revealed congenitally missing permanent lateral incisors (12,22) and permanent 1st molars showed taurodontism (Figure 5). Histopathogical examination of ground section of extracted tooth showed irregular surface with indistinct enamel rods with variable thickness of enamel which is lost at the incisal edge and few enamel lamellae running across the enamel rods. Exposed dentin shows dead tracts with sclerotic dentin at the pulpal border and smooth DEJ.

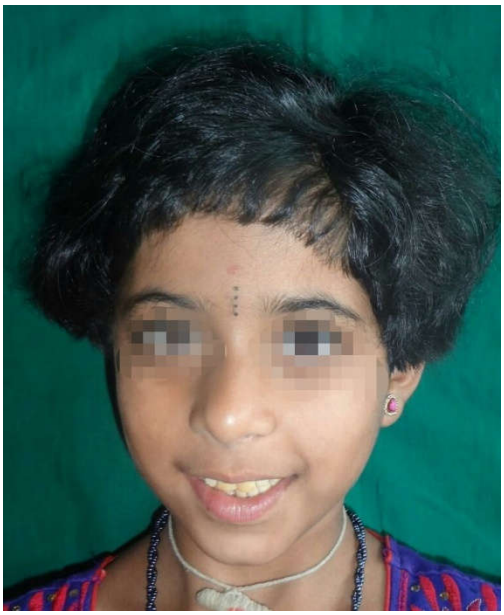

Figure 1(a). Extra oral view

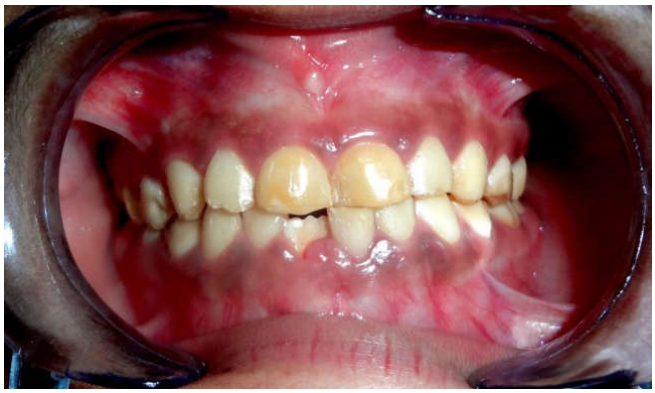

Figure 1(b). Labial view

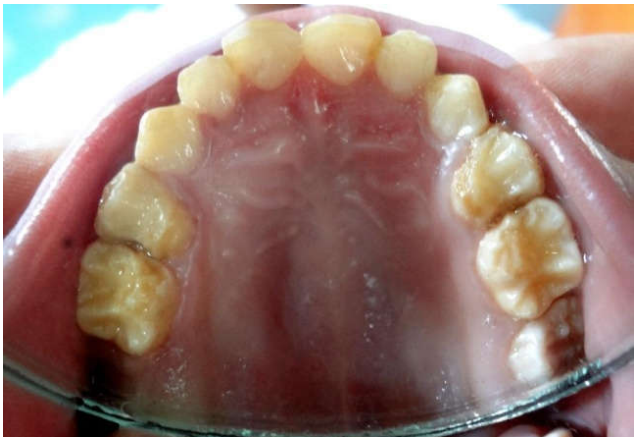

Figure 1(c) maxillary occlusal view

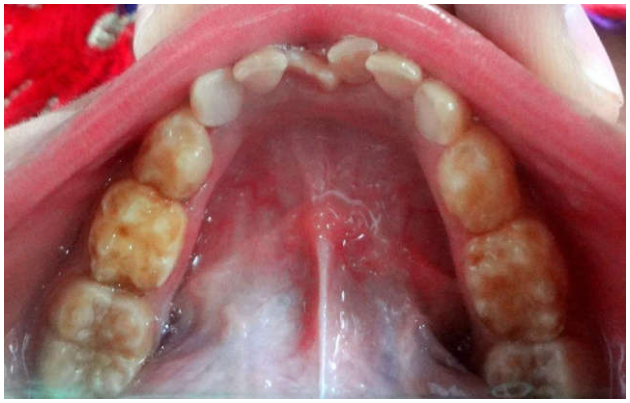

Figure 1(d) mandibular view

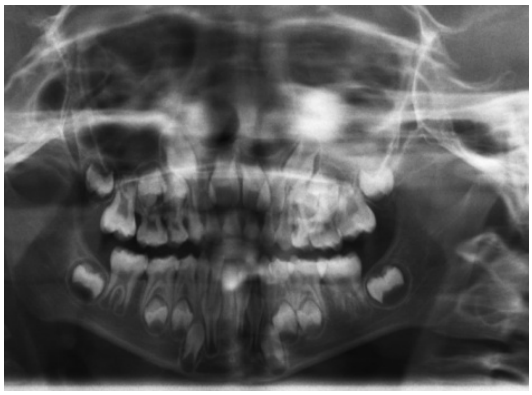

Figure 2. Pantomograph

Figure 3. Histopathological examination

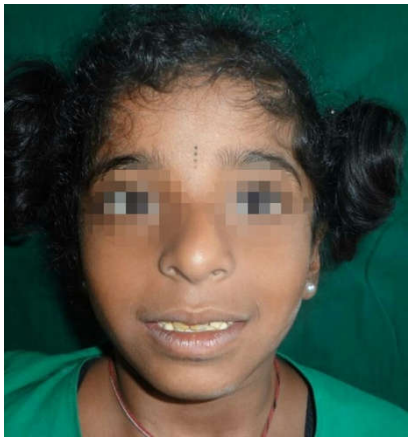

Figure 4 (a) extra oral view

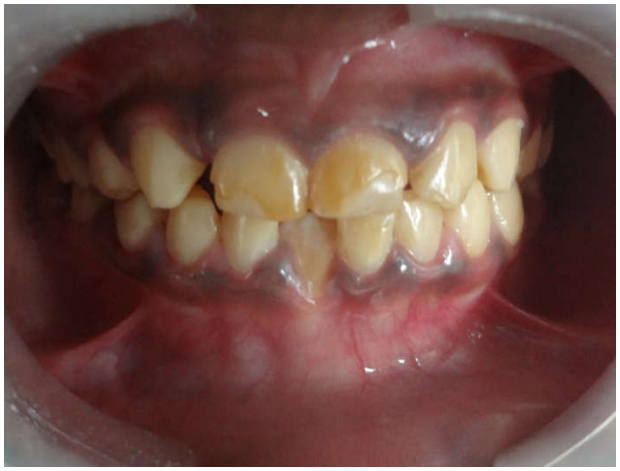

Figure 4 (b) labial view

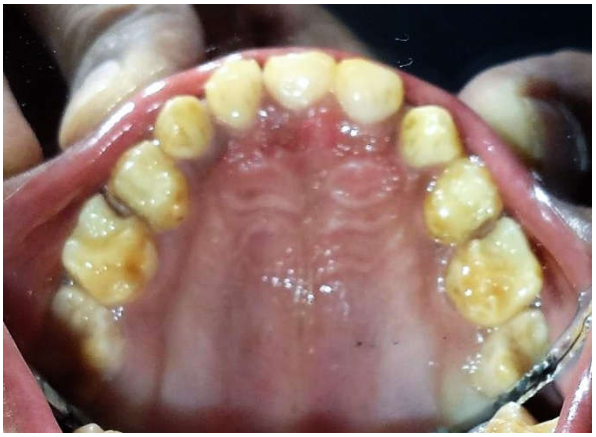

Figure 4 (c) maxillary occlusal view

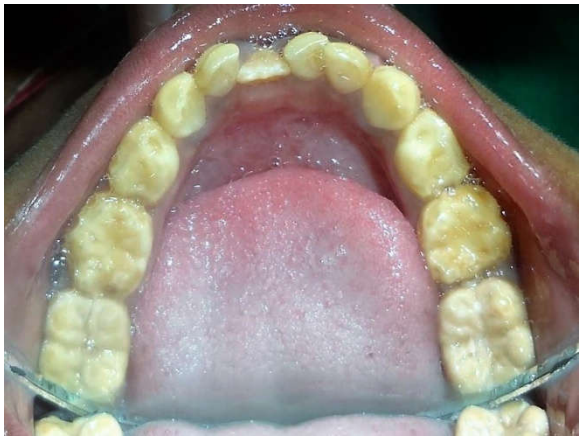

Figure 4 (d) Mandibular occlusal view

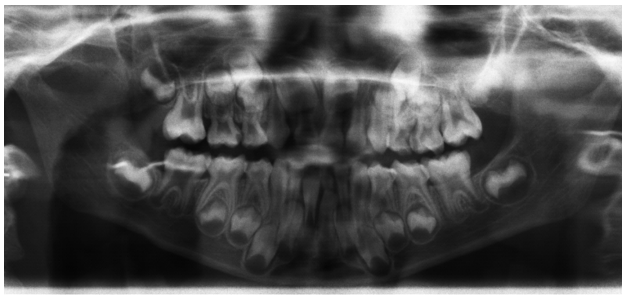

Figure 5. Orthopantomograph

Figure 6. Histopathological examination.jpg

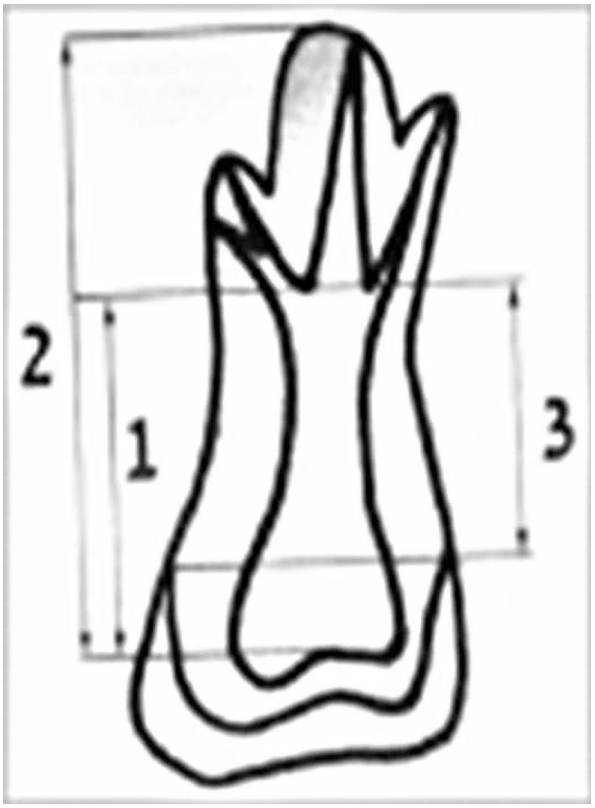

Figure 7. Caluculation of taurodontism index

Newly formed dentin also shows defects in mineralization. (Figure 6) Based on all the findings final diagnosis of Hypomaturation type of AI with taurodontism (Type IV) and congenitally missing tooth 12,22 was given. Fluorosis was

ruled out by sending the siblings household drinking water for fluoride content test which showed 0.72mg/l inferring optimal level of fluoride (Document 1).

DISCUSSION

HM-HP AI with taurodontism is a autosomal variant often confused with trichodentoosseous syndrome (Witkop, 1988) as the dental findings are similar. According to Crawford and Aldred if trichodentoosseous syndrome is inherited with complete expression, the affected individual in successive generations will show the typical kinky hair, bone, teeth changes (Crawford and Aldred, 1990) and craniofacial defects like frontal bossing, maxillary retrusion, square jaw, mandibular prognathism and dolichocephaly. (OlaB.Al-Batayneh, 2012) The kinky hair seen in children is lost in adults and the bone changes are not seen in all cases of trichodentoosseous syndrome. (Crawford and Aldred, 1990) In the present cases no such abnormalities were detected. Hence trichodentoosseous syndrome was ruled out. Siblings were born to parents of consanguineous marriage. Enamel renal syndrome also known as Amelogenesis imperfecta nephrocalcinosis syndrome runs in consanguineous family. The common features of this syndrome are the presence of thin or no enamel, delayed tooth eruption, intrapulpal calcifications, bilateral nephrocalcinosis and normal plasma calcium. (Kala *et al*., 2012) But in the present cases no such abnormalities were seen it can be ruled out. Shifman and Channel's index was used to calculate the degree of taurondontism. Taurodontism index: vertical height of the pulp chamber (1); distance between the lowest point of the roof of pulp chamber to the apex of the longest root (2); distance between the baseline connecting the two cement-enamel junction and the highest point in the floor of the pulp chamber (3). Establishment of a condition of taurodontism is made when 1/2 multiplied by 100 is above 20, and 3 exceeds 2.5 mm: (1/2) x 100 > 20 and 3 > 2.5 mm. (Shifman and Chanannel, 1978) (Figure 7) In the present cases apex is not closed only point 3 is considered. Case 1: tooth 16 – 5.5 mm, 26 – 6mm, 36 – 4.5mm, 46–4.5mm; Case 2:tooth 16-4.5mm, 26-5.5mm, 36-4.5mm, 46-5mm; indicating taurodontism. Management for AI is quiet challenging which includes three aspects of care: prevention, restoration and esthetics. Preventive aspects include oral hygiene instructions, dietary advice and fluoride supplements/topical fluoride application. Oral hygiene maintenance is important as rough enamel surface causes plaque retention and sensitivity is experienced while brushing. (Visram Salima and Mckaig Sarah, 2006) Restorative aspects include glass ionomer restorations and stainless steel crowns in primary dentition; adhesive casting or stainless steel crowns on first permanent molars and composite or glass ionomer cements in mixed dentition; adhesive casting on premolars in permanent dentition. Esthetic aspect include minimal intervention glass ionomer cement restorations in primary dentition, direct and indirect composite resin veneers in the mixed dentition and porcelain veneers, full crowns, over dentures and complete dentures in the permanent dentition. (Markovic *et al*., 2010) Psychologically AI will have huge impact even in children. So complete rehabilitation should be done starting early in age and continued till adult.

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