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Case Report

A Rare Association between Amelogenesis Imperfecta -Smooth Hypoplastic type and Anterior Open Bite: An Interesting Case Report

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ABSTRACT:

Amelogenesis imperfecta (AI) is a heterogeneous group of hereditary disorders primarily affecting dental enamel. This paper describes the clinico-radiological features of a 12-year-old female patient who was born of consanguineous parents and presented with smooth yellow discolored and misshapen teeth, small crowns, delayed eruption of permanent teeth, retained deciduous teeth, open contact areas between teeth, mild gingival involvement and anterior open bite. Radiograph revealed intrapulpal calcifications, enlarged dental sac surrounding unerupted molars and uniform radiodensity of enamel and dentin. Similar condition was observed in her 9-year-old brother. None of the other family members showed any evidence of AI. The diagnosis of smooth hypoplastic Amelogenesis imperfecta was confirmed. This case is first of its kind presenting the association of smooth hypoplastic Amelogenesis imperfecta type with anterior open bite and intrapulpal calcifications.

Key words: Amelogenesis imperfecta, Smooth hypoplastic type, Anterior open bite.

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INTRODUCTION:

Amelogenesis imperfecta (AI) is a diverse group of hereditary conditions that affects the quality and quantity of dental enamel.¹ In general the defects in AI were classified as three pathological types hypoplastic, hypocalcified and hypomaturational types, depending on the stage of enamel formation. The hypoplastic AI types are characterized by the presence of thin enamel or pits or grooves on the enamel surface. Hypocalcified AI types are characterized by soft and discolored enamel. Hypomaturational AI types are associated with maturation defects, which are clinically characterized by the presence of opaque and chalky enamel.² In the

studies by Emma LD et al, two siblings of consanguineous marriage of parents showed thin and smooth enamel, intrapulpal calcifications, delayed eruption of teeth suggestive of hypoplastic AI with nephrocalcinosis.³

Nephrocalcinosis is characterized by the deposition of calcium in renal tissues and is usually asymptomatic in the early stages of childhood. The common clinical oral findings that can be the signs for future renal nephrocalcinosis includes presence of thin or absent enamel, delayed tooth eruption and pulp calcifications.⁴ The incidence of AOB in patients with AI varies from 24% -60%. It was reported that the association between

AOB and AI was genetic. AOB is seen in both hypoplastic and hypocalcified types of AI.⁵ Consanguineous marriages are common in Middle Eastern countries. In Saudi Arabia, a prevalence rate of 51.3% and 52.0% was reported in the cities of Riyadh and Dammam.⁶ The relationship between consanguineous marriages and genetic disorders such as Amelogenesis imperfecta, were reported from many parts of the world. However, in countries such as Saudi Arabia, very limited studies were reported. Therefore, the purposes of this present study was to characterize the significance of consanguineous family presenting smooth hypoplastic amelogenesis imperfect with anterior open bite, a rare association and signify the presence of intrapulpal calcifications which indicates the future need for renal consultation.

CASE REPORT:

A 12 year-old female patient visited Riyadh Elm University Dental Hospital, with a chief complaint of unpleasant appearance of her teeth and anterior open bite. Similar condition was observed in her 9-year-old brother but was uncooperative; therefore this was not included in the present study. The family history revealed consanguineous marriage of parents and they were first cousins. Intraoral Examination revealed yellow discolored misshapen teeth with small short crown size, delayed eruption of permanent teeth, retained deciduous teeth, open contact areas and anterior open

bite (Figure 1). The Maxillary arch revealed yellow discolored 11, 21 and partly erupted 12, 22, unerupted 13, 23, retained 53,63, and delayed eruption of 14, 24 were noted (Figure 2). In the Mandibular arch yellowish –white discolored 31,41, small sized, misshaped 32,42 with sharp pointed cusps and delayed eruption of 33,34, 35, 36, 43, 44 and 45 was observed (Figure 3). Past dental history revealed extraction of 74, 75, and 84 due to caries. All the teeth showed insufficient enamel thickness with generalized smooth surfaces and discoloration. Extra oral examination was not significant. Patient's skin, Hair and nails were normal. No history of thumb sucking habit, abnormal swallowing habit and tongue thrust habit and lacks hypersensitivity of teeth. Clinically the patient was asymptomatic with no complaint of urinary tract infection, renal stones, hematuria, polyuria or enuresis. The panoramic radiograph showed thin enamel layer that could not be distinguished from the underlying dentin. It showed retained deciduous teeth and delayed eruption of several permanent teeth. No density difference between enamel and dentin was observed. Intrapulpal calcifications were seen in unerupted permanent teeth 37 and 47. Enlarged pericoronal follicular space was seen associated with unerupted teeth (Figure 4). Ultrasound of kidneys revealed no calcifications (Figure 5). The clinical and radiographic features led to the diagnosis of Hypoplastic amelogenesis imperfecta.

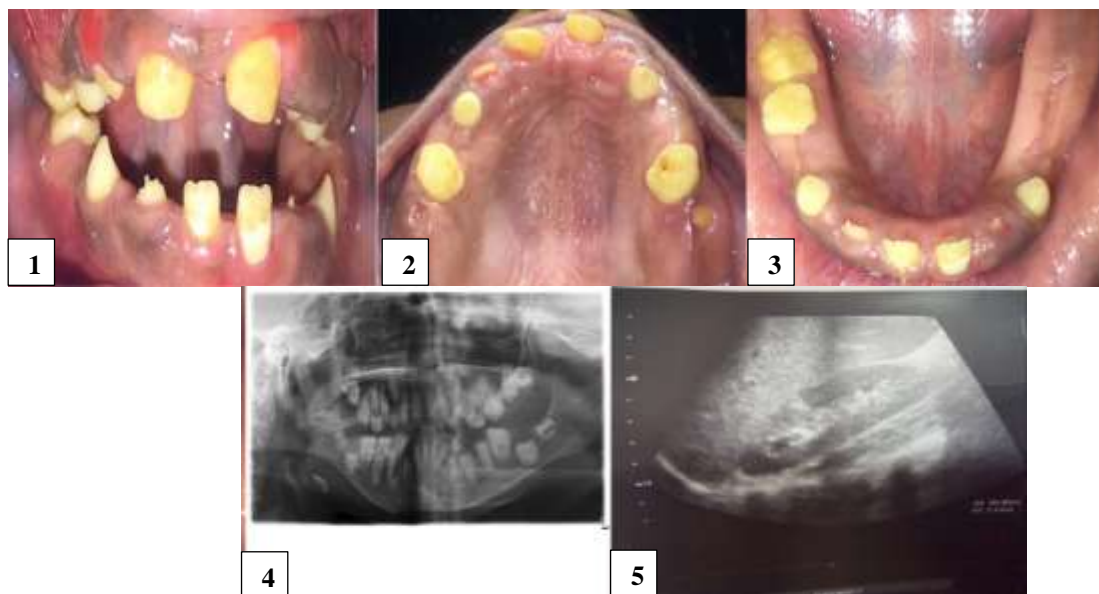


Figure 1: Anterior open bite and the teeth shows yellow discoloration and short crowns;
Figure 2: Maxillary arch showing partly erupted 12, 22 and retained deciduous teeth;
Figure 3: Mandibular arch showing yellow –white discolored 31 and 41;
Figure 4: Panoramic radiograph showing intrapulpal calcifications in unerupted molars;
Figure 5: Ultrasound revealed no calcifications in kidneys

DISCUSSION:

Among the published data, hypoplastic AI with anterior open bite cases were limited. Smooth thin hypoplastic autosomal dominant type was studied in seven patients from three different families that showed features similar to the present case but contradicts the statement that the pulp stones are formed due to local irritation, thin enamel and attrition. In the present case, intrapulpal calcifications were observed in unerupted teeth with no threat of irritation.² The first family with both siblings having AI with nephrocalcinosis was described in 1972 by Mac Gibbon and in 1985, similar studies by Lubinsky et al described autosomal recessive hypoplastic AI with dagger shaped intrapulpal calcifications and nephrocalcinosis in a brother and sister pair, but none of these studies mentioned consanguineous marriage of parents, which is in contrast to our present case which revealed consanguineous marriage of parents but without nephrocalcinosis.³ In previous studies, in Saudi Arabia, the hypersensitivity of teeth and anterior open bite was observed in hypocalcified AI and hypomaturation AI types which is in contrast to our present study that showed hypoplastic type without hypersensitivity of teeth.⁷

Hypoplastic AI with anterior open bite was observed in the cases studied by Ulas Oz et al.⁸ Clinical studies indicate that AOB is more commonly observed in patients with AI than in the general population. AOB most frequently occurred in the hypocalcification type of AI, less in hypoplastic and totally absent in hypomaturation type.⁹ In the studies by Alenka P et al.¹⁰ it was observed that the patients with rough hypoplastic amelogenesis imperfecta showed anterior open bite than in smooth types, which contradicts our present case in which anterior open bite was observed in smooth hypoplastic type of AI. A multidisciplinary treatment approach was suggested to the parent of the patient.

CONCLUSION:

AI hypoplastic smooth or rough types showing features of intrapulpal calcifications, delayed eruption and

retained deciduous teeth should be considered for a renal ultrasound scan. Further studies are required to confirm the genetic influence in AI associated AOB cases.

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