Tricho Dento Osseous Syndrome

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Abstract: Tricho-dento-osseous dysplasia is a rare autosomal dominant disorder which involves increased bone density, enamel hypoplasia, enlarged pulp chambers, and molar taurodontism. We report a case of a 12 year old female presenting with tricho dento osseous syndrome. [Gandhi B et al NJIRM 2012; 3(3) : 186-189]

Key words: : Tricho dento osseous syndrome; Amelogenesis Imperfecta; Enamel Hypoplasia

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Introduction: The term "trichodentoosseous" (TDO) syndrome first was coined by Lichtenstein et al in 1972 to describe a large kindred with an autosomal dominant syndrome of curly hair, enamel hypoplasia and taurodontism, dysplastic nails, and generalized osseous dysplasia involving the long bones and calvarium[1,2]. TDO syndrome is of interest to dentists not only because of the severe enamel defects, but also because this condition is often confused with that of primary amelogenesis Imperfecta[1]. Thus the purpose of this case report is to help shed light on the general and oral features that may be considered consistent in this syndrome.

Case Report: A 12 year old female reported to Ratnasagar dental clinic, Ahmedabad for the replacement of missing anterior teeth. Extraoral examination revealed a dolicocephalic face with a broad forehead (Figure 1a).

The nails were found to be brittle. A history of kinky hair during infancy was present which straightened out with an increase in the age. Intraoral examination revealed hyperplastic gingiva in the maxillary anterior region. Permanent maxillary central incisors were missing. Over retained deciduous maxillary incisors and molars were present. The erupting right maxillary permanent lateral incisor was labially inclined. All the teeth were yellowish in color with very thin enamel. The permanent mandibular incisors showed mottling with attrition of the incisal edges. Mottling was also present in the permanent maxillary molars and mandibular premolars and molars (Figure 2).

Figure 1 a,b - Extraoral picture of the 12 year old girl showing dolicocephalic face and broad forehead. Extraoral picture of the patient's 35 year old mother showing similar facial profile

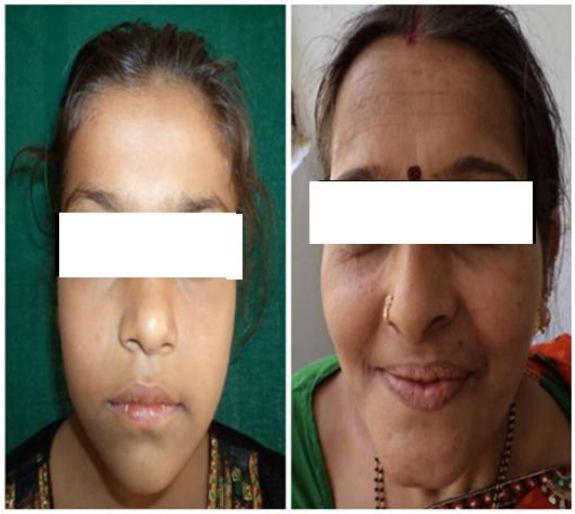
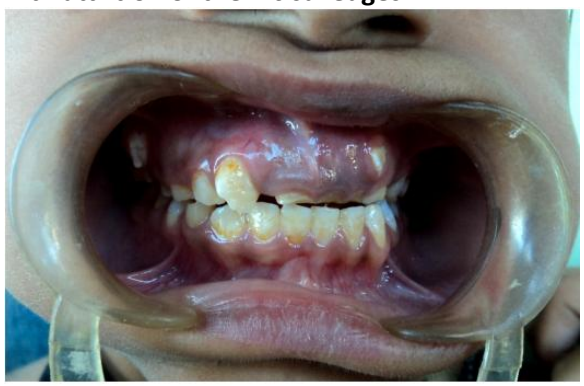


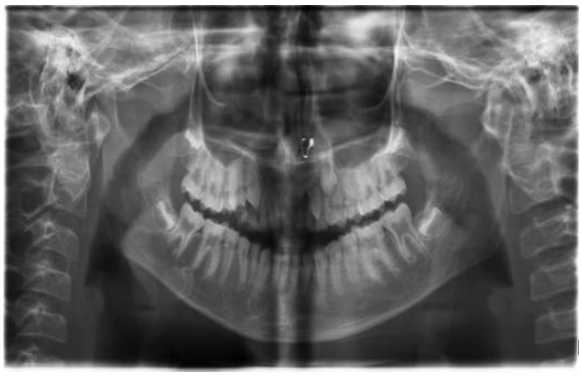
Figure 2 – Intraoral picture of the patient showing yellowish discoloration of teeth. Mottling of permanent mandibular incisors with attrition of the incisal edges.



An orthopantomogram revealed an obtuse mandibular angle on both sides. Missing permanent tooth buds of maxillary permanent central incisors were also seen. The mandibular

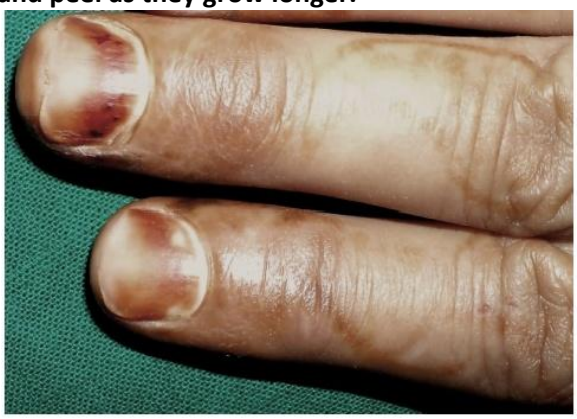
molars showed thin enamel. Roots of mandibular second molars were short with wide pulp chambers suggestive of taurodontism (Figure 3).

Figure 3 – Orthopantomogram showing an obtuse mandibular angle on both sides. Missing permanent tooth buds of maxillary permanent central incisors can be seen. Thin enamel in mandibular molars is present. Roots of mandibular second molars are short



Similar extraoral features were also exhibited by her 35 year old mother, who presented with a dolicocephalic face (Figure 1 b). She reported the presence of kinky hair which straightened out later. The nails were found to be brittle, with a tendency to break and peel as they grew longer (Figure 4).

Figure 4 – Brittle nails with a tendency to break and peel as they grow longer.



She also gave a history of wearing maxillary and mandibular overdentures, following multiple extractions two years back (Figure 5).

Figure 5 – Intraoral picture of the patient's mother showing multiple missing teeth

Based on the family history and the above described features, a diagnosis of tricho dento osseous syndrome was made. The patient was referred to a pedodontist for further treatment.

Discussion: Tricho-dento-osseous syndrome (TDO), is inherited as a highly penetrant autosomal dominant condition that demonstrates variable expression of curly hair in infancy, enamel hypoplasia, taurodontism (enlarged pulp chambers) and both thickening and increased density of the cortical bones of the skull[3,4].

Hart et al have reported the genetic linkage of TDO syndrome to chromosome 17q213. The dental defects in TDO are usually the most dramatic and consistent of all the signs. Previous authors reported the teeth to be yellow-brown in color displaying hypocalcification/hypomaturation enamel defects together with enamel hypoplasia. The enamel is estimated to be approximately 1/4 to 1/8 normal thickness[1]. These defects were associated with severe attrition, which may be why dental abscesses are commonly reported.

Histological examination of affected teeth in TDO syndrome have shown the enamel to be undercalcified and thin[1]. Furthermore, the pulp chamber may be enlarged, with the pulp horns extending close to the dentinoenamel junction, a condition similar to that seen in vitamin D-resistant rickets[1]. Enamel hypoplasia with hypomaturation and attrition of mandibular

incisal edges was present in our case. Widened pulp chambers of molars in our case was also consistent with the literature. Other reported features of the TDO phenotype include fingernail involvement including either splitting of the superficial layers or thick cornification, narrowing of the ear canal and altered craniofacial morphology, principally macrocephaly and dolichocephaly[3]. While ear changes and cranial osteosclerosis were absent in our case, dolicocephaly and nail changes were present. Kinky ''uncombable'' hair is one of the hallmarks of TDO[5,6]. In 54% cases, the kinky hair phenotype disappears after infancy, as was seen in our case[7].

The major diagnostic criteria for TDO syndrome includes all three of the following: (i) a positive family history of TDO; (ii) presence of generalized hypoplastic dental enamel (assessed by clinical and radiographic examination); and (iii) presence of taurodontism in at least two posterior teeth (assessed by radiographic examination). In addition to meeting these major criteria, to be classified as affected the presence of at least one of the following minor features is necessary: (i) curly/kinky hair at birth ; (ii) thick and/or dense bone (as assessed by cranial radiographs); or (iii) abnormal nails (e.g.brittle and peeling, cornified)[3,8]. All three major criteria and two minor criteria in relation to hair and nails was present in our case. Differential diagnosis includes osteopetrosis, Amelogenesis Imperfecta and tricho-onycho-dental syndrome[1,3].

The main aim in managing affected patients centers on preventing clinical problems as early as possible and improving dental esthetics. The discoloration of the teeth may need full-coverage jacket porcelain crowns. However, this type of restoration may not be recommended in the young patient due to large pulps. Interim veneer restorations using bonded composite resins or porcelain may be considered. Sensitivity of teeth and excessive loss of tooth structure leading to pulp exposures and decrease of occlusal vertical dimension may be prevented by stainless steel crown coverage as soon as the molars have reached adequate crown height[10]. Restoration of carious and hypoplastic defects is achieved with adhesive materials if possible since the weak enamel margins tend to fracture away from nonadhesive dental amalgams[10].

Conclusion: To conclude, dentists must be aware of the oral and general features of TDO syndrome to establish a prompt diagnosis and initiate an early treatment.

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