Ectodermal dysplasia with true anodontia

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ABSTRACT
The hereditary condition known as ectodermal dysplasia is characterized by the absence or defect of two or more ectodermally derived structures. The most commonly observed forms of ectodermal dysplasia are the hidrotic and hypohidrotic types; discrimination is based on the absence or presence of sweat glands. A case of 8-year-old male child with hypohidrotic ectodermal dysplasia with complete anodontia of primary as well as secondary dentitions is presented. The child had a short stature, low intelligent quotient (I.Q.), and was underweight. The patient experienced episodes of high fever, was intolerant to heat, and did not sweat. He exhibited smooth and dry skin, sparse light-colored eyebrows. Dental clinicians can be the first to diagnose ectodermal dysplasia due to the absence of teeth.

Key words: Anodontia, dysplasia, ectodermal

CASE REPORT
Here is the case report of a 8-year-old male child with ectodermal dysplasia. The chief complaint of the patient was difficulty in mastication due to absence of maxillary and mandibular teeth. His personal history revealed that teeth never erupted. His mother told that his birth was normal and family history was negative. The patient experienced episodes of high fever, was intolerant to heat, and did not sweat. He was unable to look directly towards a light source. Physically, the child had a short stature and was underweight. The patient exhibited smooth and dry skin, sparse light-colored eyebrows [Figure 1]. Nails were thin and brittle [Figures 2 and 3]. Skin was dry and rough [Figures 1-3]. Other characteristics of ectodermal dysplasia, such as frontal bossing, saddle nose, reduced vertical dimension of face due to total anodontia were also noticed.

Intraoral examination revealed absence of teeth with thin alveolar crests [Figures 4 and 5]. Oral mucosa appeared dry; otherwise, no mucosal defects were seen. The tongue and palate appeared normal.

Occlusal and panoramic radiographs revealed no primary and permanent teeth [Figures 6-8]. In order to improve mastication...
and aesthetics, both upper and lower complete dentures were fabricated [Figure 9].

**DISCUSSION**

Hypohidrotic ectodermal dysplasia is characterized by hypohidrosis, hypotrichosis, and hypodontia.[4] Ectodermal dysplasia is one of the most important anomalies of interest to dental clinicians because of the absent or misshapen teeth.[1] Hypodontia is known as one of the major factors of ectodermal dysplasia and is almost always present. In severe cases, no

**Discussion**

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The absence of primary teeth (true anodontia) is a rare phenomenon. In this case, the patient’s history and clinical and radiographic examination revealed the absence of primary teeth. Acikgoz et al., Vieira et al., Pirgon et al., and Pannu and Singh also reported true anodontia of primary teeth. Total anodontia denoted by complete developmental absence of teeth in both primary and secondary dentitions was reported Pirgon et al., and Pannu and Singh. Radiographs showed no signs of formation of tooth buds. It is claimed that primary teeth must be present for the development of their permanent successors. There are no permanent teeth in the oral cavity of the patient, similar finding reported by Vieira et al.,

The patient experienced episodes of high fever, was intolerant to heat, and did not sweat. The patient had sparse eyebrows. Child’s inability to perspire, more comfortable during cold weather, and absence of hair from the eyebrows with scanty eyelashes were also reported by Gupta et al., and Pannu and Singh (2002).

Short stature, underweight in relation to age and mental retardation were reported in accordance to case reported by Gupta.

Management: To improve the appearance, mastication, and speech, the child was provided with maxillary and mandibular complete dentures similar to treatment provided by Vierra et al., Pannu and Singh. The dental team should be aware of the clinical presentation of ectodermal dysplasia in order to provide the correct guidance for functional, social, and psychological needs of the patients.

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