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Oral manifestations of a pediatric patient with hyperimmunoglobulin e syndrome: A case report and review of literature

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Statement of the problem: Hyperimmunoglobulin E Syndrome (HIES) is a rare primary immunodeficiency with marked increase in serum immunoglobulin E (IgE) levels and eczematous dermatitis. Individuals with HIES share characteristic facial features, with many oral manifestations 1. The therapeutic strategies are usually directed towards prevention and management of recurrent infections, which are refractory to the conventional strategies. About 200 cases of HIES have been reported so far, the purpose of this report was to describe the dental manifestations in the dentition of a hyperimmunoglobulin E syndrome patient 2.

Methodology and Theoretical orientation: In the present case the report present the oral manifestations of a 10-year old paediatric patient diagnosed with HIES, referred to our dental clinic from the emergency department because of dental pain and oral infections.

Findings: Patients affected with HIES usually present with definitive oral manifestations including the failure of exfoliation of the primary tooth which in turn lead to impaction of the succedaneous tooth, or in case if the succedaneous tooth erupts lingual to the deciduous tooth it may result in the presence of double rows of teeth leading to

subsequent malocclusion 2. Majority of the patients show a midline fibrotic bridge on the palate and a deep midline cleft anterior to the circumvallate papillae 5. The lips and cheek may show mucosal lesions consisting of surface fissures and non-scrapable keratotic striations, patches or plaques resembling lichenoid lesions 3. Oral candidiasis is also a common manifestation This patient was presenting with the dental and oral mucosal abnormalities like retained primary teeth, tongue fissures, multiple ulcerative and keratotic lesions on the buccal mucosa, and most of the facial and morphological features which are associated with HIES syndrome.

Conclusion: The prognosis of HIES patients depends on the early diagnosis and prompt management of the complications; hence, thorough knowledge of a dental specialist especially a Paediatric dentist is important about identifying the early oral manifestations of the disease is mandatory 4

Biography

Tahani A Almutairi studied General Dentistry at the King Saud University in Riyadh then she studied and specialized in Pediatric Dentistry. She has been working as a pediatric dentist at Prince Sultan Military Medical City, in Riyadh since 2013.

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