ORIGINAL RESEARCH PAPER

Dental Science

KEY WORDS: Kindler syndrome, Epidermolysis bullosa, severe gingival bleeding, periodontal bone loss, gingivitis.

DENTAL MANAGEMENT OF A PEDIATRIC PATIENT WITH KINDLER SYNDROME- A REVIEW

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ABSTRACT

Kindler syndrome is a rare autosomal recessive genetic disorder which is a form of Epidermolysis Bullosa. It is characterized by skin fragility and acral blister formation beginning at birth. We hereby present a case of Kindler syndrome. The case is being reported for its rarity and a number of general features like blistering, cutaneous atrophy, and/or poikilodermatous skin changes and oral features such as gingival swelling, advanced periodontal bone loss, mild-to-severe gingivitis, dental caries, and leukokeratosis of buccal mucosa along with its management.

INTRODUCTION

Pedodontics is the speciality which gets to deal with every dental problems of child from birth through adolescence. It is considered to be one of the challenging branches as it not only deals with the dental aspect but also with the behaviour shaping of a child. A Pediatric dentist must be aware of various disorders and syndromes associated with dental anomalies for better diagnosis and treatment.

Kindler syndrome is a rare autosomal recessive genetic disorder which is a form of Epidermolysis Bullosa. It is characterized by skin fragility and acral blister formation beginning at birth. Later followed by clinical features like diffuse cutaneous atrophy, photosensitivity usually most prominent during childhood and decreases after adolescence, poikiloderma, diffuse palmoplantar hyperkeratosis, and pseudosyndactyly.[1] It was first mentioned in 1954 by Theresa Kindler in a 14-year-old girl with acral blistering since childhood who subsequently developed poikiloderma and photosensitivity.[2] It results from mutation in the KIND1 gene (FERMT1), localized on the short arm of chromosome 20.[3] These genes encode a protein called kindlin-1 which is involved in actin-extracellular matrix linkage and affects cell adhesion, signaling, morphogenesis, differentiation and migration. Other areas that get affected are the mouth, eyes, esophagus, intestines, genitals, and urinary system, causing these tissues to be very fragile. These children often experience constipation and dysphagia since birth. Onset of periodontal disease most commonly began in early adolescence.[4] Literature has briefly reported the dental findings which includes oral lesions, atrophy of buccal mucosa, limited oral opening, malocclusion, dystrophic teeth, ankyloglossia, bleeding gums, lip erosions and geographic tongue, atrophy of gingiva, erosion of the hard palate, gingival swelling and desquamative gingivitis.[5,6,7]

CASE REPORT

Chief Complaint And Medical History

A 16-year-old female patient presented with the chief complaint of bleeding gums and burning sensation in mouth while chewing food and also difficulty in swallowing food from past 5-6 months for which they have consulted a physician.

As reported by patient's mother her daughter got diagnosed with kindler syndrome at 5th day after her birth as the child had blisters seen on both the feet which later started appearing all over the body for which they took medical attention. Gradually the skin became scaly, papery and patchy. Patient was undergoing medical treatment since then. According to the medical records, patient was hospitalized a month back to get her right eye operated as she was diagnosed a neurotropic ulcer.

GENERAL EXAMINATION

On cutaneous examination multiple hypopigmented and hyperpigmented poikilodermatous patches were seen all over the face, neck, back, trunk, upper and lower limbs Figure. 1. Keratoconjunctivitis and neurotropic ulcers was also present for which the patient got operated Figure. 2. Skin over the neck, hands and feet was dry, atrophic and photosensitive to the sunlight. Dorsum of hands and feet showing characteristic cigarette paper like wrinkling Figure. 3.

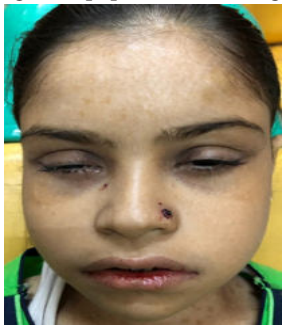

Figure. 1 Face showing hypopigmentation and hyperpigmentation patches

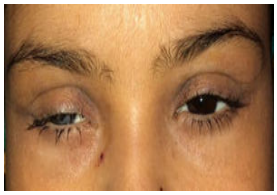

Figure. 2 Right Eye Affected With Neurotropic Ulcer

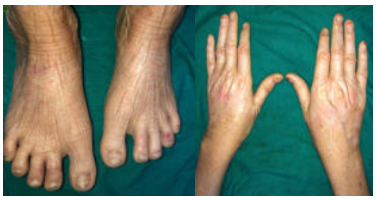

Figure. 3 Dorsum Of Hand Showing Cigarette Paper Wrinkling

ORAL EXAMINATION

On examination of oral cavity, chronic gingivitis along with bleeding marginal gingiva, periodontitis and ulcerations of buccal mucosa are was observed Figure.4,5,6.

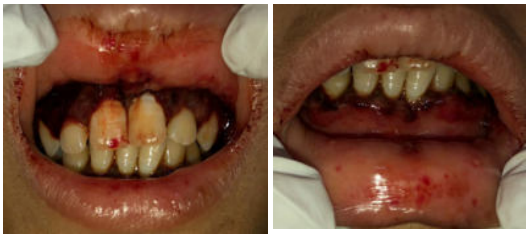

Figure. 4 Bleeding Gums And Ulcerated Lips

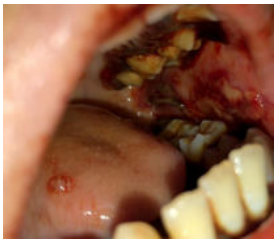

Figure. 5 Ulcerated buccal mucosa and tongue

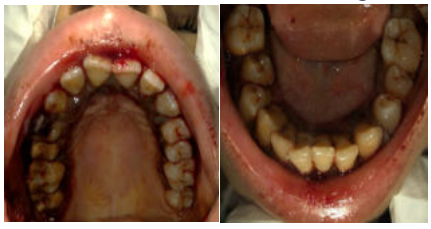

Figure. 6 Occlusal View Of Maxillary Arch And Mandibular Arch

DIFFERENTIAL DIAGNOSIS

Kindler syndrome needs to be differentiated from Weary syndrome, Hereditary Sclerosing Poikiloderma, Rothmund-Thomson syndrome, Bloom syndrome, Cockayne syndrome, dyskeratosis congenita, epidermolysis bullosa, and xeroderma pigmentosum. Immunostaining with anti-kindlin-1 antibody can be used as recent diagnostic test for Kindler syndrome.

TREATMENT CONSIDERATIONS

- In such cases an interdisciplinary approach is required to understand the disease better and plan out the treatment well.
- Good wound care including the use of topical and systemic antibiotics for infected bullous lesions and ulcerations might reduce the morbidity.
- Zinc and multivitamins must be supplemented before and after any invasive procedure or dental procedures like oral prophylaxis and gingivectomy followed by prescribing mouth rinse with peroxide solution.
- Diet rich in sucrose is considered necessary to provide sufficient caloric intake to maintain the patient's growth.

For the above case oral prophylaxis was carried out followed by oral hygiene maintenance using soft baby-size toothbrush and peroxide mouth wash (for 5 days). Due to excessive bleeding the post-operative pictures were not taken and also to avoid discomfort to the patient. Patient was kept under regular follow up.

DISCUSSION

Kindler syndrome has been recently added in the classification of Epidermolysis Bullosa.[8,9] A very few cases have been reported in literature with this syndrome along with its oral features and dental management. Such patients are often presented with mucosal manifestations where oral and gastrointestinal mucosa are frequently affected. As it is a type of Epidermolysis Bullosa similar dental management can be carried out for patients with Kindler syndrome. Wright in 1999 has mentioned dental problems can be taken care of along with anaesthetic and restorative techniques.[10] Skogedal et al. in 2008 demonstrated that caries can be successfully prevented by continuous follow-up on diet, oral hygiene habits, frequent professional cleaning, and fluoride therapy.[11]

It was observed in a cross-sectional study involving 20 individuals with Kindler syndrome showed periodontitis with an early onset (teenage years), and its rate of progression was rapid, resembling aggressive periodontitis, compared to individuals without the syndrome of the same geographic and ethnic origin.[4] The treatment strategies included the use of a disclosing agent to reveal dental plaque, professional prophylaxis, topical applications of 0.12% chlorhexidine gluconate and 1.23% acidulated phosphate-fluoride gel. The goals are mainly to provide the patient with an effective psychological conditioning, motivation and stimulate commitment with the treatment.[12] Tooth paste containing Menthol can cause discomfort for children with Epidermolysis Bullosa. In this case a tooth pastes without menthol, yet containing fluoride is a good alternative. Depending on the severity of disease and the ability to open the mouth, soft bristles or a small head brush should be prescribed to improvise the oral health of the child affected by the syndrome. Ultrasonic powered toothbrushes with a very small brush head have proven useful. Also aids such as interdental brushes, Q-tips or gauze swabs can be used, the dentist or dental assistant should find the aids that are most suitable for the child. Dental treatment under local anesthesia can be performed depending on the patient cooperation. General anesthesia is indicated in patients with small of a mouth opening, severe pain and blistering, or the need for extensive treatment.

CONCLUSION

For patients with Kindler syndrome a regular follow up along with good oral hygiene is mandatory as these patients often show severe periodontal problems which may lead to difficulty in performing the daily routine activities.

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