

## Periodontal Management of a Patient with Kindler Syndrome

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### ABSTRACT

**Background and aim:** Kindler syndrome is a subtype of epidermolysis bullosa with gingival fragility and periodontitis as common oral manifestations of these patients. Because of the early onset and rapid progression of periodontitis in these patients, clinical management of their oral status is an important aspect of their multidisciplinary care and treatment.

**Case presentation:** We present a successful maintenance case report of a patient who has been followed in Hamadan University of Medical Sciences Periodontology Department. The patient was given conservative nonsurgical periodontal treatment and followed every 3 months for one year.

**Conclusion:** The periodontal status of these patients could be well managed by accurate treatment and a good maintenance program

**Keywords:** Kindler syndrome, oral manifestations, recessive epidermolysis bullosa.

### INTRODUCTION

Epidermolysis bullosa (EB) is composed of a group of genetically determined skin disorders characterized by blistering and

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mechanical fragility of the skin and mucosa. In the most recent classification there are four major EB types and over 30 EB subtypes. The four major EB groups include intraepidermal EB (simplex), junctional EB, dermolytic EB (dystrophic), and mixed EB (Kindler syndrome, KS).<sup>(1)</sup> Depending on the specific EB type there

can be significant morbidity involving the soft and hard tissues of the craniofacial complex. KS can be difficult to diagnose in neonates and infants because of the clinical overlap with other forms of EB. However, with increasing age, the appearance of signs such as progressive skin atrophy and poikiloderma and the reduction in blisters and photosensitivity may help in the correct diagnosis of KS.<sup>(2)</sup> From a molecular point of view, KS results from disruption of the actin cytoskeleton-ECM anchorage network.<sup>(3)</sup>

This syndrome was first reported by the German pediatrician Theresa Kindler, in 1954.<sup>(4)</sup> The main clinical features of KS are blistering, especially in trauma-prone sites, skin atrophy and progressive poikiloderma on the dorsal aspects of the hands and feet,<sup>(3,4)</sup> early-onset destructive periodontal disease and severe desquamative gingivitis causing bleeding gums and loss of teeth.<sup>(3)</sup> Mucosal involvement including anal, vaginal, urethral, and oesophageal stenosis,<sup>(5,6)</sup> severe colitis and bloody diarrhea<sup>(7)</sup> is frequently seen in individuals with KS. Finger webbing, pseudosyndactyly, squamous cell carcinoma<sup>(8,9)</sup> and nail dystrophy are other reported features. Their histological abnormalities depend on the site where their skin biopsy is taken. Transmission electron microscopy (TEM) may be helpful in the diagnosis of KS but is often nonspecific or it may show different findings in affected individuals.<sup>(10)</sup> We have no reports of effective gene, protein, cell or

drug therapies so far; therefore, treatment is largely symptomatic. Genetic counseling can be given to the patient and his or her family. Periodontal health is a main area of concern for dental therapy.<sup>(11)</sup> Dental treatment of patients with KS and other types of EB should be carried out carefully to avoid the formation of new bullae during management; use of soft toothbrushes and irrigation techniques can be recommended. Purée diets might be recommended because of the lesions involving the oral mucosa and esophagus. There is also a need for diet supplements, such as vitamins, proteins and iron in order to avoid anemia.<sup>(12)</sup> Use of corticoids and immunosuppressive drugs has also been suggested for the treatment of EB.<sup>(13)</sup> **CASE REPORT**

A 23-year-old male patient referred to the Department of Periodontology, Hamadan University of Medical Sciences, with a chief complaint of calculus and bleeding gums with any small trauma. In his clinical appearance poikiloderma and skin atrophy, especially on the dorsal aspects of the hands, and pigmented skin could be observed (Figures 1 and 2).



**Figure 1. View of the patient's face with pigmented skin.**



**Figure 2. Poikiloderma and skin atrophy, especially on the dorsal aspects of the hands.**

His dermatological examinations had diagnosed him with autosomal recessive mixed type of EB (KS). In his oral finding we could see high plaque index (40%) and severe generalized inflammation of the gingiva which bled quite easily (Figures 3 and 4).



**Figure 3. High plaque index.**



**Figure 4. Severe generalized inflammation of the gingiva which bled quite easily.**

There were quite a few teeth with caries needing endodontic and restorative treatments. The upper right first molar had been extracted. There was no severe bone loss observed in radiologic examinations except for the anterior maxillary incisors and upper left first molar area (Figure 5).



**Figure 5. There was no severe bone loss in radiologic examination except for the anterior maxillary incisors and upper left first molar area.**

There were gingival enlargements and pockets that could be probed. Probing depths were less than 5 mm in most areas.

In the first session of treatment we tried to perform conservative supragingival scaling due to the extremely bleeding gums. We also gave the patient proper and careful instructions and guidance on how to improve his oral health by brushing properly and using antiseptic chlorhexidine mouthwash. Complete scaling and root planing of each arch was performed in a separate session taking care not to cause irritations and trauma to the gingiva and oral mucosa. When the patient was assessed 4 weeks later the gingival status was extremely better. He reported mild irritation of the mucosa after treatment. There was gingival enlargement and probing depth in

an area which still had bleeding on probing. Therefore, we decided to treat this region by gingivectomy with electrocautery to prevent bleeding (Figure 6).



**Figure 6. Treatment with electrocautery to prevent bleeding.**

The patient was then referred to the Restorative Department for his restorative treatments. We scheduled a follow-up maintenance phase for him to be followed every 3 months. He tried to cooperate as much as he could. At present his periodontal status is satisfactory and quite stable with no bleeding and no pockets.

## DISCUSSION

There is tremendous genetic heterogeneity and marked variations in clinical phenotypes in the group of EB disorders.<sup>(1)</sup> A careful approach to KS is advised as blisters can form after dental treatment such as scaling. The scarce literature available for these patients suggests periodontal health as a main area of concern for dental therapy.<sup>(11)</sup> The management of KS is largely symptomatic. The skin in KS is often dry and pruritic and may require frequent topical application of emollients. Photo protection is recommended because of the development of photosensitivity in

KS; skin in many affected individuals becomes red within minutes of sun exposure. There is an increased risk of squamous cell carcinomas; therefore, repeated screening checks are indicated for premalignant keratosis and early detection of malignancy. Regular dental care is advised because of erosive gingivitis and periodontitis in KS. In cases with severe esophageal dysfunction, temporary parenteral nutrition may be necessary. Affected individuals with colitis-like symptoms may develop iron-deficiency anemia. Sometimes surgical bowel resection may be required for severe colitis. In pregnant women with KS, careful obstetric planning, such as consideration of an elective cesarean section, should be considered because vaginal stenosis may occur in KS.<sup>(14)</sup>

Our patient was given conservative nonsurgical periodontal treatment and we tried to manage his specific complication, including bleeding gum, and also his skin lesions by advising him to regularly visit his dermatologist. Treatment of this patient was managed and he was under a strict maintenance program at Hamadan Medical Sciences University Periodontology Department in consultation with his physician.

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