Oral Manifestations and Dental Management of Epidermolysis Bullosa Simplex

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ABSTRACT

Epidermolysis bullosa (EB) is a group of hereditary chronic disorders, characterized by fragility of the skin and mucous membranes in response to minor mechanical trauma. The objective of this study was to report the case of a young girl diagnosed with epidermolysis bullosa simplex (EBS), transmitted by an autosomal dominant gene. Cutaneous findings included blisters and dystrophy following minimal friction. Recurrent blisters and vesicle formation on the hard palate were the main oral findings. In conclusion, publications concerning the oral and clinical manifestations of EBS are important for providing knowledge and an early multidisciplinary approach that prevents blister formation and improves these patients’ quality of life, with the dentist playing an important role in oral health management.

Keywords: Epidermolysis bullosa simplex, Oral manifestations, Alternative therapies.

CASE REPORT

The most prevalent type of EB is the EBS, which mostly involves feet, hands and neck. Histological analysis reveals that its cleavage level is above the basement membrane.5 Local pain is the most common symptom and avoiding friction will prevent lesions.6 The maintenance of skin integrity is a serious challenge for dental practice.7 Therefore, the aim of this study was to report the case of a girl with EBS, describing the clinical features and the precautions that help improve patient’s quality of life, particularly in relation to dental treatment.

INTRODUCTION

Epidermolysis bullosa (EB) is a heterogeneous group of hereditary disorders characterized by extreme fragility of the skin and mucous membranes, which gives rise to the formation of blisters following minor trauma.1 This dermatological condition is a severe autoimmune disease.2,3

There are four major types of EB that differ phenotypically and genotypically: simplex (EBS), junctional (JEB), dystrophic (DEB) and Kindler’s syndrome.4 Transmission electron microscopy (TEM) is considered the ideal method for diagnosing this pathology.4

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The recurrent blister lesions continue to develop mostly on the hard palate, but she never had any systemic complications related to EBS.

**DISCUSSION**

Epidermolysis bullosa is a challenge to health professionals because there is no definitive cure. Skin care attempts to minimize the severity of blister lesions due to the pain, risk of infection and dissatisfaction with appearance. Epidermolysis bullosa is a prime example of a dermatological condition that has a profound psychological impact across all aspects of health. Depression and shame are very common as a result of the appearance. The patient described in this study is shy.

All major types of EB are characterized by blisters following mild mechanical trauma. Many patients with EB can present systemic complications, such as ocular, genital and oropharyngeal infections, involving difficulty in swallowing. The patient described in this study was diagnosed early and has not developed any complications or disturbances in swallowing which is in agreement with Fortuna et al.

Epidermolysis bullosa patients require special precautions during dental treatment because of the greater probability of lesioning the soft tissue when handling cutting instruments close to the skin and oral mucosa. Cariogenic food, limited mouth opening caused by wounds and poor oral hygiene caused by pain are predisposing factors to dental caries. In this case, minimal intervention has so
far preserved the oral cavity and monthly topic fluoride application helped to control dental caries. The patient maintains continuous contact with the health team to avoid complex treatments.

Numerous alternative therapies are used as first aid treatment for blisters. The application of aloe vera gel (bright sparkling, forever living products, Scottsdale, Arizona, USA) diminish the subdermal temperature, providing a refreshed sensation, reducing the healing period and promoting antimicrobial activity.\(^{13}\) The decrease in blister formation due to oral moisturizing and saliva stimulation is the reason Biotene mouthwash (GlaxoSmithkline, USA) was prescribed. This product possesses buffering capacity, an immunological effect, antimicrobial activity and a self-cleaning effect.\(^{8}\)

Epidermolysis bullosa treatment is generally focused on support. Perforating the blisters contributes to accelerating the healing process and prevents continued lateral spread of the blisters. Currently, researchers are focusing their attention on gene and cell therapy, recombinant protein infusions, intradermal injections of allogenic fibroblasts and stem cell transplantation. Other developing therapies are directed toward the enhancement of wound healing and better quality of life for EB patients.\(^{14}\)

A multidisciplinary approach involving the following health professionals is essential: nutritionist, pediatrician, dermatologist, plastic surgeon, hematologist, gastroenterologist, ophthalmologist, cardiologist, pediatric dentist, nurse and occupational therapist.

The girl comes to the dental office every month to maintain her oral health. She attends dermatological reevaluations sporadically and, once a year, returns to her pediatrician for control exams. This girl has gotten used to soft food and to avoiding certain physical activities during dental treatment because of the greater probability of blister formation. Moreover, those patients require an early multidisciplinary approach to improve their quality of life, with the dentist playing an important role in oral health management.

CONCLUSION

This case emphasizes that patients with EBS need special precautions during dental treatment because of the greater probability of blister formation. Moreover, those patients require an early multidisciplinary approach to improve their quality of life, with the dentist playing an important role in oral health management.

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