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Dental Management of a child with Dystrophic Epidermolysis Bullosa under general anaesthesia.

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Abstract

Epidermolysis Bullosa (EB) is a group of rare genetics vesiculobullous disorders affecting the skin and mucous membrane. Affected patients usually present with continuous blister formation even after minor trauma. A three-year-old boy with dystrophic epidermolysis bullosa (**DEB**) referred to the Dental and Maxillofacial Surgery Department with dental pain. On examination, the child had extensive blisters and scars all over the body, pseudo-syndactyly of the hands and feet and loss of the toenails and fingernails. Intra-oral features included microstomia, limited mouth opening, poor oral hygiene, generalized enamel hypoplasia and grossly carious teeth. The dental treatment was done under general anaesthesia (GA), and it included multiple restorations and extractions. Post-operatively, the patient recovery was uneventful, and he was discharged a day following the surgery. Many oral and systemic manifestations usually complicate dental treatments for EB patients. Careful planning and liaison with the anaesthetist are important when dental care anticipated to be under GA.

Keywords: Epidermolysis Bullosa (**EB**), Vesiculobullous disorder, General anaesthesia (**GA**), Blister, Dentistry

Introduction

Epidermolysis bullosa is a rare genetic disorder characterized by skin and mucous membrane fragility and the systemic manifestation of variable severity [1, 2, 3]. It was reported for the first time in 1871 by Herba [2]. The vesiculobullous lesions usually appear in response to trauma, exposure to heat or occurs spontaneously without obvious cause [4]. The lesions slowly heal and are accompanied by scar formation. The onset of the disease is usually immediate after birth up to the age of 5 years [3]. The disease diagnosed by different special tests such as immunofluorescence mapping and transmission electron microscopy [5]. This rare disease inherited as an autosomal dominant or autosomal recessive trait with the prevalence ranging from 1:50,000 to 1:500,000 of live birth [1, 4]. Many dentists have limited knowledge of the disease and how to deal with it in the clinical setting. There are four major types; simplex (EBS), junctional (JEB), and dystrophic (DEB), with classification based on the target

protein's location and the level of the blister within the dermal-epidermal junction [4, 5, 6].

This disorder can be associated with high morbidity and mortality [3], ranging from mild form limited to blister formation only to the severest form causing premature death [2, 7]. The general finding of EB may show the involvement of the whole skin, including blisters, erosions, pigmentations changes, and the absence of the finger and toenails. The extra-cutaneous manifestation may extend to involve the eyes, teeth, oral mucosa, gastrointestinal tract, anus and musculoskeletal system [7]. The oral manifestation varies according to the severity of the disease and the type from localized blister to a more severe form, which involves the whole oral mucosa. In more severe cases, microsomia, ankyloglossia and obliteration of the oral vestibule is seen [1, 8]. Involvement of the tooth structure occurs in the form of hypodontia, defective crowns, cementum, and enamel hypoplasia, leading to high caries risk [2]. With continuous scar formation and trauma from even gentle brushing, patients presented difficulties in maintaining adequate oral hygiene (OH), making them more susceptible to caries [1,2].

Up to date, there is no definitive cure for EB, and whatever treatment is to control the recurrent infection, pain and improve quality of life [6, 9]. Providing adequate dental care to children with this disorder can be challenging.

Furthermore, in most cases, dental treatment under GA is the choice to manage the poor cooperation level and less blister formation [1]. General anaesthesia can be challenging in epidermolysis bullosa, causing multiple complications. Some of these include chronic anaemia, securing venous access and blister formation during intubation. The following case report presents the dental management of a 3 year old boy with DEB.

Case report

Three years old male with DEB referred to the Paediatric Dentistry clinic at the Dental & Maxillofacial Surgery department at Sultan Qaboos University Hospital (SQUH) to manage recurrent dental pain from the mandibular molars. The child was born with normal delivery with a birth weight of 2.8 kilograms to consanguineous parents. He was diagnosed with EB at birth, and one of his siblings was a known case of DEB.

The child was 11kg at presentation; physical examination revealed generalized blisters, pseudo-syndactyly of the hands and feet, loss of the toenails and fingernails and extensive skin scarring (Figure 1&2).

Psychologically, he appeared to have a normal cognitive function. The child was complaining of recurrent pain from lower molars teeth. Extra-oral examination showed multiple blisters and erosions on the face, some on the healing stage.

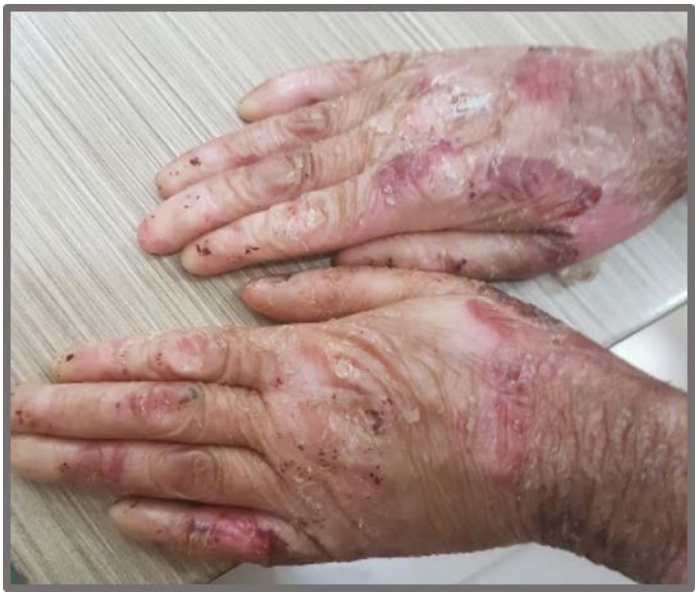

Figure 1: Generalized blisters and pseudo-syndactyly of the hands

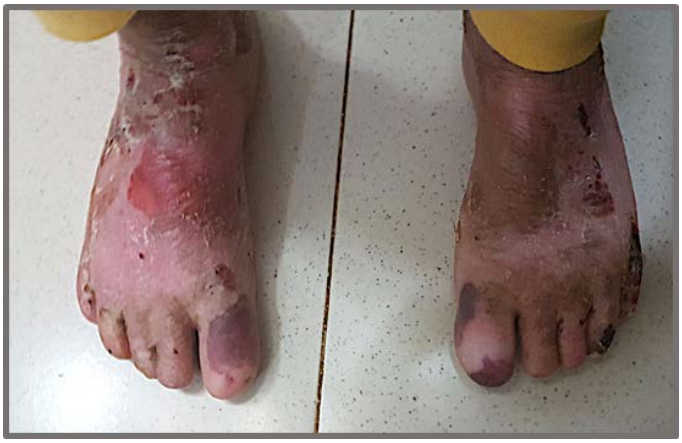

Figure 2: Generalized blisters and pseudo-syndactyly of the feet's

The perioral structure showed fibrous bands bilaterally at the commissures of the lip with the limited aperture. Intra-orally he had microstomia, poor oral hygiene and primary dentition with a normal count, generalized enamel hypoplasia and grossly carious teeth (Figure 3). The child had poor cooperation and showed negative behaviour.

The clinical findings and treatment options with risks and benefits discussed with the patient's parents, and the agreed plan was comprehensive dental care under GA. The treatment's primary phase involved preventive care, including oral hygiene instructions, diet counselling, and fluoride advice. The patient referred to the pre-anaesthetic clinic for an anaesthesia assessment.

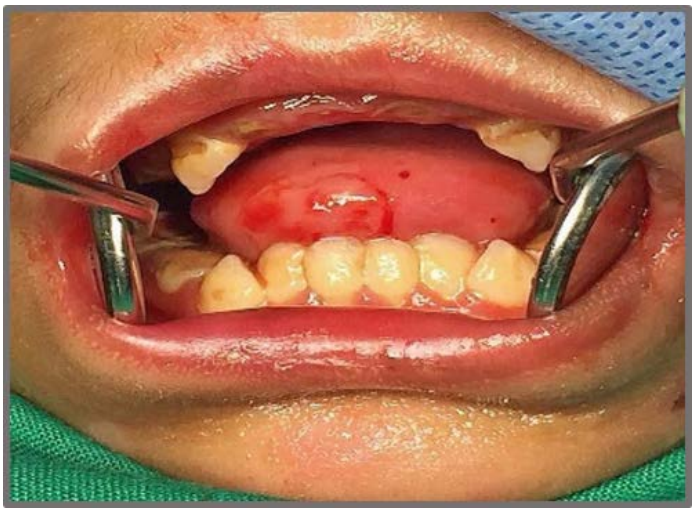

Figure 3: Pre-Operative photo showing the severe hypomineralisation of the teeth

Admission was arranged at a paediatric ward one day before the procedure. Pre-operative blood investigations avoided due to the poor cooperation and risk of trauma. Intraoperatively blood was collected as recommended by the physician and anaesthetist, and it included full blood count, coagulation and electrolytes profiles. The significant finding was a low haemoglobin level (Hb) (7.5 g/dL). In the operating theatre, a soft sheet was placed on the operating table to avoid skin friction. Defibrillator pads used to attach the electrocardiography (ECG) electrodes to the chest (Figure 4), and a non-invasive blood pressure cuff applied along with cotton pads around the arm.

The dental treatment included restoration of all primary canines with composite resin, extraction of maxillary primary molars and incisors and mandibular primary molars. The mandibular primary incisors disked, and fluoride varnish was applied. The lips and perioral tissues generously lubricated throughout the procedure with petroleum gel, and any bullae formed during the treatment were evacuated (Figure 5).

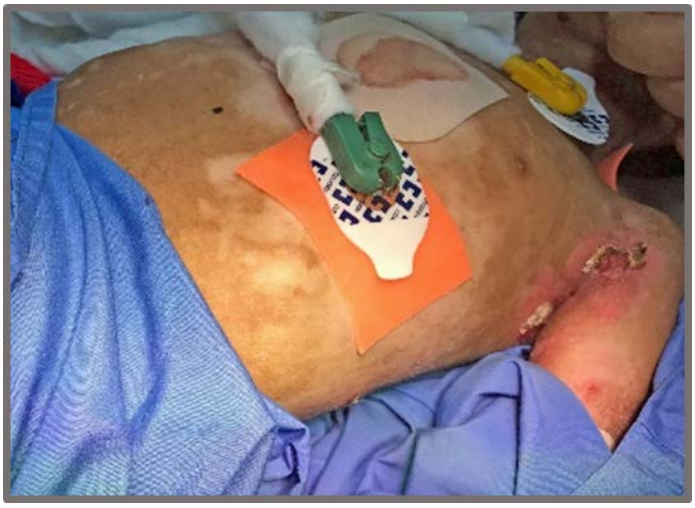

Figure 4: Defibrillator pads to reduce trauma caused by the ECG lead.

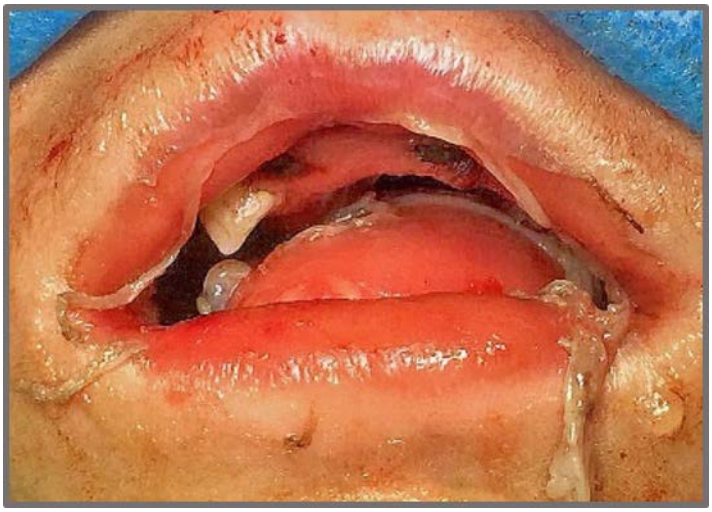

Figure 5: Sloughing of perioral skin and mucosa of the lip.

As the patient comes from an area long distant where tertiary medical care is not available, we referred him to the Paediatric Ophthalmology, dermatology, and genetics team to assess and provide needed care.

Post-operative recovery was uneventful, and he was discharged after 24 hours with analgesics.

Follow up appointment arranged; however, the patient failed to attend as they live in an area far from SQUH. Review appointment arranged in the local dental center but again they did not attend.

Discussion

EB is a rare group of genetic disorder affecting the skin and mucous membrane and characterized by the development of blisters to minimal trauma [10]. Among the four types of the disease, dystrophic epidermolysis bullosa is the severest form and caused by a mutation in the type VII Collagen gene [6]. DEB further subclassified into severe generalized recessive, recessive and dominant.

DEB characterized by deformities of the skin, including coalescence of the fingers, blistering, scarring, nails changes and milia formation [6]. Acquired syndactyl in hand and feet following cicatricial tissue formation may eventually lead to loss of finger and toenails and recurrent skin infections. In our case report, the child presented with many of these dermatological deformities like generalized blisters, pseudo-syndactyly of the hands and feet, loss of the toenails and fingernails and extensive skin scarring.

Patients with EB are usually associated with many systemic complications extending to the ocular, genital and oropharyngeal structures. The gastrointestinal tract can be severely compromised by scarring and tissue contraction, leading to difficulties in swallowing [4,11]. However, in our case, the child did not exhibit any swallowing difficulties, but he had inadequate oral intake. The anaemia in EB is multifactorial in origin, and its severity varies with the type of epidermolysis bullosa [5]. In our patient, many factors were responsible for the low Hb level (7.5 g/dL), like repeated skin infections, inflammation, poor nutrition, and extensive blistering, which causes blood loss.

DEB found to have the most severe oral manifestation involving tooth structure, gingiva, and oral mucosa. Patients with EB require special care during the provision of dental treatment as they are more prone to lesion formation even with the gentle examination [11].

The physical difficulties in maintaining OH, enamel defect and cariogenic diet make these children more susceptible to dental caries [1,2]. Dental treatment under GA is the treatment of choice due to the extensive work needed and the difficulty associated with mouth opening and fragility of epithelial tissues [1]. In this case, many factors favoured the dental treatment to under GA; the extensive dental caries involved almost all the primary teeth, limited mouth opening, ankyloglossia, and obliteration of the vestibule and poor cooperation.

Regular reviews and long-term monitoring are crucial for caries prevention, and squamous cell carcinoma (SCC) screening reported to occur in the tongue in many cases with recessive DEB [5,7].

Conclusion

The sensitive oral mucosa complicates dental care for EB patient to the minimum contact and challenging airway maintenance, necessitating a multidisciplinary approach in management.

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