

EPIDERMOLYSIS BULLOSA ACQUISITA (EBA): A CASE REPORT

Ahmad EL-Nassry¹, Abrar EL-Gedaily²

1. Ph.D ,Professor, King Fahad Hospital,Dental Center,Pediatric Dentist consultant, Jeddah,Saudi Arabia
2. BDD, Ibn Sina College, Pediatric dentistry Resident, Jeddah, Saudi Arabia

ABSTRACT:

Epidermolysis bullosa acquisita (EBA) is a chronic autoimmune bullous disease characterized by the presence of IgG tissue and type VII collagen antibodies. It is rare in humans and animals with an incidence ranging from 0.2 to 0.5 new cases per million and per year. In this case 2 years male patient came to the dental pediatric department complaining of itchy pain related to his oral mucosa and toothache.

Key words: Epidermolysis Bullosa Aquisita(EBA), childhood, immunofluorescence.

INTRODUCTION:

Epidermolysis bullosa is a general term that combines several types. It can be divided into three main hereditary types based on the mechanical defect in the basement membrane of the skin.

The symptoms of each type are determined according to the location of the defect. Junctional EB is characterized by lack of adhesion of the skin through the basement membrane. It composes approximately 5% of all EB types.

The outlook of junctional EB is usually rather bleak; half of the children die within the first two years as the result of malnutrition and anaemia, which are

caused by serious blistering in the pharynx and the oesophagus. Characteristic clinical features are damage to and marking of the skin tissue on the face.(fig.1)

Figure1



Dystrophic Epidermolysis bullosa is characterized by a lack of adhesion of the skin under the basement membrane. It composes Approximately 25% of all EB sufferers. It derives its name from the tendency of the blisters to heal with scarring.

This process can lead to contraction of the joints, fusion of the fingers and toes, contraction of the mouth membranes and narrowing of the oesophagus. There is a wide variation in the severity of dystrophic EB. In general, dystrophic EB is not life--- threatening in childhood.(fig.2)



Figure 2

Epidermolysis bullosa simplex is characterised by lack of adhesion of the skin right above the basement membrane. It composes Approximately 70% from EB simplex. The most common form of EB simplex has blistering confined to hands and feet. In another form of EB simplex blistering can occur all over the body. Blistering may appear during the neonatal period but it can also

manifest itself in later childhood (or even in adult life). (fig.3)



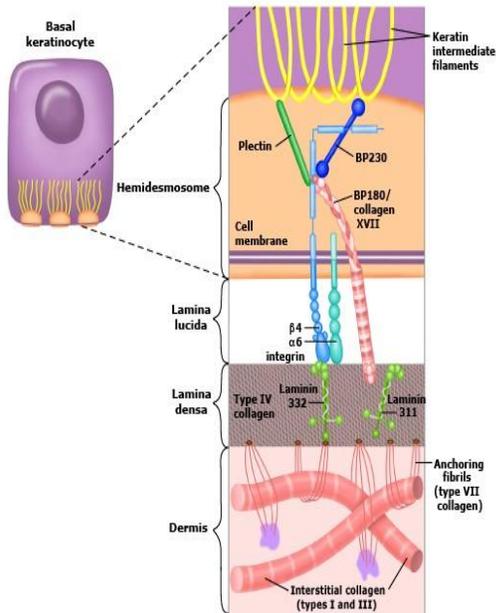
Figure 3

And one acquired rare type called Epidermolysis bullosa acquisita.

CASE DETAIL:

A 2 – years--- old male patient came to the pediatric Dentistry Department complaining from severe pain associated to multiable cairase and oral ulcers .Medical history showed that the patient diagnosed with Epidermolysis bullosa acquisita) (EBA) which is defined as a chronic autoimmune bullous disease of the skin and mucus membrren characterized by the presence of IgG tissue and type VII collagen antibodies located below the lamina densa of the basement membrane, which shares greater than 80% homology in amino acid sequence with the human NC1 domainleading to the formation of bullae in the dermoepidermal junction and blistering. ^[1, 3]

The skin and mucus membrane have three clinical variant in case of (EBA),A noninflammatory or mildly inflammatory Variant affecting trauma---prone extensor skin surfaces, Clinically



this feature resembles porphyria cutanea in elderly patients, and it resembles the dominantly inherited form of epidermolysis bullosa dystrophica in children. And the generalized inflammatory form, which usually heals with minimal scarring formation, this form of epidermolysis bullosa acquisita clinically, resembles bullous pemphigoid or linear IgA bullous dermatosis.

Clinically this case presented with tense vesicles, bullae, and erosions primarily on the extensor surfaces of hands, knuckles, elbows, knees, and ankles. The blisters may be hemorrhagic. Blisters on mucus membranes rupture easily. (fig.4)



Figure 4:



Figure 5

Mucous membranes of buccal, conjunctival, gingival, palatal, nasopharyngeal, rectal, genital, esophageal was involved with nails dystrophy and loss. [2,3,4]. Intraoral Examination shows redness, mucosal peeling, scars, and several eroding ulcers on the tongue, lip, palate compounded with the additional complication of candidiasis, which explain the burning and itchy feeling that patient complains of it. [8] (fig.5)

DISCUSSION:

EBA is a rare condition in humans and animals and it has been reported in dogs only, the incidence of EBA in human ranging from 0.2 to 0.5 new cases per million and per year, some studs shows that EBA doesn't affects certain race or gender but others shows that females are more susceptible than males and it also shows African and Korean are more affected by EBA than others. Significantly, both agreed that most cases are in middle age, but generally the rang of onset is very wide from childhood to the adult life. [4, 7]

A good communication between dentist and patient physician to get the proper diagnosis and treatment in the proper time is very important which is depending in the severity of the case. Some literatures shows that Systemic corticosteroids are the gold standard in treatment but there is no proof that they are the best treatment, [8] others claimed, that the Direct

immunofluorescence is the gold currant standard to establish a diagnosis for EAB or by using of Serological tests (Indirect IF microscopy) of the patient's serum which is act as a screening test for circulating antibodies. [3] But there is another opinion said that colchicine is the first line of treatment due to its effectiveness and low side effect profile. [4] Physicians could also consider using cyclosporine, extracorporeal photochemotherapy, intravenous immunoglobulin and intravenous infusion of anti-CD20 antibodies (rituximab). [5, 8]

Patients who are treated with systemic corticosteroid for longer than 1 month, a combined supplement of calcium and vitamin D should be instituted to prevent osteoporosis. Infection due to open erosions and wounds, Malignancies secondary to chronic inflammation and immunosuppressive treatments, Bone marrow suppression, Growth retardation, adrenal insufficiency and osteoporosis all are possible complications secondary to medication. [6, 8]

Patients with epidermolysis bullosa acquisita (EBA) should receive supportive care to reduce the risk of skin trauma which is includes proper wound care and strategies for avoiding trauma (They can wear protective pads to cover their extensor skin surfaces), not using harsh soaps, reducing vigorous rubbing of their skin on wash cloths and towels, and to avoid over washing, especially with hot

water. Sunscreen should be used because prolonged sun exposure may aggravate or promote new lesions. Skin protection prevent infection, patient should be educated about identifying superimposed skin infections and when it is appropriate to seek medical care to improve quality of the patient's life. [4, 8]

Careful manipulation and Co-operation between dentist and parents or caregiver is very important in this case to prevent more trauma and ulcers, achieve more dental care and to maintain a good oral hygiene. Tooth brushing with small head and soft bristle is possible with most EB cases but Cotton buds, disposable mini brushes or gauze can be used to clean the teeth if a patient not able to brush because of soreness, [9, 10,11,12,13,14] rinsing with water after meals or with chlorhexidine 0.12% mouthwash, [9,10,12,15,16,17,18] Topical fluoride applications of high-dose varnish are recommended every 3 months at each dental visit, daily fluoride supplement for children who live in a non fluoridation community is very important. [9,10,15,16]

Proper diet should be obtained to minimize oral damage i.e. soft food, low acidity foods. [6, 8]

Limited mouth opening is the most challenging case could face the dentist. Some exercises by mouth opening stretching, resin plugs or mouth trainer before the dental treatment by half an hour would be useful; some cases

recorded a progression from 19 to 23 after 10 minutes of exercises. [19,20, 21, 22]

CONCLUSION:

The prognosis for (EBA) is highly dependent on the degree of the disease that is present but if the patient treated and cared properly, should expect to live a normal life span. Patients with scarring secondary to epidermolysis bullosa acquisita may develop dysfunction. But significant morbidity may complicate some. [6,8]

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