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Case Report on Epidermolysis Bullosa Acquisita

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ABSTRACT

Epidermolysis bullosa acquisita (EBA) is rare autoimmune blistering of the skin disease but not inherited and characterized by auto antibodies to type-VII collagen. It is a rare with an incidence rate of 0.26 /million population. Treatment include oral corticosteroids (E.g.: prednisone), anti-inflammatory agents, and immuno suppressants (e.g.: azathioprine). Patients who are on long-term oral steroid treatment may be advised to take calcium and bisphosphonates to reduce the risk of osteoporosis that can be associated with steroid use. A case of 13 years old boy initially presented with bullous vesicular lesions all over the body. 5 months history of progressive trauma persuaded papules and vesicles on his hands, and face.

Key words:
Epidermolysis bullosa acquisita, type-VII collagen, Immuno suppressants, Blistering.

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INTRODUCTION

Epidermolysis Bullosa (EB) is a group of rare inherited connective disease that results in blister formation on the skin[1].Alike EB, Epidermolysis Bullosa Acquisita (EBA) is a rare and acquired auto immune sub epidermal blistering of the skin disease but not inherited. The record of public autoimmune sub epidermal blistering disease, bullous pemphigoid (BP), is reported to have a yearly predictable incidence between 2.4 and 21.7 per million. By difference, the estimated incidence of EBA is <0.5 per million. Higher than that of previous reports of the incidence and prevalence of EBA estimated in South Korea. 2.8 cases per million has recently been reported in Germany[2].

EBA patients' presence with auto antibodies, tissue bound IgG directed against Type-VII collagen (COL7) within anchoring fibrils located at the dermal-epidermal junction. The loss of anchoring fibrils leads to formation of blisters under the lamina densa[3]. Clinically, it is two types, Classical mechanobullous (non-inflammatory), Non-classical/non-mechanobullous (inflammatory) [4].

Classification of EBA	Clinical features
Classical mechanobullous (non-inflammatory)	• This is the most common form of EBA in Europeans • Tense vesicles and bullae primarily on extensor surfaces of hands, knees, knuckles, elbows and ankles • Mucous membrane blisters rupture easily • Lesions heal with significant scarring and milia (small white spots) • Resembles the inherited form of dystrophic epidermolysis bullosa
Non-classical/non-mechanobullous (inflammatory)	• Widespread tense vesicles and bullae not localised to trauma-prone sites • Often affects the trunk and skin folds • Generalised redness, itching and plaque formation • Heals with minimal scarring and milia formation • Resembles bullous pemphigoid

COL7 expressed in gastrointestinal tract that involves in oral cavity and other mucosal sites has been frequently reported, thus EBA must be consider as mucocutaneous disease. There is involvement of other mucosal membranes in ocular and genital have been repetitively noted in EBA patients and extra cutaneous involvement may also occur. In addition, EBA associated with cancer as well as inflammatory, infectious, cardiovascular, metabolic and neurological diseases were reported. Mostly crohn's disease and systemic lupus erythematous were reported[5].

DIAGNOSIS

The autoimmune diseases can be diagnosed according to their clinical, histo pathological and immunological grounds[6].

Based on clinical

- Onset adult symptoms
- Negative family history

Based on immune grounds

- Immuno fluorescent techniques
- Salt-split skin technique
- Immuno blot procedures
- Electron microscopy
- Serology

TREATMENT

EBA is resistance to treatment. Similar to other AIBD's(Autoimmune skin blistering diseases***)***, the first choice of drugs are systemic corticosteroids use alone or combination with agents including colchicine, diamino diphenyl sulfone (DDS, dapsone), methotrexate (MTX), azathioprine (AZA), cyclosporine (CSA), mycophenolate mofetil (MMF), and cyclophosphamide (CPA) have been reported in treatment of EBA[7]. Prednisolone, Gold and vitamin E are also used. Children who experienced with EBA responds better with treatment of dapsone and prednisone[8]. Other therapeutic options like high-dose intravenous Immunoglobulin (IVIG), rituximab (RTX), plasmapheresis and immuno adsorption (IA), Daclizumab (3 cases only show clinical improvement) and extracorporeal photo chemotherapy (ECP) have been used. Sometimes patients not respond to any treatment[9].

CASE REPORT

A 13-year-old boy admitted with bullous vesicular lesions all over the body, small white spots, pus-filled blisters on both the leg.5 months history of progressive trauma persuaded papules and vesicles on his hands, and face. The first lesion developed on the left humerus region followed by left shoulder, face, and legs (figure 1. A, B, C). His family history was negative for other dermatological diseases. The vitals are normal on examination. His height is 130 cm and weight is 30kgs, BMI is normal. Examination of skin biopsy specimen revealed that the presence of bullae in the supra dermis, perivascular infiltration of eosinophils and plasma cells in the dermis. Based on the subjective and histo pathological evidence the patient was diagnosed as Epidermolysis Bullosa Acquisita. Patient treatment plan includes T. Prednisolone (10mg-oral-0D), T. Cefixime (100mg-oral-BD), T. Cetirizine (10mg-oral-BD), Framycetin skin cream (30gm-Topical), T. vitamin C (500mg-oral-OD), T. Calcium (500mg oral-OD), T. Pantoprazole (40mg-oral-OD), T. Azathioprine(50mg/day-OD), 0.05% Betamethasone ointment and wound management is done.

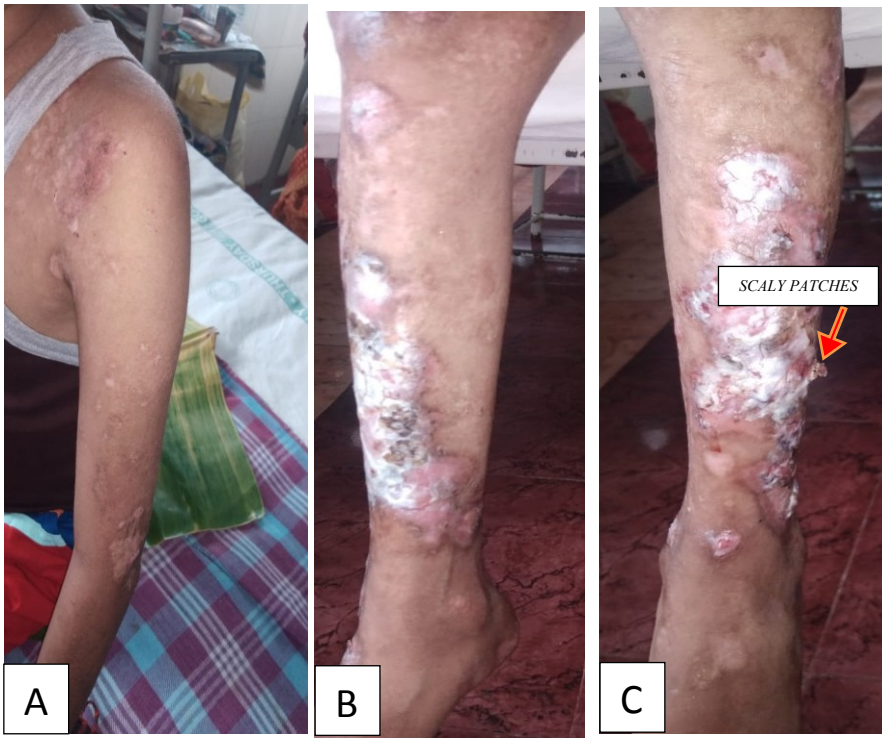

Figure 1. A. first lesion developed on the left humerus region B. small white spots, pus-filled blisters on Left leg, C. scaly and hypopigmented patches on Right leg.

DISCUSSION

In this case, the patient initially presented with bullous vesicular lesions all over the body, small white spots, pus-filled blisters on the both legs. Later these bullae burst leading to formation of scaly and hypopigmented patches involving almost all over the body. He had a past history of progressive trauma persuaded papules and vesicles on his hands, and face at the age of 5 months. Based on physical examination, we initially consider this patient as bullous pemphigoid and started the treatment, includes T. Prednisolone (10mg-oral-0D), Inj. Dexamethasone (8mg-IV-BD), T. Cefixime (100mg-oral-BD), T. Pheniramine (5-15mg-oral-OD), T. Cetirizine (10mg-oral-BD), Framycetin skin cream (30gm-Topical), Inj. Cefotaxime (500mg-IV-BD), T. Metronidazole (700mg-oral-BD-half tablet), T. vitamin B. Complex, T. vitamin C (500mg-oral-OD), T. Vitamin A&D (5400IU-oral-OD), T. Calcium (500mg-oral-OD), T. Pantoprazole (40mg-oral-OD), T. Azathioprine (50mg/day-OD), 0.05% Betamethasone ointment, 1% Silver Sulfadiazine cream and wound management is done.

After 2 week, histopathology revealed that the presence of bullae in the supra dermis, perivascular infiltration of eosinophils and plasma cells in the dermis. And there was no improvement in the symptoms with the treatment. Considering both clinical and histopathological aspects, the patient confirmed with epidermolysis bullosa acquisita. At first, the patient was treated with antibiotics, corticosteroids agents due to exacerbation of his condition. By the end of 3 weeks his symptoms were slowly relieved with the treatment provided in the case report and discharged with the following medication:

His discharge medication includes: T. cefixime(100mg-BD-15days), T. prednisolone(20mg-OD-15days), T. pantop(40mg-OD-15days), T. Levocetirizine+ Montelukast(5mg+10mg-oral-15days), soframycin(30gm) + betamethasone

ointment(0.05%), T. vitamin-b-complex(75mg-OD-15days), T. iron folic acid, T. calcium(500mg-OD-15days), T. vitamin c(500mg-OD-15days)

CONCLUSION

As Epidermolysis Bullosa pemphigoid and Epidermolysis Bullosa Acquisita look like, there is a miss perception in our case initially considered as Bullosa pemphigoid based on the symptoms. And started the treatment but there was no change in the complaints. Then after the scaly patches were sent for histopathology study. The study results are conformed epidermolysis bullosa acquisita. Based on the subjective and histopathology the case was diagnosed as epidermolysis bullosa acquisita. And the treatment was started as described in the case report, patient where get relief from the symptoms.

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