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EC PULMONOLOGY AND RESPIRATORY MEDICINE

Case Report

A Case Report on Epidermolysis Bullous Acquisita

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Received: May 10, 2018; **Published:** June 19, 2018

Abstract

Epidermolysis bullosa acquisita (EBA) is an ideal autoimmune disease with a well-categorized pathogenic consequence of autoantibody binding to the target antigen. EBA is a sporadic illness with an incidence of 0.2 new cases per million and per year. The nature of the disease usually hints to skin firmness and secondary scarring that often causes restriction in the movement of skin surfaces. The onset of disease to some degree is rapid and more extensive. The clinical presentation of this subset of patients is indefinite from other type of pemphigoid. In this article, we present a case of 30 year elderly old female with complaints of itching all over the body and fluid filled lesions over right forearm and upper chest. She was prescribed with topical steroids and prednisolone.

Keywords: *Autoimmune Disease; Type VII Collagen; Clinical Presentation; Therapy*

Introduction

Epidermolysis bullosa acquisita (EBA) is a very rare disease in which blisters appear at sites of trauma mostly occurs to skin and mucus membranes. In this disease condition the auto antibodies targets the type VII collagen which is associated with the connection of basement membrane to the dermal layer. It is the main substance involved in anchoring fibrils EBA is rare in humans but mostly reported in dogs. When EBA is occurred, blisters are most commonly seen with some mucosal discharge at the affected areas and are left with dark scars after healing. The blisters that are formed in the epidermolysis bullosa acquisita are first seen on the trunk followed by flexural areas. It would be occurred at any age but most commonly seen in elders than the children even though reported at 3 months of age. EBA can be discriminated from other skin diseases i.e. bullous condition based on the signs and symptoms like the presence of the characteristic lesions, onset of disease and family history [1]. The current therapy for EBA generally relies on immunosuppressive therapy.

Case Report

A 30-year-old elderly female presented with chief complaints of itching all over the body and fluid filled lesions over right forearm and upper chest since five days. She was known case of EBA and she was on therapy with T. prednisolone 20 mg and stopped medication since 20 days. Present illness started as itching all over the body, on scratching fluid filled lesions appeared which ruptures spontaneously to form raw area over right forearm, upper chest and buttocks. Patient attained menopause 8 months back. Her lab investigations found to be normal and on cutaneous examination multiple, hemorrhagic, crusted plaques with erosions present over inframammary region, right forearm and over both foot and soles. Her oral mucosa, hair and nails were found to be normal. Patient was prescribed with topical clobetasol and gentamicin cream, tablet prednisolone. Her symptoms were alleviated during hospital stay and she was discharged with oral and topical steroids.

Discussion

EBA is a bullous dermatosis resulting from the formation of anti-collagen VII autoantibodies and associated with HLA-DR2 [2,3]. Its presentation normally takes place in adult age (higher incidence at the age of 50), with greater preponderance in female patients. various systemic diseases, preferably IBD, systemic lupus erythematosus, amyloidosis, thyroiditis, multiple endocrinopathy syndrome, rheumatoid arthritis, pulmonary fibrosis, chronic lymphocytic leukaemia, thymoma, diabetes, multiple myeloma and other autoimmune diseases are associated with EBA [4,5]. The two different clinical subtypes in EBA could be classified as inflammatory and noninflammatory condition of EBA. Most commonly EBA is a cutaneous inflammation which could resemble various diseases like bullous pemphigoid, brunsting-perry pemphigoid but varies with the signs and symptoms, time period of the disease in the patients or they can possess both the disease conditions at the same time. IgG auto antibodies are composed of heterogeneous subclass and light chain, specific for anchoring fibrils but their complement-activating capacity don't be similar to the inflammatory phenotype. The antibodies by attacking the collagen the strength of the basement membrane is collapsed so results in fragility of skin and the formation of blisters. Although there are no guidelines for the treatment of EBA, conventional therapy includes high potency topical steroids, oral prednisone and dapsone [6,7]. EBA is a long term inflammatory condition in which the symptoms are decreased by proper therapy or else aggravated. The drugs which are used to treat this condition can result in adverse effects that requires symptomatic therapy.

Conclusion

As most types of EBA are inherited it has no cure, mild forms may improve with age. Treatment focuses on symptomatic relief and to prevent complications. Among treatments available corticoid therapy is the most commonly used treatment in patients with inflammatory EBA which is of best choice and is also evidenced.

In this case, clinical improvement was significant after introduction of topical clobetasol and gentamicin cream, tablet prednisolone.

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Volume 7 Issue 7 July 2018