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Bullous Pemphigoid – A Rare Disease Case Report

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Abstract

Bullous pemphigoid is a rare, chronic and a most common blistering disorder, characterized by the appearance of large fluid-filled blisters on the skin. It can be diagnosed based on clinical, immunologic, and histological criteria and is most commonly found in elderly persons. These blisters are often found on areas of the skin in the regions such as – lower abdomen, upper thighs or armpits. It manifests clinically with diffuse eczematous (inflamed or irritated skin), pruritic (unpleasant sensation that desires to scratch), urticarial-like lesions (reddish flat patches or swellings), eventually with the appearance of tense bullae or blisters typically filler with clear fluid. It is an auto-immune disorder, in which our body produces antibodies to the fibers that connect the outer layer of skin (epidermis) and the next layer of skin (dermis) and finally triggering the inflammation process, which leads to the production of blisters. The major aim of this case report is to present a typical case of this condition, to bring awareness on several treatment options, and to advocate referral to a dermatologist given its potential severity. Multiple treatment opportunities have been found for this condition, including anti-inflammatory medications that reduce antibody formation, and treatments to increase the elimination of antibodies [1].

Keywords: bullous pemphigoid, blistering disorder, autoimmune disorder, diffuse eczematous, pruritic urticarial lesions, dermatology

Introduction

Bullous pemphigoid is a rare skin disorder, in which there is an appearance of blisters on the skin, and most commonly found in older individuals. Bullous Pemphigoid can be resolved within five years, and it can be goes away on its own in a few months. The major aim of this case report is to highlight the several treatment options, to bring awareness about the disease and to advocate referral to a dermatologist given its potential severity.

Patient information, clinical findings, timeline

A 52 years old female patient was admitted in the hospital with the chief complaints of large and fluid filled blisters all over the body, with weight loss since 5 years.

Diagnostic assessment

The patient is in a conscious and in a coherent state, with lymphocytosis and with afebrile temperature. The patient is diagnosed with the elevated levels of Serum glucose –fasting and Serum Total Iron with the abnormal values of **140mg/dl** and **24mg/dl** respectively, in Biochemical profile. Haematological profile is abnormal with normal to the platelet count. Microbiological record shows that Serum has been taken as a Specimen, in which the C-reactive Protein levels has been elevated with an abnormal value of **371.16mg/L**, by Immunoturbidometry method. Anti-Nuclear Antibody (ANA) has performed by ELISA method and value obtained is 0.40, considered as a negative value. AntiDS-DNA antibody (ELISA) has also conducted, in which the serum has been taken as a specimen, where the test value was obtained as 8IU/ml which is a negative value.

Therapeutic interventions

She was previously diagnosed with the disease i.e. bullous pemphigoid in a farther hospital, at where those doctors prescribed the following medications:

- Inj. Folitrax
- Inj. Hydrocortisone
- B. Plex Forte
- T. Azitpiral
- T. Folic acid

But still the condition has been worsening, so the patient admitted in another hospital for management and to control her disease.

Follow-up and outcomes

According to all the Laboratory profiles, mentioned above, the patient has been diagnosed with Bullous Pemphigoid disease and physicians prescribed the following medications in order to control or to prevent the further complications of the disease.

S.No	Trade Name	Generic Name	R.O.A	Dose	Frequency

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1.	Tab. OROFER – XT	Ferrous Ascorbate	P/O	1tab	Once in a day
2.	Inj. FERINJECT	Ferric carboxymaltose	IV	500mg	SOS
3.	Inj. EMROK	Levonadifloxacin	IV	800mg	twice a day
4.	Cap. CUDPRO	Streptococcus thermophiles Lactobacillus acidophilus Bifidobacterium longum	P/O	1 cap.	twice a day
5.	Vimpro Powder				SOS
6.	Tab.OMNA CORTIL	Prednisolone	P/O by gargling		Thrice a day
7.	HHFUCTIC CREAM	Mometasone and Fusidic acid	All over the body at ulcers and scalp L/A		Morning - Night
8.	DERMADEW ALOE CREAM	Aloevera gel+Jojoba+Vitamin+Glycerine	L/A		Morning - Night
9.	INJ. DEXONA	Dexamethasone	IV	1ml	Once in a day
10.	KENACORT OROPASTE	Triamcinolone	P/O	300mcg	3 times a day
11.	INJ. FORCAN	Fluconazole	IV	200mg	Once in a day
12.	INJ. AMPITRUST	Sulbactam	IV	1.5g	twice a day
13.	INJ. GRAFEEL	Filgrastim	S/C	300mg	Once in a day

Discussion:

Bullous pemphigoid is a rare, sub-epidermal, chronic, auto immune, inflammatory and a most common blistering skin disease characterized by the formation of diffuse eczematous, pruritic, urticarial-like lesion, with the later appearance of tense bullae on skin in the regions such as upper thighs or armpits and lower abdomen. It can be fatal in the patients who are debilitated.

Bullous pemphigoid is a rare or uncommon disease and its frequency is unknown. It is known to be reported throughout the world. It has no racial predilection or predisposition, and significantly affects both men and women equally. It primarily appears in the elderly individuals, especially in 60s [2]

The exact cause of Bullous pemphigoid is Unclear, but it's thought that in a person with a genetic pre-condition, it can be triggered by various medications such as Furosemide, Captopril, Pencillamine, Non-steroidal anti-inflammatory drugs (NSAIDs) and Antibiotics.

Bullous pemphigoid is a Type II Hypersensitivity reaction, which is an antibody, mediated reaction, in which IgG and IgM antibodies directed against our own body cells and recognizes as antigen.

This chronic, inflammatory blistering skin disease is also characterized by the presence of immunoglobulin G (IgG) which is an auto-antibody that is specific for the hemidesmosomal bullous pemphigoid antigens i.e., BP230 (BPAg1 – Bullous pemphigoid Antigen 1 or Dystonin) and BP180 or Type 17 collagen (BPAg2 – Bullous pemphigoid Antigen 2). BP antigen 2 is the usual pathogenic antibody [3].

Immune cells, called B-cells or B-lymphocytes produces IgG antibodies, which are a "Y" shaped molecules that involves Fab (Fragment antigen binding site) and Fc (Fragment Crystallizable region) regions, in response to any triggering event.

In Bullous pemphigoid, the Fab region of IgG antibodies binds to the proteins that make up the Hemi desmosomes (which are multiple protein complexes that helps in the stable adhesion of basal epithelial cells to the underlying basement membrane) and recognizes as an antigen [4].

In this case, the Fab region of IgG antibody binds to the hemidesmosomal proteins i.e., BPAg1 (Bullous pemphigoid Antigen 1) or Dystonin and BPAg2 (Bullous pemphigoid Antigen 2) or BP180 or Type Collagen 17.

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As the IgG antibody also contains the Fc region i.e., Fragment Crystallizable region, activates the Complement system. In return the C1, the first of Complement proteins, binds to the Fc region of IgG. C1 then engages or stimulates other members of the Complement family i.e., from C2-C9, some of which are activated by being Cleaved or Chopped off by different enzymes [5].

The cleaved fragments C3a, C4a and C5a acts as Chemotactic factors or Chemokines, and attracts certain cells, in this case, the Mast cells. So, in response to that, Mast cells degranulate and releases several inflammatory mediators, particularly:

- Tumour Necrosis Factor - α
- Leukotriene's
- Cytokines

And these attract inflammatory cells called Neutrophils, Macrophages, Eosinophils and T-cells. So as to fulfill their roles, these inflammatory cells releases proteolytic enzymes against the proteins of Hemi desmosomes (BPAg1 and BPAg2) and destroy them. So as a result blisters or bullae (Sub-epidermal bullae) form between the dermis and epidermis [6].

Bullous pemphigoid can be differentiated from other blistering disorders such as pemphigus vulgaris, dermatitis herpetiform, epidermolysis bullosa acquisita etc., by using clinical histologic and immunologic direct immunofluorescence findings.

Treatment

- First line: topical or systemic corticosteroids
- Systemic corticosteroid therapy should be accompanied by steroid sparing agents whenever possible (Am J Clin Dermatol 2017;18:513)
- Steroid sparing agents (mycophenolate mofetil, methotrexate)
- Tetracycline
- IVIG in steroid resistant disease
- Rituximab or IVIG for refractory disease (not resistant)[7]

Bullous pemphigoid can be treated by prescribing three categories of drugs, in which the first category includes anti-inflammatory drugs, such as topical steroids, sulfonamides, and antibiotics that possess anti-inflammatory properties like tetracycline.

Another class includes the drugs that decrease the production of antibodies such as systemic steroids, azathioprine, methotrexate, mycophenolate, cyclosporine, and ritumixab [8].

And last but not least, drugs that increase the elimination of abnormal antibodies like plasmapheresis and intravenous immunoglobulin (IVIG) can be prescribed [9].

Conclusion:

Overall, Bullous pemphigoid is potentially a fatal disease and can be treatable under the appropriate level of care. The major aim of this case report is to highlight the several treatment options, to bring awareness about the disease and to advocate referral to a dermatologist given its potential severity [10].

Ethical committee approval: Obtained approval from the Manipal hospital ethical committee. We also obtained informed consent from the patient which we have attached with this case report for the journal's reference.

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