International Journal of Nursing and Health Research
www.nursingjournal.in
Online ISSN: 2664-6420, Print ISSN: 2664-6412
Received: 03-10-2021, Accepted: 16-10-2021, Published: 29-10-2021
Volume 3, Issue 1, 2021, Page No. 30-33

Case report on bullous pemphigoid-A challenge with co-morbidities

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Abstract
Bullous pemphigoid (BP) is a rare chronic autoimmune disease of the skin which typically affects people over the age of 60. It might present with blisters seen on arms, legs, groin, mouth or abdomen less commonly involve the mucous membranes including the eyes, oral mucosa, esophagus and genital mucosa. In overall pemphigoid prevalence was 0.012% per 100000 adults and more than 60 years was 0.038% per 100000. In this paper, a 63-year old Female is presented with the complaints of exacerbation of fluid filled lesions all over body since 5 days. The main diagnosis was Bullous pemphigoid with hypertension and Hypothyroidism. Patient was on Corticosteroids and appropriate wound care, diabetes and hypothyroidism was managed with oral drugs. Multiple tense bullae over an erythematous base, few in group and few in discrete pattern over bilateral forearm, neck, face, trunk, proximal part of lower limbs. All blisters were varying sizes ranging between 1-5 cm in diameter, It was successfully managed with appropriate treatment. BP is a life-threatening, especially for older peoples who are already in poor health.

Keywords: bullous pemphigoid, vesicles, bullae, blisters, hypertension and hypothyroidism

Introduction

The aim of this study is to report a case of an elderly woman who presented in the hospital with complains of exacerbation of fluid filled lesions all over body since 5 days. Bullous pemphigoid blisters are less commonly involve the mucous membranes including the eyes, oral mucosa, esophagus and genital mucosa. Pemphigoid is caused by a malfunction of the immune system and results in skin rashes and blistering on the legs, arms, and abdomen.It is a chronic mucocutaneous disease manifesting from the skin and spreading to the other mucous membranes. It is life threatening immunopathologicand dermatologic disease occurring in people at the age of 60-80 years. It is a type II hypersensitivity reaction characterised by the formation of anti-hemidesmosome antibodies.

Case Report

Mrs. V.V is a 63-year old lady, housewife. She is a known case of bullous pemphigoid, biopsy proven on 16.07.2020 since seven months. Now complaints of exacerbation of fluid filled lesions all over body since 5 days. Past history of the patient which initially started with itching over the bilateral forearm, reddish raised lesions, it progressed to involve bilateral thigh, trunk, face, scalp and buttocks are over duration of 5 days. The fluid filled lesions are spontaneously ruptured, which showed that healed within 3 days with hyper pigmentation. There is no history of oral lesions. She is a known case of systemic hypertension since 4 years on T. Amlodipine 5 mg OD, and known case of hypothyroidism since 2 years on T. Thyroxine 25 mcg OD on regular treatment. She has taken C. Doxycycline 100 mg BD for 2 months, T. Dapsone 100mg OD for 3 months, T. Prednisolone 40 mg OD for 22 days, on November 2020 was tapered to 30mg for 1month and the month of December tapered to 20mg for 2 weeks. She wasa history of stoppage T. Predniosolone since 10 days. After stoppage of Steroid Drugs she had developed fluid filled lesions all over body.

There was no associated history of neurological, chest, urinary symptoms, ear pain or discharge and history of similar complaints in the family.

She was admitted on 01.02.2021 with the complaints of exacerbation of fluid filled lesions all over body since 5 days.On general examination the patient was conscious and oriented. The patient's vital parameters were pulse rate was 92 beats per minute, blood pressure was 140/80 mmHg and afebrile.

On Cutaneous examinations she had bullae present on various folds of her body. Multiple tense bullae over an erythematous base, few in groups and few in discrete pattern over bilateral forearm, neck, face, trunk, proximal part of lower limbs. Multiple urticarial plaques seen over the lower back and buttocks region, Few erosions seen over posterior part of thigh and axillary area. Hyperpigmentedmacules seen over the bilateral upper and lower limb, single erosion seen over left groin fold and few vesicles present in mons pubis.All blisters were varying sizes ranging between 1-5 cm in diameter.

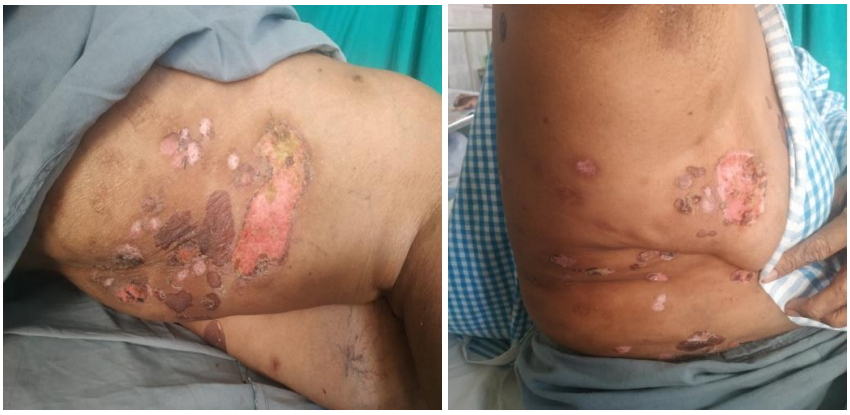

Fig 1

Initial investigations done were a Complete blood count showed TWBC 15600 cells/cumm, blood glucose test, liver function test, renal function test and urine routine are normal. On 04.02.2021 Pap smear sent to the lab, it showed moderate inflammation present and no evidence of infection and abnormal cells.

Discussion

This is a case of Bullous pemphigoid (BP) is a rare, autoimmune, chronic skin disorder characterized by blistering, urticarial lesions (hives) and itching. Less commonly blisters are involved the mucous membranes including the eyes, oral mucosa, esophagus and genital mucosa. Pemphigoid is caused by an immune system deficiency which manifests as skin rashes and blistering on the thighs, arms and abdomen.. In overall pemphigoid prevalence was 0.012% per 100000 adults and more than 60 years was 0.038% per 100000. The management of this patient was Tab. Pradnisolone 40 mg BD, Tab. Symbiotik 500 mg TDS, Fucidin Cream L/A BD, Clobetasolproprionate Oinment HS over the lesion, Tab. Shelcal (Calcium Carbonate) 500mg BD, Tab. FST (Ferrous Sulphate) OD, Tab. Amlodipine 5 mg OD, Tab. Thyroxine 25 mcg OD. After four days the bullae were found to decrease in size and itching reduced much far better.

Through physical examination and appearance of bullae, it was clearly predicted that the patient had Bullous Pemphigoid. Multiple drug therapy was given in order to resolve the symptoms as there was no known etiological cause of the condition along with various medications given to the patient. Having a healthy diet plan to built up the immunity strong along with Calcium, Vitamin supplements, antibacterial, antifungal agents would help overcome the symptoms and condition. Other care patient received such as barrier nursing, wound care, rehydration and treatment of infections.

Definition

Bullous pemphigoid is a chronic, inflammatory, autoimmune pruritic skin blistering disease preferentially in older people, that may involve the formation of blisters (bullae) in the space between the epidermal and dermal skin layers. The disorder is a type of pemphigoid.

Epidemiology

Very rarely seen in children, bullous and non-bullous pemphigoidmost commonly occurs in people 60 years of age and older.

Causes

- A malfunction of immune system
- **Medications:** Etanercept (Enbrel), sulfasalazine (Azulfidine), furosemide (Lasix), and penicillin are examples of prescription medications that can induce bullous pemphigoid.
- **Light and radiation:** Ultraviolet light treatment for some skin disorders, as well as radiation therapy for cancer, may cause bullous pemphigoid.
- **Medical conditions:** Psoriasis, lichen planus, diabetes, rheumatoid arthritis, ulcerative colitis, and multiple sclerosis are all conditions that can cause bullous pemphigoid.

Pathophysiology

Due to the etiological factors Development of auto antibodies against the basement membrane antigens (i.e. BPAG1 and BPAG2) that leads to antibodies activate the complement system and recruit inflammatory cells leading to autoimmune destruction of the basement membrane that leads to weakened adhesion between epidermis and dermis that leads to accumulation of extracellular serous fluids in a pseudo pocket between the epidermisand the dermis that lead to development of wide spreads serum filled bullae in all regions of the body.

Clinical Manifestations

- The first lesions may manifest as a hives-like red raised rash.
- Itchy skin that lasts for weeks or months before blisters appear
- Blisters don't easily rupture when increase in size of the blister and while touching also, often along creases or folds in the skin.
- Minor blisters or sores on the lips, tongue, or other mucous membranes (benign mucous membrane pemphigoid)

Investigations

- A biopsy reveals that the vesicles are intraepidermal, with rounded keratinocytes circulating freely within the blister cavity (acantholysis).
- Direct immunofluorescence of the adjacent natural skin reveals the IgG and C3 epidermal intercellular deposits.
- Diagnostic diagnose can be confirmed with indirect immunofluorescence or tests (ELISA).

Management and Treatment of Bullous Pemphigoid

- Bullous pemphigoid is most commonly treated with corticosteroids drugs such as prednisone, clobetasol, and halobetasol these can be taken by mouth or topically applied creams.
- Antibiotics or other anti-inflammatory medications may also be useful for mild cases such as tetracycline or erythromycin.
- Severe cases may require the use of immunosuppressant medications such as methotrexate, azathioprine or mycophenolatemofetil.
- The objective of steroid therapy is to reduce blister formation, encourage blistering and corrosion healing and to assess the minimum dosage of medication required for disease prevention.Oral corticosteroids can have serious side effects, so it is important to closely follow your doctor's instructions.

The prognosis of bullous pemphigoid

Bullous pemphigoidmay be self-resolving in a period of within 5 years without any treatment, and generally responds well to treatment. When the blisters are ruptured that become infected, can lead to a life-threatening condition called sepsis As a result, it is important to get care as soon as you notice signs. Patient with Poor general health due to old age is associated with a poor prognosis.

Nursing Management

1. Wound Care

- Skin treatments such as dressings and wound care, which will be needed if patient, have areas of raw skin. The four main goals of wound care are
- Prevention of infection,
- Maintenance of a moist environment,
- Protection of the wound, and
- Minimizing scar formation
- Gentle cleansing is an early priority and that can be accomplished by normal saline or antiseptic solution twice a day and apply the ointment to a dressing.
- Cool wet dressings are protective and soothing.
- Before beginning skin care, patients with painful and extensive lesions should be pre-medicated with analgesics.
- After the patient's skin is bathed, it is dried carefully and dusted liberally with non-irritating powder, which enables the patient to move freely in bed.
- Dress in loose fitting cotton clothes. This helps protect your skin.

2. Oral Hygiene

- Oral hygiene is important to keep the oral mucosa clean and allow the epithelium to regenerate.
- Frequent mouth rinsing is recommended to remove debris and soothe ulcerated areas.
- Lip balm is used to keep the lips moisturised.

3. DIET

- All patients to take more amount of Vitamin C rich foods, minerals and water
- If the patient have a oral lesions avoid eating Spicy, hard and crunchy foods, such as chips, raw fruits, and vegetables.
- Orange juice, hot tea and coffee, and spicy drinks can irritate your mouth, causing new sores.
- Soft foods which may be easier to swallow
- Avoid sun and heat if you have pemphigus foliaceus

4. Psychological Support

Reducing anxiety of the patient

- The patient is encouraged to express freely anxieties, discomfort, and feelings of hopelessness.
- Making arrangements for a family member or close friend to spend more time with the patient may be beneficial.
- The patient can be helpful in coping with fears, anxiety and stress through referral for clinical advice.
- Advice the patient, Oral corticosteroids can have serious side effects, so it is important to closely follow your doctor's instructions.

Side effects can include

- Weight gain.
- High blood pressure.
- Diabetes mellitus.
- Upset stomach or indigestion.
- Hip pain
- Advice the patient, BP is self-resolving in a period of within 5 years without any treatment.
- Educate the importance of regular follow-up and monitoring.
- Various forms of psychological therapies such as relaxation techniques, meditation and stress management training to the patients with skin complaints

Complications

- Secondary bacterial infection
- Fluid and electrolyte imbalance
- Hypoalbuminemia

Conclusion

Bullous pemphigoid is primarily a disease of older adults and it rarely occurs in children. Women are affected twice as frequently than in men. It is life threatening immunopathologic and dermatologic disease occurring in people at the age of 60-80 years. It can be resolving in a period of within 5 years. It was successfully managed with appropriate steroids therapy, proper wound care, Skin care (Avoid sun exposure, apply a skin creams and ointments), Good Oral Hygiene and proper diet and Psychological therapy (Keep stress levels are low, pain-relieving muscle Massage and keep the patient family members along with the patient) are important components of the treatment plan to the patients.

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