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Case Study

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A RARE CASE OF BULLOUS PEMPHIGOID WITH SUBCORNEAL PUSTULAR DERMATOSIS AND SECONDARY CELLULITIS: A UNIQUE TRIAD

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ABSTRACT

A 68-year-old female with a long-standing history of Bullous Pemphigoid (BP) presented with widespread, painful pustular lesions and right lower limb swelling. Histopathology and immunofluorescence confirmed BP and Subcorneal Pustular Dermatosis (SPD) while clinical and USG Doppler findings indicated secondary cellulitis. The patient had hypersensitivity to Dapsone and Amoxyclav limiting therapeutic options. She was managed with systemic corticosteroids (Prednisolone), immunosuppressants (Cyclosporine, Cyclophosphamide) and broad-spectrum antibiotics (Ceftriaxone, later Meropenem) along with supportive care. Gradual clinical improvement was observed with resolution of lesions and post-inflammatory hyperpigmentation. This case represents a rare triad of BP, SPD and Secondary Cellulitis posing significant diagnostic and therapeutic challenges. It underscores the importance of early diagnosis, multidisciplinary care, and a carefully balanced treatment approach in managing complex autoimmune and infectious dermatological conditions.

KEYWORD:- Bullous Pemphigoid (BP), Subcorneal Pustular Dermatosis (SPD), Secondary Cellulitis, Immunosuppressants, Broad-Spectrum Antibiotics, Histopathology and Immunofluorescence.

INTRODUCTION

Bullous Pemphigoid (BP) is an autoimmune blistering disorder primarily affecting older individuals, associated with significant morbidity and mortality. Timely diagnosis is crucial as BP is a chronic, relapsing condition that presents histologically with spongiosis, eosinophils, neutrophils, and superficial dermal inflammation. Direct immunofluorescence reveals linear IgG and/or C3 staining along the basement membrane.[1-2] First-line treatment typically involves topical and/or systemic glucocorticoids, steroid-sparing agents, and anti-inflammatory antibiotics.[3] Subcorneal Pustular Dermatosis (SPD), or Sneddon-Wilkinson disease is a rare, chronic, sterile pustular eruption associated with systemic diseases, including immunoglobulinopathies, neoplasms and autoimmune disorders.[4] SPD is characterized by neutrophil accumulation in the subcorneal epidermis and is typically treated with Dapsone.[5] It is marked by the formation of sterile pustules, particularly on flexural areas and the trunk, with the hallmark "Hypopyon Sign" presenting as pustules with a pus-filled lower half.[6] Cellulitis, a bacterial infection often caused by *Staphylococcus aureus* or *Streptococcus pyogenes*, can complicate BP when blisters rupture, offering an entry point for bacteria.[7] The rare coexistence of BP, SPD and cellulitis poses significant diagnostic and therapeutic challenges, requiring a careful balance of immunosuppressive and antibiotic therapies.

CASE PRESENTATION

A 68 year old female patient admitted in the SKIN and STD department, female dermatology ward in Karnataka Medical College and Research Institute (KMCRI), Hubballi on 25th February 2025.

Chief complaints

Painful pus-filled lesions on the scalp, neck, both upper limbs, lower limbs, and trunk for the past 3 days. Itchy, fluid-filled lesions present over both upper limbs (UL) and trunk for 8 years. Exacerbation (Worsening) of lesions for 1 week. Swelling of the right lower limb (RLL) for the past 3 days, associated with pain. On examination the findings confirm the presence of lesions.

History of presenting illness

The patient was previously admitted for the same condition with, k/c/o Bullous Pemphigoid (BP) and Subcorneal Pustular Dermatosis (SPD). The patient was previously admitted for BP with Dapsone hypersensitivity. She received DCP pulse therapy (Dexamethasone-Cyclophosphamide Pulse) Phase I, Cycle 8 on 17/06/22. Her diagnosis of BP was confirmed by biopsy on 03/09/21, and SPD was confirmed on 10/06/24. The patient has been on T. Cyclophosphamide 50mg OD for 2.5 years. She developed hypersensitivity to Dapsone after 4 days, leading to her hospitalization on 13/03/24, and the medication was discontinued. Additionally, she had a severe cutaneous drug reaction to T. Amoxyclav in July 2022, diagnosed as SPD, which required further DCP therapy (Phase I, Cycle 3). The patient's condition began 8 years ago with itchy, fluid-filled lesions on her trunk, which started small and gradually enlarged. Over the past month, the lesions progressively increased in size and number, now involving the upper and lower limbs and trunk. She also exhibited drug hypersensitivity to Amoxyclav and Dapsone.

Head-to-toe examination

Affected Part	Cause
Face	Dryness, Erythema and Exfoliation.
Neck	Lesion present, Rashes.
Oral Cavity	Poor oral hygiene but no lesions.
Palms & Soles	Slight lesions present.
Trunk	Slight pustules with lesions.
Upper and Lower Limbs	Erythema, Edema and Desquamation (Skin peeling)

o/c/e: Recently developed multiple discrete and coalescing pus-filled vesicles on the neck, upper limbs (UL), lower limbs (LL), and trunk. There were multiple lesions with yellow crusting over the bilateral upper and lower limbs, which suggests an infection or secondary bacterial involvement.

The patient appetite was normal, sleep was decreased, diet was mixed, bowel and bladder regular and not addictive to smoke and alcohol.

Vital parameters

Temperature	98^{o}F (normal)
Pulse Rate (PR)	100 beats per minute (Slightly elevated)
Respiratory Rate (RR)	18 breaths per minute (normal)
Blood Pressure (BP)	130/80 mmHg (prehypertension range)
Pain Score	Not recorded
SpO2 (Oxygen Saturation)	99% (normal)
GRBS (Glucose Levels)	110 mg/dl (normal)

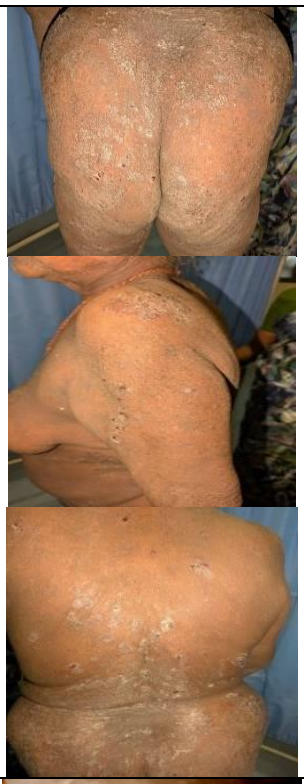
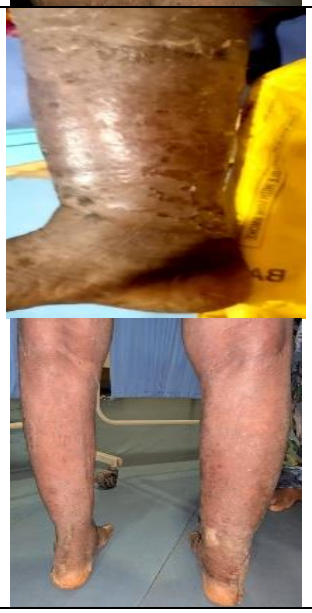
Systemic examination

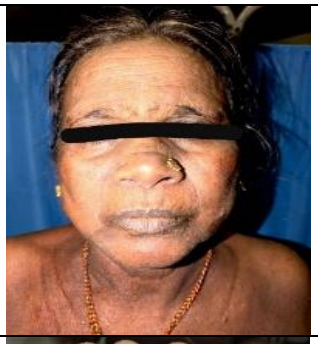
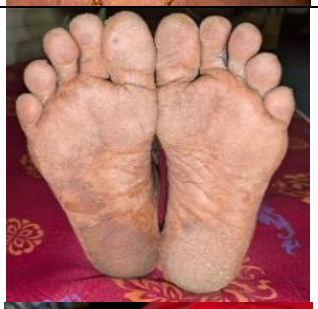
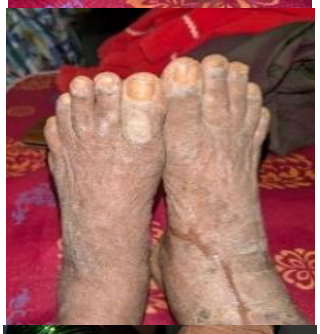
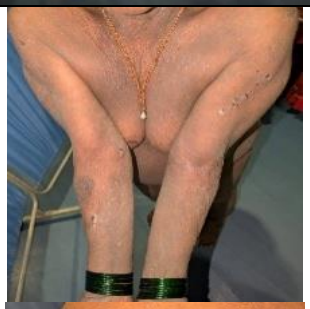
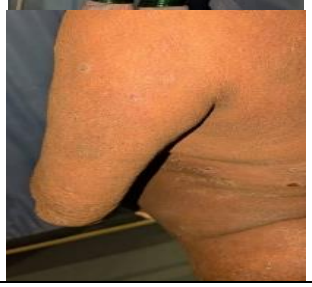
Cardiovascular System	Normal heart sounds (S1, S2) with no murmurs.
Respiratory system	Normal vesicular breath sounds with no added sounds, indicating no lung abnormalities like wheezing or crackles.
Central Nervous System	Higher Mental Functions (HMF) intact. Conscious orientated.
Per Abdomen	Soft, non-tender and no organomegaly (no enlargement of liver or spleen).

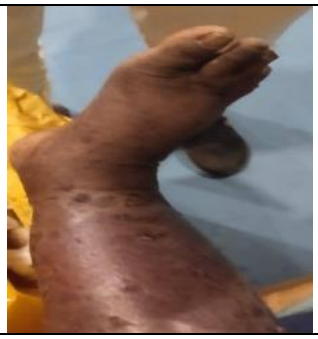
Hematology report

Items	Observed Values	Normal Values	Alarm
WBC	8.2	4.5 to 11.0 × 10^{9}/L	Normal
LYM%	11.5	20 - 40%	Decreased
GRAM%	80.2	50 - 70%	Increased
HGB	9.5	11 - 16g/dl	Decreased
HCT	31.6	36 - 48%	Decreased
MCV	102.4	80- 100 fL	Decreased
RBC	3.09	3.5 – 5.5 10^{6}/uL	Decreased
RDW-SD	47.0	39 – 46 fL	Increased
Sodium	133.2	136 – 149 mmol/L	Decreased
Potassium	3.29	3.5 – 5.3 mmol/L	Decreased
Albumin	2.77	3.2 – 5.5 g/dl	Decreased

The patient was finally diagnosed with Bullous Pemphigoid, Subcorneal Pustular Dermatosis and Secondary Cellulitis. Investigations included CBC, RBS, LFT, RFT, urine analysis, immunofluorescence and blood culture to assess infection, organ function and blood sugar levels. USG Doppler confirmed Cellulitis in the leg, while CBC revealed anemia due to nutritional deficiency. HbA1c test showed increased in percentage under immunoturbidometric method. The biopsy report suggests subcorneal pustular dermatosis**,** a skin condition with neutrophil-filled blisters below the outer skin layer, showing inflammation, acantholysis, and dermal capillary dilation. Treatment included systemic corticosteroids (Prednisolone), immunosuppressants (Cyclosporine) and broad-spectrum antibiotics (Ceftriaxone, later Meropenem). Surgery consultation and MgSO4-glycerine dressing were applied for cellulitis. The patient was also treated for B12 deficiency anemia with Vitamin B12 injections (Vitiofol) and managed for diabetes with insulin, later transitioned to DHA. The patient received care from 25th February in the female dermatology ward and discharged on 8th March, 2025.

Day 1		• Multiple well- to ill-defined hyperpigmented plaques with erythematous erosions. • The lesions are coated with yellowish-brown crust and multiple discrete pustules. • Diffuse erythema, edema, and desquamation (skin peeling) over the face, trunk, and limbs. • Presence of bullae (fluid-filled blisters) on the right lower limb. • Diffuse edema and discrete bullae present over the right lower limb (R/L).
Day 4		• Hyperpigmented plaques with erythematous erosions. • Covered with yellowish-brown crusting. • Discrete pustules and diffuse dryness on face, upper limbs, and trunk. • Reduction of multiple well-ill-defined post-inflammatory hyperpigmentation (indicating healing).

		
Day 7	  	• Multiple well- to ill-defined hyperpigmented plaques. • Covered with erythematous erosions and yellowish-brown crusting. • Discrete pustules with diffuse dryness and exfoliation over face, both upper limbs (BL UL), and trunk. • Post-inflammatory hyperpigmentation noted (indicating healing).
Day10	 	• Multiple well-defined hyperpigmented plaques. • Erythematous erosions covered with yellowish-brown crusting. • Diffuse dryness and exfoliation. • Involvement of the face, both upper and lower limbs (BL UL, LL), and trunk. • Multiple well-defined post-inflammatory hyperpigmentations (suggests healing progress).



Treatment

Si. No.	Medication	Frequency	Route of administration
1	Injection Ceftriaxone (Xone)	1-0-1	IV
2	Tablet Chymoral Forte (Trypsin + Chymotrypsin)	1-1-1	PO
3	Glycine + Magnesium Sulfate Dressing	-	-
4	Injection Pantoprazole	1-0-0	IV
5	Tablet Paracetamol	1-0-1	PO
6	Tablet Ferrous Sulphate / B-Complex / Calcium Citrate (FS/BC/CaZt)	0-1-0	PO
7	High Protein Diet	-	-
8	Tablet Chlorpheniramine Maleate (CPM)	0-0-1	PO
9	Tablet Cefuroxime (CZR)	1-0-0	PO
10	Liquid Paraffin	1-0-0	PO
11	Tablet Cyclophosphamide	1-0-0	PO
12	6th Hourly Glucose Random Blood Sugar (GRBS) Monitoring	-	-
13	Injection Gentamycin (GA)	1-0-0	IV
14	Tablet Prednisolone	1-0-1	PO
15	Injection Vitcofol (Vitamin B12, Folic Acid, Iron)	1-0-0	IV
16	Injection Insulin (Actrapid or PSS)	1-1-1	SC
17	DASH Diet (Dietary Approaches to Stop Hypertension)	-	-
18	Injection Meropenem	1-1-1	IV
19	Tablet Cyclosporine	1-0-1	PO

DISCUSSION

Bullous Pemphigoid (BP) is a non-scarring blistering disease, often presenting with flexural skin lesions, though it can be localized or generalized. Mucous membranes, particularly the oral mucosa, are involved in about 50% of cases. BP is typically self-limiting, remitting within 5 years, with mortality rates ranging between 6% and 41% prior to the use of corticosteroids, as reported by Lever in 1953, and more recently in studies.[8-10] Diagnosis is established clinically, histologically, and immunopathologically using immunofluorescence (IF) testing, though prolonged treatment can reduce IF positivity.[11] Subcorneal Pustular Dermatosis (SPD) is characterized by neutrophil infiltration in the subcorneal epidermis, typically affecting intertriginous and flexural areas, though, in our case, it presented with severe lesions across the body. Cellulitis most often affects the lower extremities, presenting as acute, erythematous, and swollen areas, sometimes with blisters, ulcers, and lymphatic involvement. The area of cellulitis should be reviewed daily for progression or regression to assess antibiotic efficacy.[12] Anti-inflammatory agents can help resolve the infection by reducing immune mediator production.[13-14] This case underscores the rare coexistence of BP and SPD, further complicated by secondary cellulitis, requiring an individualized therapeutic approach.

This case highlights the rare coexistence of Bullous Pemphigoid and Subcorneal Pustular Dermatosis, a combination that has been scarcely reported in literature. The additional complication of secondary cellulitis further complicated management, necessitating an individualized therapeutic approach.

CONCLUSION

This case presents a rare and complex triad of Bullous Pemphigoid (BP), Subcorneal Pustular Dermatosis (SPD) and secondary cellulitis highlighting significant diagnostic and therapeutic challenges. The 68-year-old female, with a history of recurrent BP and hypersensitivity to Dapsone, was diagnosed with BP and SPD through histopathology and immunofluorescence, while cellulitis was confirmed clinically and via USG Doppler. Treatment involved a cautious regimen of systemic corticosteroids (Prednisolone), immunosuppressant (Cyclosporine, Cyclophosphamide)

and broad-spectrum antibiotics (Ceftriaxone, Meropenem), along with supportive care. The patient showed gradual improvement with lesion resolution and post-inflammatory healing. This case emphasizes the importance of a multidisciplinary approach, early diagnosis, histopathological confirmation and individualized therapy in managing such complex, overlapping autoimmune and infectious dermatological disorders. Continued research should focus on steroid-sparing strategies and long-term management to optimize patient outcomes in rare, challenging cases like this.

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