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Clinical pharmacist intervention in exacerbated Pemphigus vulgaris

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ABSTRACT

Patient had a history of Pemphigus Vulgaris for past 3 years and was now admitted due to the sudden extensive flare up. The subject complained painful erosions covering more than 40% of the body surface area. Lesions started orally and were short lived and formed peripheral extensive erosions. Periorbital and Perioral lesions were present . Patient was immediately initiated on Corticosteroids and antibiotics. Protein supplementation, Condy's Compression along with other local applicants were prescribed. A partial pulse with Dexamethasone was given for two days. The overwhelming number of medications that created confusions of administration for the care taker were resolved through elaborate patient counseling. Apart from adverse drug reactions monitoring which forms a part of clinical pharmacist responsibilities, this case shows the inevitability of clinical pharmacist in educating patients and their care takers in both medication management and need for the compliance.

INTRODUCTION

Pemphigus is a group of chronic, potentially fatal diseases that are autoimmune and bullous in nature. It is usually classified into six, based on their characteristics. Pemphigus vulgaris with its many clinical variants is a mucocutaneous disease where initial blisters are mostly oral in origin[1]. IgG antibodies against the Desmoglein 3 in the circulating system is diagnosing factor of the most common variant[2]. Here the blister formation is seen in the intraepithelial layer due to the destruction of tight junctions of the epithelial cells by the auto antibodies and thus resulting in the acantholysis[3,4].

Pemphigus Vulgaris is a vulnerable disease with potential to aggravate on slight ignorance to the regular medication and changes in dietary and environmental factors[5]. As such apart from the genetic predisposition, environmental changes including viral infections, contact allergens and numerous others and the endogenous variations triggering the disease makes it imperative to provide immense education regarding the management as well as the provoking agents to the patient and their care takers[6].The emergence of systemic corticosteroids in the treatment of pemphigus vulgaris in early 1950s brought a drastic reduction in fatality of the disease[7,8]. Along with the systemic steroids, use of oral corticosteroids and nonsteroidal agents like Mycophenolate mofetil, dapsone, cyclophosphamide, azathioprine, cyclosporine and even rituximab in certain cases is used today for the management of pemphigus vulgaris[4,9]. The prime objective of therapeutic management of this disease is to heal the mucocutaneous lesions and bullae and rectify any functional impairment induced by the disease. The prevention of a relapse and management of adverse reactions caused by the long term use of immunosuppressive therapy and corticosteroids forms the secondary objective of the management[10].

CASE REPORT

A 52 year old female with known case of pemphigus vulgaris of three years was admitted in the dermatology department of the Government medical college hospital with the complaints of painful erosions all over the body . She had no history of diabetes mellitus or hypertension and had no other comorbidities. Patient was on combined therapy of oral corticosteroids and azathioprine when 3 weeks back she stopped all the medications and started ayurvedic treatment which caused a flare up. Lesions initially started as painful erosions over the oral canal and then as fluid filled lesions on upper trunk. All lesions where short lived (1-2 days) to form erosions with peripheral extension . On general examination ,raw , oozy lesions over neck, trunk , abdomen , thigh , periorbital region were found along with bilateral pitting pedal edema. Nikolsky's sign was positive . Patient's vital signs remained stable with difficulty in obtaining the blood pressure due to pervasiveness of the painful erosions.

Extensive Cutaneous examination showed multiple raw lesions over neck, bilateral mammary area, upper limb , axilla, abdomen , groin , thighs, popliteal fossa , few of them having hyper pigmentation and crusting. Multiple raw erosions with hemorrhagic crusting were seen in the periorbital and perioral region involving lips. Hyper pigmented macules over thighs and ventral portion of the trunk and well defined ulcers on the lower side of the right breast and upper side of abdomen were visible. In the oral cavity erosions over soft palate and dorsal aspect of tongue were seen. Raw erosions over labia minora, crusted erosions and fungal infection on the scalp were also noted. Perilesional biopsy showed Keratinocytes arranged in "tomb stone" pattern at the base of the blisters. Dermis showing moderate perivascular lymphocytes in the follicle. Pus culture sent on the second day of admission showed the abundance of Acinetobacter and Pseudomonas species . (Table 1)

DISCUSSION

Patient was immediately started on 8 mg of DEXAMETHASONE injection twice daily which was continued till the second day. On the third day onwards the duration was increased to three times a day. It was continued for three days during which LINEZOLID 600mg was given orally every 12 hourly to heal the secondary infections and ulcerations. GENTIAN VIOLET PAINT was stopped after three days due to the complaints of extreme dryness of the skin. After the first seven days of LINEZOLID treatment , PIPERACILLIN(4000mg) along with beta lactamase inhibitor TAZOBACTAM(500mg) were initiated . DOXYCYCLINE 100 mg was added to suppress the inflammation. On the sixth day of the treatment DEXAMETHASONE was given as a partial pulse, that is 100mg in 500ML of 5% DEXTROSE over 2 hours for two days. The short term high dose therapy being highly efficacious and purported to cause less adverse drug reactions.

A rising trend of the fasting blood glucose was detected after the partial pulse therapy and short acting INSULIN (HUMAN ACTRAPID) of 6-6-8 units was recommended for immediate initiation. Persisting elevated blood glucose level indicates

Table 1 : Laboratory investigations

Investigations	Day 1	Day3	Day5	Day7	Day 10	Day14	Normal Range
Total WBC	17.8	15.6	15	13.5	7.1	12.9	4.3-10.3x10^3/µL
Neutrophils	95	89.1	90.8	92.5	81.2	87.6	41.4-73.0%
Lymphocytes	3.0	5.9	4.1	2.4	12.6	9.7	19.4-44.9%
RBC	3.4	3.68	3.53	3.36	3.54	3.37	4.38-5.7x10^3/µL
HB	10.3	10.0	9.7	9.3	9.6	9.4	12.5-15.5g/dL
Platelet	369	331	323	291	285	287	156-373x10^1/µL
ESR	100	100	95	60	66	90	0-25mm/hr
S.Total Protein	5.2	5.1	5.4	5.6	6.0	5.9	5.5-8.5 gm%
S.Albumin	2.1	2.1	2.6	2.7	3.0	3.0	3.0-5.0 gm%
SGOT	16	18	22	21	23	17	5-38 IU/L
SGPT	32	36	40	53	69	78	5-42 IU/L
S.ALP	135	141	157	159	165	171	20-112 IU/L
FBS	124	148	163	194	185	162	70-110mg/DL
C-reactive protein	111					34	0-10 ng/L

Table 2 : Drug Chart

	DRUGS	Day 1-2	Day 3-5	Day6-7	Day8-9	Day 10-14
1.	Inj.Paracetamol IV Q8H	X	X	X	X	X
2.	Inj.Albumin 100mL IV OD		X			X
3.	Inj. Human Actrapid (short acting insulin) 6-6-8U s/c			X	X	X
4.	Inj.Dexamethasone 8mg IV BD	X			X	X
5.	Inj.Dexamethasone 8mg IV TID		X			
6.	Inj.Dexamethasone 100mg in 500mL 5% Dextrose over 2 hrs			X		
7.	Inj.Piperacillin Tazobactam 4.5g IV Q8H (ATD)				X	X
8.	T.Linezolid 600mg Q12H	X	X	X		
9.	T.Calcium and Vit D3 500mg	X	X	X	X	X
10.	T. Ranitidine 150mg BD	X	X	X	X	X
11.	T.Vitamin Bcomplex 1OD	X	X	X	X	X
12.	T.Clonazepam0.5 mg 1 HS		X	X	X	X
13.	T. Zinc acetate 50mg 1 HS			X	X	X
14.	T. Vitamin C 500 mg 1 OD			X	X	X
15.	T. Mycophenolate Mofetil 500mg TID				X	X
16.	T. Ferrous fumarate 152 mg 1 OD				X	X
17.	C.Doxycycline and Lactic acid Bacillus 100mg				X	X
18.	SYP.Sucralfate 2tspTID	X	X	X	X	X
19.	Protein Powder 2 tsp TID in 1 glass of milk	X			X	X
20.	Cuticell dressing	X	X	X	X	X
21.	Condy's Compress LA BD	X	X	X	X	X
22.	Amlexanox gel 5%LA BD					
23.	Gentian Violet Paint 1%LA BD	X				
24.	Moxifloxacin eye ointment 0.5% LA BD		X	X	X	X
25.	Fusidic acid cream2 % LA BD			X	X	X
26.	Moxifloxacin with Dexamethasone eye drops (Quin D) 1 drop every 3 hourly	X	X	X	X	X

corticosteroid induced hyperglycemia. During the interaction with patient a mild case of muscle cramps owing to the high dose corticosteroid therapy was recorded. The gradually decreasing pattern of the red blood cells, hemoglobin and platelet during LINEZOLID therapy raised the suspicion of the drug induced anemia and thrombocytopenia. FERROUS FUMARATE 152 mg (containing 50mg of elemental iron) was recommended on the eighth day of therapy. (Table 2)

Patient compliance to the prescribed medication is a prerequisite for the remission and prolonged period without a relapse of an autoimmune disease such as Pemphigus Vulgaris. Noncompliance to the medication arise from the multiple drug regimen , varying and confusing frequencies of these multiple drugs, intolerable and nauseating taste of these medications and exorbitant cost of the therapy [11].

Here the patient and her care takers were primarily re-educated on the importance of long term concordance with the medication. The potential of this disease to relapse on minute

triggering factors and the fatality associated with the abandonment of the therapy, owing to the life threatening secondary infections were detailed by the clinical pharmacist. The need for a food calendar as per the recommendations of International Pemphigus and Pemphigoid foundation to easily trace out the triggering agent was specified and a model was illustrated .Importance of high protein diet for the rapid healing and the list of protein rich food items and other specifically useful diet was provided to the patient. The frequency of the long list of medications and the proper method of administration which was often confusing to the care taker was provided in a neatly printed sheet along with the illustration of a drug calendar to assure the daily drug administration .The compounding of POTASSIUM PERMANGANATE crystals for CONDY'S COMPRESSION was detailed to the care takers. Patient and their care takers opted out the long term DCP (DEXAMETHOSONE, CYCLOPHOSPHAMIDE PULSE) therapy and preferred treatment with Immunosuppressive agent MYCOPHENOLATE MOFETIL (500mgTID) .The need for routine monitoring of Complete blood count , hepatic and renal functions while taking this medication was emphasized.

CONCLUSION

The concept of pharmaceutical care which is a responsible provision pertaining the drug therapy for the prime objective of obtaining defined therapeutic outcome that ultimately improve quality of a patient' life includes patient counseling .Apart from the monitoring of adverse drug reactions and potential contraindications and interactions, the inevitability of a clinical pharmacist in patient education especially in the management of chronic diseases with polypharmacy is notable. Unlike in case of an acute ailment , chronic diseases dictate a prolonged hospital admission, self management ,long term drug therapy and various non pharmacological methods for the improvement . Extensive information about the frequency and duration of the multiple drugs makes it confusing for the care taker which often can result in the noncompliance .By being an integral part of the clinical team that cares for the patient , clinical pharmacist can thus assure the patient concordance and patient awareness.

LIMITATIONS

We couldn't follow up the case further and the rarity of a case with this extensive manifestations made it difficult for us to assemble similar cases.

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CONFLICT OF INTEREST

Authors declare no conflict of interest

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