



Case Study

AN INTEGRATED APPROACH IN MANAGEMENT OF PEMPHIGUS VULGARIS

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ABSTRACT

Pemphigus refers to a group of autoantibody-mediated intra-epidermal blistering diseases characterized by loss of cohesion between epidermal cells (a process termed acantholysis). Applying pressure to the skin of these patients may elicit the separation of the epidermis which is known as Nikolsky's sign. Pemphigus vulgaris can be compared with Agnivisarpa based on similar manifestation. The mainstay of treatment is systemic glucocorticoids in allopathy science. The current mortality is around 5% with glucocorticoids treatment. Case **History:** This case report is of 40 years old male patient who presented with blisters formation on his whole body perspiration, itching and burning since the last 6 months. He was treated in indoor department with Snehana Cikitsa mainly along with oral glucocorticoids for 25 days. The dose of oral glucocorticoids was tapered with improvement in patient's condition. Result: At the time of discharge, patient was prescribed 10mg prednisolone and oral Ayurvedic medicines. After 25 days of treatment patient had more than 90% relief and glucorticoids was stopped. On the 2nd follow of treatment patient was kept only on Ayurvedic medicines. During first two follow-ups, patient had significant relief.

INTRODUCTION

Pemphigus Vulgaris is a mucocutaneous blistering disease that mainly found in patients over 40 years of age. Pemphigus Vulgaris typically starts on mucosal surfaces and often involve the skin. Pemphigus Vulgaris is manifested as fragile, flaccid, blisters and it ruptures to produce extensive denudation of mucous membranes and Pemphigus Vulgaris typically involves the mouth, scalp, face, neck, axilla, groin and trunk. Pemphigus Vulgaris is sometimes associated with severe skin pain; some patients also experience pruritus as well. Most of times, lesions heal without scarring. Postinflammatory hyperpigmentation is usually present at sites of healed lesions after few days. Pemphigus Vulgaris can be life threatening. Before availability of glucocorticoids, the mortality ranged from 60-90%.

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The common causes of death are infection and complications of treatment with glucocorticoids. Factors include advanced age, diffused spread, and the requirement of high doses of glucocorticoids (with or without other immunosuppressive agents) for control of disease suggest bad prognosis. The course of disease in individual patients is variable and not easy to predict. Some patients of Pemphigus Vulgaris achieve remission, while others may need long-term treatment or succumb to complications of their disease or its treatment. Pemphigus Vulgaris is usually started on prednisolone, 1mg/kg per day. If new lesions continue to appear after 1-2 weeks of treatment, the dose may need to be increased and/or combined with other immunosuppressive agents such as azathioprine, mycophenolate mofetil or cyclophosphamide. Patients with severe, treatment resistant disease may derive benefit from plasmapheresis, IV immunoglobulin or rituximab. It is urgent to bring severe or progressive disease under control as soon as possible to lessen the severity and/or duration of this disorder[1].

Clinical features of Pemphigus Vulgaris and Agnivisarpa are almost same so Pemphigus Vulgaris can be correlated with Agnivisarpa in Ayurveda. In *Agnivisarpa*, patient has feeling as if his whole body is sprinkled with burnt charcoal. The areas of body which the Agnivisarpa spreads appears like the extinguished charcoal (black) or excessively red. The areas become surrounded with pustules like those caused by burns. Patients of *Agnivisarpa* succumb to death instantaneously if not treated timely. Thus, there is need to treat this life threatening disease with integrative approach.

Patient Information

A 40-year old patient working as farmer came with complaints of blisters formation whole over body, increased perspiration, itching and insomnia since last 6 months. Initially, blisters appeared on chest and within three days whole body was covered with blisters. Patient took medicine from local physician for 10 days and he didn't get any relief. Then patient was referred to dermatologist on 8/12/2022. Patient was advised for hospitalization but patient was reluctant. Patient was prescribed Tab. Glasiprin (Azathioprine)-50mg twice a day, Tab. Winself-16 (methylprednisolone) twice a day. Patient took medicines for 40 days but he didn't get relief and his problem was worsening so he stopped all medicines. After few days at home he came to Ayurveda hospital on 16/02/2023. He was advised hospitalization after complete examination.

Clinical Examination Timeline and Therapeutic Interventions

Patient came with complaints of blisters formation whole over body, perspiration and itching. Blisters were found whole over body; some of them ruptured and produced extensive denudation of skin. Patient's look was gloomy and depressed. Clinical findings regarding other systems were normal. Hemodynamically, patient was stable.

Trividha Pariksha (Three-fold examination)

Darshana: Agnidagdha Sphota (pustules like those caused by burns) whole over body and Raktavrnata (reddish discoloration). Jihva was slight coated and Sveta, Akshi was little red.

Sparshana: Nadi (pulse) was Vatapaitika in dosika character.

Prasna: Purisha frequency twice a day, semisolid in consistency, no mucus, no foul smell, not observed whether floated or sunk in pan. *Mutra* frequency around 8-10 times per day without any associated complaints.

Diagnostic Assessment

Diagnosis of pemphigus vulgaris was made on basis of clinical presentation. Patient was confirmed as a patient of pemphigus vulgaris by dermatologist. The cardinal features of the pemphigus vulgaris i.e., blisters formation whole over body and itching were present in this case.

Sr.no	Drug	Dosage	Adjuvant	Duration
1	Panchatikta Grita ^[2]	1st day- 40ml twice a day- empty stomach	water	
		2 nd day – 55ml twice a day- empty stomach		
		3 rd day – 70ml twice a day- empty stomach		
		4th day – 85ml twice a day- empty stomach		
		5^{th} day – $100ml$ twice a day- empty stomach		
		6 th day – 120ml twice a day- empty stomach		
		7 th day – 40ml twice a day- empty stomach	water	12 days
2	Kaishor Guggalu ^[3]	3 tablets thrice a day – before food	water	12 days
	(1 tab = 300mg)			
3	Maṃjishthadi Kvatha	40ml twice a day before meal.	-	12 days
4	Panchavalkal churna	For dusting	-	18 days

Table 1: First 18 days in IPD

Conventional medication

Sr.no	Medicine and dose	Dose	Duration
1	Prednisolone	40mg - Once a day	6days
		30mg- Once a day	6 days
		20 mg- Once a day	6 days
		10 mg - Once a day	On the day of discharge

Table 2: last 7 days -follow up

Sr.no	Drug	Dosage	Adjuvant	Duration
1	Panchatikta grita	40ml- before food	Water	7 days
2	Kaishor Guggalu (1 tab = 300mg)	3 tablets thrice a day – before food	Water	7 days
3	Maṃjishthadi Kvatha⁴	40 ml twice a day before meal.	-	7 days
4	Panchavalkal churna	For dusting	-	7 days
5	Prednisolone	10mg - Once a day		7days only



1. Before treatment

2. At the time of discharge

3. During first follow-up



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1. Before treatment

2. At the time of discharge

3. During first follow-up

Follow-Up and Outcome

A significant improvement in clinical features was noted after one week of treatment. There were no new blister formation and old lesions disappeared gradually. 10mg oral glucocorticoids was tapered on every sixth day and on 25th day (on first follow-up) it was stopped. Even after stopping glucocorticoids, any new blister formation was not noted and there was marked improvement in old lesions.

DISCUSSION

There are many life threatening ailments which are not cured by allopathic science. Ayurveda has treatment of these ailments but in critical cases saving of patient's life must be considered. Advanced diseased condition of pemphigus vulgaris may cause death instantaneously. Acharya Caraka has also mentioned that if treatment is not started immediately, patient will succumb to death instantaneously. [5] *Agnivisarpa* is caused by Vata-pitta.[6] Medicated ghee is drug of choice in case of vitiation of Vata-pitta.[7] Continuous use of ghee alleviates Pitta, as ghee is Madhura rasa (sweet taste), Sita (coldness) and Mamda; Pitta being Amadhura (opposite to Madhura) in taste Ushna (hot) and Tikshna (sharp) is of the opposite nature. The stronger dominates over the weaker, when interaction between two substances having mutually opposite qualities, therefore use of ghee for long time alleviates Pitta.[8] In this case Panchatikta ghee was given in increasing doses. Medicated ghee having bitter taste is drug of choice in Visarpa cikitsa.[9] It was started with 40ml twice a day on first day. On second day, 55ml twice a day was given. On third day 70ml twice a day, on fourth day 90ml twice a day, on fifth day 90ml twice a day. On fifth day, patient had five times loose motions, so from sixth day patient was kept on 100ml twice a day and continued for next 12 days. For local application Panchavalkal Churna was Panchavalkal Churna having Kashhaya rasa, Kashhaya

rasa absorbs fluid and also do *Sandhana karma* which is necessary in denudation of skin.^[10] *Panchavalkal* was used as local application.

CONCLUSION

This case report suggests important of integrative approach in management of life threatening condition such as Pemphigus vulgaris. Glucocorticoids is lifesaving drug if they are used rationally. Glucocorticoids has also serious side effects if it's used long time irrationally. Glucocorticoids doesn't have any role in breaking the pathogenesis, there is also high chances of recurrence and resistance of therapy if glucocorticoids used alone. In Ayurveda science, there are very good therapies which cures the disease completely and there is very less chances of recurrence. This case report is good testimony of good outcome of combined approach in management of pemphigus vulgaris.

Declaration of Patient Consent

Authors certify that they have obtained patient consent form, where the patient has given his consent for reporting the case along with the images and other clinical information in the journal.

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