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Case Study

PEMPHIGUS FOLIACEOUS WITH SPECIAL REFERENCE TO *VISPHOTA KUSHTA*

K Ambika[1]*, Aswathy Dev S[2], Vishnu P[2]

*[1]Professor and HOD, [2]MD Scholar, Dept. of Kayachikitsa, Govt. Ayurveda College, Thiruvananthapuram, Kerala, India.

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ABSTRACT

Pemphigus is an autoimmune blistering disease which can be fatal if left untreated. In case of secondarily infected skin lesions, life threatening sepsis may develop. There is no specific treatment protocol for the management of pemphigus. The present study is aimed to treat a 57-year-old male patient clinically diagnosed with pemphigus foliaceus admitted in Kayachikitsa department in Govt. Ayurveda College, Thiruvananthapuram. Treatment was aimed at controlling the progression of disease, preventing infection and inducing healing of the affected area. In Ayurveda symptoms are more similar to *Kaphapitha kushta* especially *Visphota kushta*. *Langhana, Snehapana* and repeated *Sodhana* were the treatment protocols adopted. Ayurvedic approach in this single case of pemphigus is beneficial in correcting the inherent *Agnimandya* at *Koshta* and *Dhatu* level and thus provides significant result. PDAI score was used for assessment. Before treatment PDAI was 96, reduced to 24 after *Sodhana* and after treatment it was 6. There was no recurrence of symptoms after 2 weeks.

INTRODUCTION

A large number of diseases which were earlier considered as cutaneous manifestations of autoimmune diseases are now recognized as diseases having its own clinical histologic and immunopathologic findings. Pemphigus is one among them [1]. Pemphigus is an immunologically mediated blistering disease with pain, pruritus disfigurement and in some cases may lead to death. Pemphigus includes various subtypes like pemphigus vulgaris, pemphigus foliaceus, paraneoplastic pemphigus, bullous pemphigoid, dermatitis herpetiformis etc [2]. Pemphigus foliaceus is an autoimmune condition in which immune system releases IgG autoantibodies that causes a characteristic inflammatory reaction. The sub corneal layer of skin is involved where intercellular adhesion of keratinocytes is lost leading to the formation of sub corneal blisters within the epidermis. Skin lesions may remain localised and over the time can coalesce to cover the entire skin [3]. Nikolsky's sign is present classically which shows separation of epidermis on applying manual pressure.

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Loss of cohesion between epidermal cells (acantholysis) is responsible for mucocutaneous blisters [4]. Advanced age, widespread involvement, requirement of high doses of glucocorticoids are bad prognostics for the disease. In modern system of medicine this condition is considered as an emergency situation which needs immediate intervention. Immediate administration of corticosteroids is the treatment of option but these in high doses may cause serious side effects including diabetes, increased risk of infection stomach ulcers etc [5]. PDAI (Pemphigus Disease Area Index) score is used to assess the severity of the disease which ranges from 0 to 263[6].

Considering the etiological factors, clinical features, mode of spread and pathogenesis, different types of skin manifestations described in the classical textbook of Ayurveda can be correlated. Spreading nature of the disease is similar to that of *Visarpa* as explained in *Ashtangahrudaya*[7]. But while considering all the diagnostic factors and *Dosa* predominance pemphigus closely resembles *Visphota kushta* which is characterized by erythematous and blackish eruptions associated with pain[8]. *Dosa* predominance is *Kaphapithadhika tridosha*. A large number of formulations like *Tikthaka ghrita, Mahatikthaka ghrita, Vajraka ghrita* etc. are explained in the classical texts in the context of *Kushta chikitsa*[9]. In the initial stage there is always *Kapha pitha utklesha* and *Amanubandha* manifested as inflammation. Hence

Snehana is totally contraindicated. Considering the *Visarpa* nature of the disease *Snehana* cannot be done initially. *Langhana* therapy in the form of *Rookshana* and *Deepana pachana* is done initially till *Utklesha* is reduced and *Niramavastha* is attained. In this phase *Snehana* can be started followed by proper *Sodhana*. Repeated administration of *Sodhana* therapy and *Rasayana* helps to reduce the chance of recurrence and further progression of the disease.

AIMS AND OBJECTIVES

To evaluate the role of a selected treatment protocol in the management of pemphigus.

Study setting

The present case study was conducted in the Dept. of *Kayachikitsa*, Govt. Ayurveda College, Thiruvananthapuram.

Case Report

Basic information of the patient

Age- 57, Sex- male, Religion – Muslim, Socioeconomic Status – poor, Occupation – works in a juice shop.

Presenting complaints

Erythematous crusted exfoliating skin lesions all over the body since 4 months.

H/o presenting complaints

The patient with a known history of type 2 DM, HTN, DLP was leading a normal life since 4 months ago. Before 4 months he had to go through an extreme stressful situation followed by which he developed mild itching over his face. Within a few days he noticed vesicular lesions over the back of head which on gruttage ruptured with oozing of clear fluid. The lesions were extremely painful associated with low grade fever and he consulted a dermatologist. Some topical applications were advised. He got mild relief from pain and lesions subsided to a small extend. Reddish patches started to appear over face, around eyes, neck, upper limb and lower limb associated with itching and low-grade fever. The patient consulted GAVC, TVM. For consultation and underwent op management. Within a few days fluid filled bullae developed which after getting ruptured with purulent oozing turned skin over affected area into crusting erosions during the course of healing. The ruptured lesions turned scaly, erythematous and crusted within a few days. Hyperpigmentation of the skin was also noticed. Thus, the patient was admitted here for further management.

H/o past illness

H/o Type 2 DM since 3 years (under irregular medication)

H/o HTN since 20 years (under irregular medication)

H/o DLP since 3 years (under irregular medication)

H/o CAD –3 yrs

H/o hyperuricemia (since 2013)

H/o chickenpox (2 episodes – childhood and 4 years before)

H/o recurrent fungal infection (upper limb and groin region)

H/o onychomycosis

H/o food poisoning – 4 months

Drug history

T. Metformin 600 mg 1-0-1

T. Glimepiride 1 mg 1-0-1

T. Atorvastatin 10 mg 1-0-1

T. Ecospirin 0-1-0

T. Clinidipine 10mg 1-0-1

Family history

Father had history of having psoriatic lesions (not diagnosed)

Personal history

Bowel – Irregular, loose in consistency, Appetite- adequate, Micturition – 3-4 times per day, urgency present, increased frequency, Sleep – disturbed due to skin lesions, Addiction – Alcohol – 240 ml per day

Occupational history

Worked in a juice shop for 30 years

Psychosocial history

The patient is leading a stressful life for the last 4 months. Financial stress- present. In a good relationship with family members.

Socioeconomic history

Lower middle-class family

On Examination

General condition was normal, afebrile, conscious and oriented, respiratory system, gastrointestinal system and musculoskeletal system showed no abnormalities. In cardiovascular system, the patient felt palpitation occasionally. HR– 84/min, PR- 84/min, BP- 116/76mmhg, RR- 16/ min.

Integumentary System Examination

Site of onset- scalp, mode of spread – centripetal, colour- erythematous, primary lesions – vesicle, secondary lesion – crust, configuration – coalesced, margination – ill defined, distribution – asymmetrical, genitalia – not involved, mucus membrane – not involved, nail changes- onychomycosis, Nikolsky's Sign - positive

Ashtasthana pareeksha

Nadi – Vata-Pitha, Mutram– Anavilam, Malam– Gradhitam, Jihwa – Upaliptham, Sabda- normal*, Sparsha – Rooksha khara, Drik – Vyaktham, Akruti –* normal

Diagnosis

Clinical diagnosis of pemphigus foliaceus had been made after history taking and systemic examination. Nikolsky's sign was also positive. In Ayurveda,

symptoms are found to be more similar to that of *Visphota kushta*

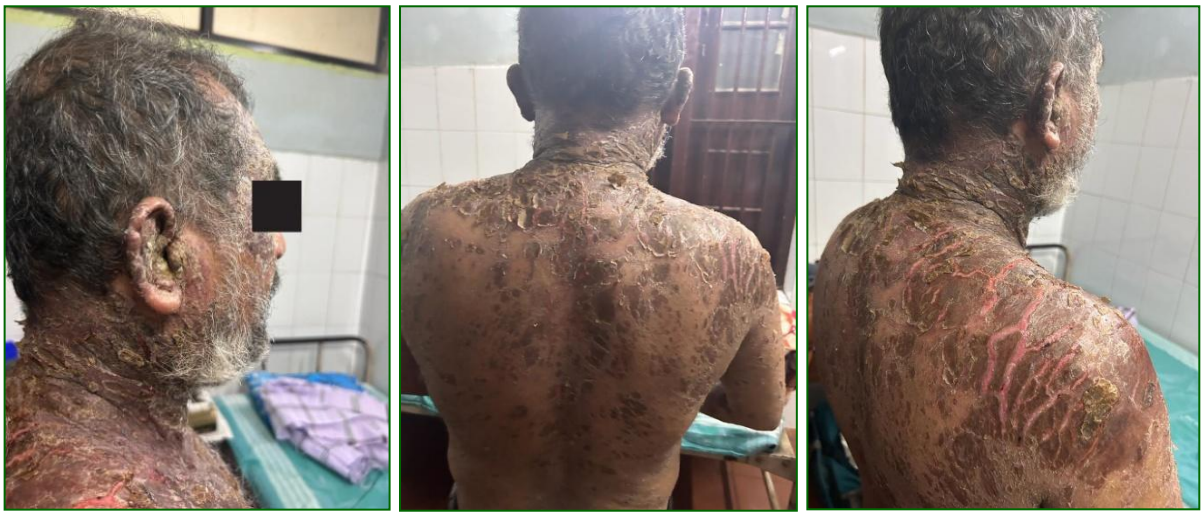
Treatment protocol and Assessment

		Internally	**Externally**	**PDAI Score**
stage 1	*Deepana pachana*	*Chiruvilwadi kasayam*– 90ml bd b/f *Aragwadharishtam* - 30ml bd a/f *T. Sudarshanam 2-0-2* *T shaddharanam 1-0-1*	*Dhara – Veppinpatta and Konnatholi Kashaya* prepared with leaves of *Azardirachta indica and Cassia fistula*	**96**
Stage 2	*Snehana*	*Aragwadha mahatikthaka ghritam* – starting dose 25mg		
Stage 3	*Sodhana*	*Virechana– Avipathi choornam 25 g*		
		Peyadi -3 days		**69**
Stage 4	*Samanam*	*Patoladi kasayam 90 ml bd b/f* *Avipathi choornam 5g bd with kasayam* *T. vilwadi 1-0-1*	Soaking with *Triphala + Karanja patram*	
Stage 5	*Snehana*	*Manjishtadi kasayam 90ml bd b/f,* *Aragwadhamahatikthakam ghritam 5g bd with Kasaya*		
Stage 6	*Shodhana*	*Virechanam – Avipathi choorna 25 g*		
		Peyadi krama – 3 days		**24**
Stage 7		*Tikthaka ghritam 5g bd b/f*	*Takradhara*	
Stage 8			*Nasyam – Anutailam 2.5 ml* each nostril	
Stage 9	*Samanam*	*Aragwadhadi kasayam* *T. Sudarshanam*	*Udgharshana - Vara choornam +* honey	**6**

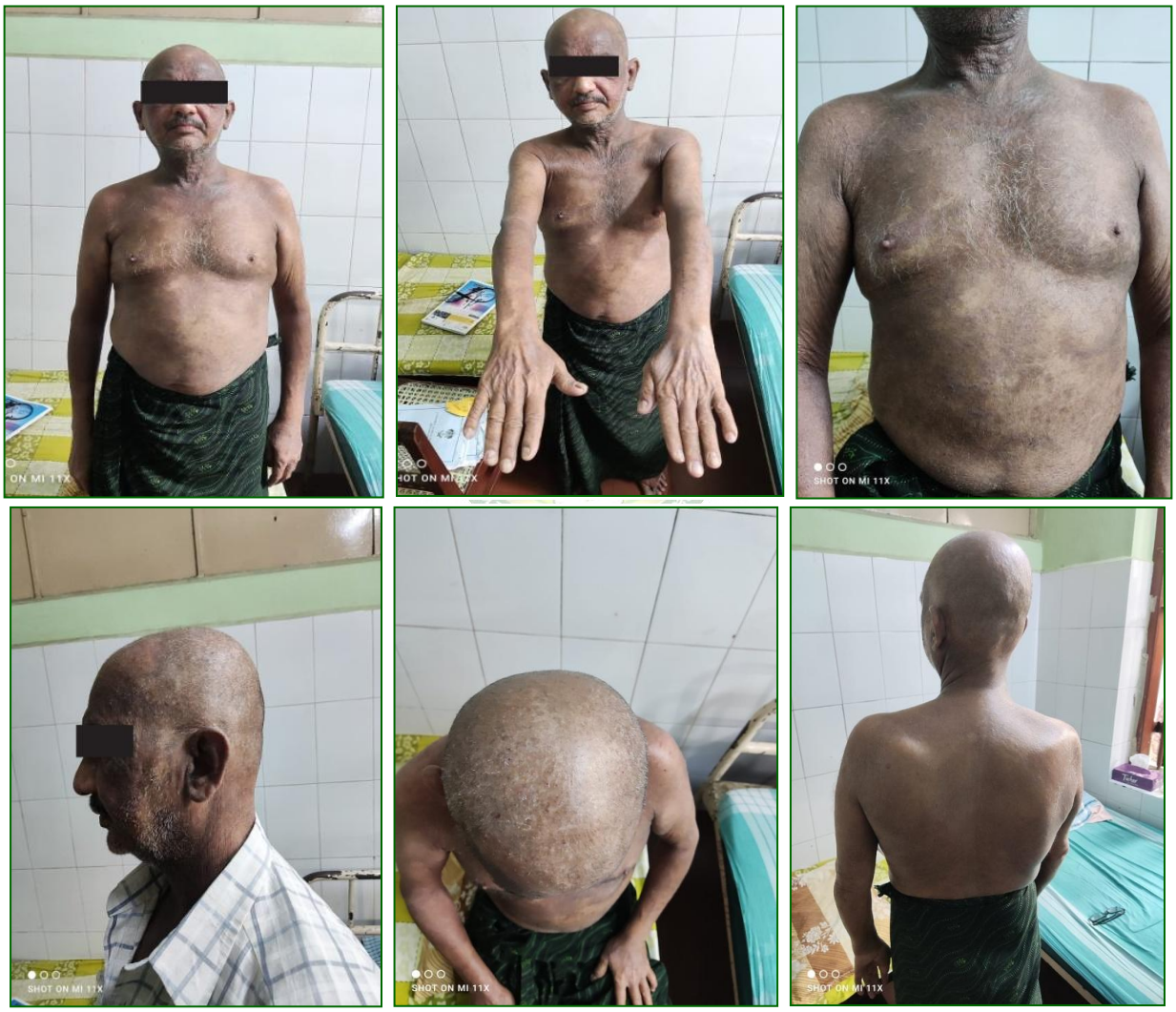
Advice on Discharge

1. *Manibhadra gulam*
2. *Eladi keram* – External application





Before Treatment



After Treatment

RESULT AND DISCUSSION

Pemphigus foliaceus is a rare autoimmune disease characterized by painful itchy blisters and sores in the skin. The most common places are scalp, face, neck and back. Considering various factors favouring diagnosis it has been diagnosed as *Visphota kushta* and the treatment principles mentioned in the context of *Kushta chikitsa* has been adopted. [10] *Kushta* is a disease characterised by vitiation of all the three *Dosas*. Initial phase manifests as inflammation and the involvement of *Kapha pitha dosas* has to be taken into account. Considering *Amatwa* and *Kapha pithadushti* initially *Langhana* in the form of *Deepana pachana* was adopted. After the inflammatory phase on attaining *Niramavastha, Snehapana* can be started. *Aragwadha mahatikthaka ghrita* was taken for *Achapana* considering role of *Pitha kapha dosas* in pathogenesis.

After attaining *Samyak snigdha lakshana*, *Abhyanga* and *Ushnambusnana* has been done for *Dosotklesha*. The *Utklishta dosas* are expelled through *Sodhana*. Here the method of *Sodhana* was *Virechana* even though lesions were more in *Jatroordhwa* because the patient was unfit for *Vamana*. He had a h/o CAD and showed ECG variations. After giving *Vyadhi* specific *Samanoushadha*, *Shodhana* therapy is repeated. After the body is completely cleansed, *Sirodhara* followed by *Nasya* is done for *Sodhana* of *Dosas* localised in *Siras*. Finally, *Manibhadragudam* was given as discharge medicine for *Dosa sesha sodhanam*. Autoimmune diseases are characterised by continuous formation of antibodies. In this case, the patient already had a family history of psoriasis and h/o recurrent fungal infections. Hence any *Dosadushti* has the tendency of getting *Sthanasamsraya* at *Twak*. Autoimmune diseases are characterised by continuous formation of antibodies. Hence intermittent *Sodhana* is beneficial to clear out the accumulating antibodies. *Manibhadra gula* [11] is given as discharge medicine for 1 month since it contains *Rasayana* drugs like *Amalaki*, *Abhaya* etc and helps in *Nitya sodhana*.

Significant reductions in lesions were noticed after *Sodhana*. PDAI score was taken as the criteria for assessment. At the time of admission PDAI was 96 which reduced to 24 after *Sodhana*. At the time of discharge PDAI was 6. There was no recurrence of lesions on review.

CONCLUSION

The mentioned treatment protocol was found to be effective in *Visphota kushta*. Recurrence of disease was also prevented. Treatment protocol was selected on the basis of *Dosa* predominance. Repeated *Sodhana* and *Samanoushadhas* showed significant results in the management of disease.

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***Address for correspondence**
Dr. K Ambika
Professor and HOD
Dept. of Kayachikitsa,
Govt. Ayurveda College,
Thiruvananthapuram.
Email: ambikachitharanjan@gmail.com