Case Report

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Ayurvedic Modalities in the Management of a Suspected Hailey-Hailey Disease: A Case Report

Ayurveda Section

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ABSTRACT

Hailey-Hailey Disease (HHD), or familial benign chronic pemphigus, is a rare autosomal dominant genetic disorder caused by mutations in the *ATP2C1* gene, leading to impaired calcium homeostasis in the Golgi apparatus. A 28-year-old male presented with recurrent painful blisters, erosions, and crusting in the axillary region, accompanied by intense pruritus and burning sensation, persisting for two years. Diagnosis was clinically established based on characteristic lesions in intertriginous areas and a positive family history. This case explores an integrative treatment approach combining Ayurvedic and modern dermatological approaches and highlights the potential of evidence-based integrative management in HHD.

Keywords: ATP2C1 gene, Calcium homeostasis, Intertriginous areas, Skin blisters

CASE REPORT

A 28-year-old male patient reported the Outpatient Department (OPD) of Kayachikitsa with complaints of painful skin blisters, erosions, and crusting localised to the axillary region, associated with intense itching, burning sensation, and discomfort, severely aggravated for the past two years. The symptoms followed a relapsing and remitting course, with periodic aggravations especially during hot and humid weather, excessive sweating, tight clothing, and friction. Temporary relief was noted with cool environments. Previously, the patient had consulted with a dermatologist and received topical corticosteroids (Clonate-F cream), a tablet Wysolone 10 mg once daily and tablet Amoxyclav 625 mg. There was no history of systemic illnesses like hypertension, diabetes, thyroid dysfunction, tuberculosis, asthma, or allergies. The family history was positive, as his father had a history of HHD. The patient followed a mixed diet, had good sleep, and had regular bowel habits. For objective evaluation, standardised dermatological assessment tools were employed, including Dermatology Life Quality Index (DLQI) [1], Physician's Global Assessment (PGA) [2], and Visual Analogue Scale (VAS). Photographic documentation was also maintained as part of the follow-up. The following *Ashthavidha Pariksha* (Eight folds of examination) was done [Table/Fig-1].

S. No.	Examination	Observation
1.	Nadi (Pulse rate)	80 times/minute, *kapha pitta*
2.	Mutra (Frequency of micturition)	4-5 times per day, samyak
3.	Mala (Bowel)	Regular, Prakrita
4.	Jihva (Tongue)	Saam
5.	Shabda (Sound)	Spashta
6.	Sparsha (Touch)	Anushnasheet
7.	Drik (Vision)	Prakruta
8.	Akriti (Body built)	Madhyam

[Table/Fig-1]: *Ashthavidha Pariksha* (Eight folds of examinations).

The patient's vital signs were within normal limits, with a blood pressure of 126/80 mmHg, respiratory rate of 16 breaths per minute, pulse rate of 80 beats per minute, and body temperature of 98.6°F. Haematological Investigations: Complete Blood Count (CBC), Erythrocyte Sedimentation Rate (ESR), fasting blood sugar, and post-prandial sugar were within normal limits.

The diagnosis of HHD was established based on characteristic clinical features and a positive family history, further supporting the provisional diagnosis, consistent with the autosomal dominant inheritance pattern of HHD. The diagnosis was confirmed clinically without genetic testing, as the patient declined genetic counselling. A dermatologist made the final diagnosis after evaluating lesion morphology, clinical presentation, and family history. Differential diagnoses such as pemphigus vulgaris, fungal and bacterial infections (including impetigo), and contact dermatitis were considered; however, HHD was deemed the most likely.

The treatment began with *Shodhan Chikitsa* (bio-purification therapy) to eliminate vitiated doshas and improve therapeutic outcomes. treatment therapy started with *Deepana* and *Pachana* using *Trikatu churna* for three days, followed by *internal Snehapan* with gradually increasing doses of *Panchatikta Ghrita* from Day 4 to Day 9. *Local Snehana* with *Marichyadi taila* and *Swedana* were given on Days 10 and 11. On Day 12, *Virechana* was performed using *Icchabhedi rasa*, resulting in effective purgation [Table/Fig-2].

Procedure	Medication	Dose and frequency	Duration	Route of medication
Deepana and *Pachan* (appetiser and carminative)	*Trikatu powder*	5 gm twice a day with lukewarm water	(Day 1, Day 2, Day 3)	Orally
Internal Snehapan (internal administration of medicated ghrit)	*Panchtikta Ghrit*	1st day (8 am)-30 mL 2nd day -60 mL 3rd day -90 mL 4th day- 120 mL 5th day -150 mL 6th day -180 mL	(Day 4, Day 5, Day 6, Day 7, Day 8, Day 9)	Orally
Local *Snehan* and *Swedan* (Oleation and sudation)	*Marichyadi taila*	Once a day	(Day 10, Day 11)	massage
Virechan (Purgation)	*Icchabhedi rasa*	2 tabs with cold water	Day 12	orally

[Table/Fig-2]: Shodhan Chikitsa.

Following the *Virechana procedure*, a structured *Sansarjana Krama* (post-purification diet regimen) was advised in a phased manner

over five days, beginning with easily digestible and light preparations [Table/Fig-3].

After completing Shodhan and *Sansarjana Krama*, a three-month *Shaman Chikitsa* was initiated as shown in [Table/Fig-4]. The combined regimen provided effective symptomatic relief and helped prevent recurrence.

Diet	Route of administration	Date
Morning – lukewarm water Evening -PEYA (Rice +14 times water)	Orally	Day 13
Morning - PEYA (Rice +14 times water) Evening-*Vilepi* (Rice +6 times water)	Orally	Day 14
Morning – *Vilepi* (Rice +6 times water) Evening-*Akrut Yush* (moong dal + 16 times water)	Orally	Day 15
Morning – *Kruta Yush* (moong dal + 16 times water + spices) Evening-*akruta mamsa rasa* (mamsa + 4 times water)	Orally	Day 16
Morning - *Krut Mamsa rasa* (*mamsa* + 4 times water + Spices) Evening- normal food	Orally	Day 17

[Table/Fig-3]: Diet regime for 5 days after *virechan*.

S. No.	Medications	Dose	Anupan and frequency	Duration
1.	*Sutshekhar rasa*	2 tab	Thrice a day before food for chewing	3 months
2.	*S -kin vati*	2 tab	Twice a day after food with lukewarm water	3 months
3.	*Mahamanjishthadi kwath*	4 tsp	Twice a day, after food, with an equal amount of lukewarm water	3 months
4.	*Avipattikar churna*	1 tsp	Twice a day before food with lukewarm water	3 months
5.	*Shatdhaut ghrit cream*	For external application	Thrice a day	3 months
6.	*Paadchira ointment*	For external application	Twice a day	3 months
7.	*Marichyadi taila*	For external application	At night	3 months

[Table/Fig-4]: *Shaman* treatment.

Clinical progress was assessed at regular intervals using key parameters [Table/Fig-5] [1,2]. Initially, the patient presented with painful blisters, erosions, crusting, and extensive axillary skin involvement. Over the 90-day treatment period, lesions showed gradual healing with complete resolution of crusting and erosions by day 90.

The impact of the condition on the patient's quality of life was assessed using the DLQI, as presented in [Table/Fig-6]. Initially, the DLQI score was 13, indicating a moderate effect on daily life due to discomfort, embarrassment, restrictions in clothing choices, and limitations in social and physical activities. With treatment, there was progressive improvement across all parameters. By day 90, the DLQI score had reduced to 2, reflecting minimal impact on the patient's quality of life, with most concerns such as pain, social limitations, and emotional distress significantly alleviated.

Visual assessment of the lesion progression was documented at four key intervals, as shown in [Table/Fig-7]. On the 0th day, the bilateral axilla showed painful blisters, erosions, and extensive crusting. By the 90th day, significant healing was observed with no active lesions, indicating complete resolution and healthy skin regeneration.

DISCUSSION

The HHD, or familial benign chronic pemphigus, is a rare autosomal dominant genodermatosis marked by recurrent vesicular and

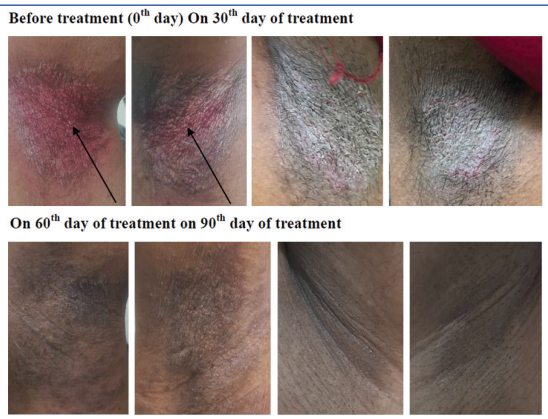
S. No.	Assessment parameters	Before treatment (0th day)	1st follow-up (on 30th day)	2nd follow-up (on 60th day)	After treatment (on 90th day)
1.	Lesion Appearance	Painful, blisters, erosions and crusting	Reduced pain and crusting; partial healing of lesions	Further healing; very few residual marks	Significant reduction in lesion size; healing of erosions; no crusting
2.	Skin involvement	Extensive armpit affected	Decreased area of involvement; fewer active lesions	Minimal skin involvement; healthy skin starting to restore	No active lesions and improved skin condition
3.	Pain level (VAS score)	3	2	1	0
4.	Itching/ Burning sensation	Persistent and intense	Noticeably reduced itching and burning	Occasional mild itching, no burning	Minimal or absent
5.	Dermatology Life Quality Index (DLQI) [1]	13	8	4	2
6.	Physical Global Assessment (PGA) scale [2]	3	2	1	0

[Table/Fig-5]: Observation and results [1,2].

S. No.	Assessment parameters	Before treatment (0th day)	1st follow-up (on 30th day)	2nd follow-up (on 60th day)	After treatment (on 90th day)
	How much discomfort (itching, soreness, pain, or stinging) have you experienced?	2	1	1	0
	How embarrassed or self-conscious have you felt because of your skin?	1	1	0	0
	Has your skin made it difficult to shop, maintain your home or garden?	1	1	0	0
	How much has your skin affected the clothes you wear?	2	1	1	1
	How much has your skin interfered with your social or leisure activities?	2	1	0	0
	Has your skin made it difficult for you to do any sports or physical activities?	2	1	1	1
	Has your skin prevented you from working or studying?	1	1	0	0
	Has your skin caused issues with your partner, friends, or relatives?	1	1	1	0

	Has your skin caused any difficulties with sexual activity?	1	0	0	0
	Has your skin treatment been problematic (time-consuming, messy, etc.)?	0	0	0	0
	Total score	13	8	4	2

[Table/Fig-6]: Dermatology Life Quality Index (DLQI) Grading.



[Table/Fig-7]: Difference in lesions of bilateral axilla before, 30th, 60th, and 90th day of treatment.
Note: On the 0th day, clinical image arrows showed the small blisters

crusted lesions in intertriginous areas such as the axillae, groin, and inframammary folds [3]. Typically presenting in the second or third decade [4], HHD follows a chronic-relapsing course, with flare-ups triggered by heat, sweating, friction, and secondary infections. In this case, the patient exhibited painful blisters, erosions, and crusting localised to the axillae, accompanied by intense itching and burning. Baseline scores indicated moderate disease burden VAS 3/10, DLQI 13/30) and PGA moderate severity, highlighting both physical and psychosocial impact. Conventional treatments like corticosteroids, antimicrobials, and immunomodulators offer symptomatic relief but carry risks such as skin thinning and systemic complications. Surgical options like excision or CO_2 laser ablation may be considered in refractory cases, though recurrence and scarring remain concerns [4]. While HHD lacks a direct Ayurvedic correlate, it may be interpreted under Kushtha Roga or Tvak Vikara, involving *Pitta Pradhan Kapha Dushti* and *Raktadosha* [5,6]. Ayurvedic management in this case combined *Shodhana* (bio-purification) and *Shamana* (palliative) therapies. *Virechana Karma* was advised to eliminate vitiated *Pitta* and *Rakta Dosha*, followed by *Samsarjana Krama* with light, digestible preparations such as *Peya*, *Vilepi*, *Akrita/Krita Yusha*, and *Akrita/Krita Mamsa Rasa*. After 90 days, the patient showed marked improvement, VAS 0/10, DLQI 2/30, and PGA mild severity. Treatment began with *Deepana-Pachana* to strengthen *Agni* and reduce *Pitta* aggravation, followed by *Snehapana* and *Snehan* for tissue lubrication, Vata pacification, and toxin mobilisation. *Swedan* therapy helped alleviate burning and irritation [7], preparing the body for *Virechana* to eliminate systemic toxins and reduce flare-ups. Ayurvedic formulations such as *Sutshekhar Rasa*, S-Kin tablets, *Mahamanjishtha Kwath*, *Paadchira ointment*, *Shatdhaut Ghrita*, *Marichyadi Taila*, and *Avipattikar Churna* were used to balance doshas, purify blood, reduce inflammation, and support skin healing. Although no specific Ayurvedic protocols for HHD exist, this case suggests that integrative approaches may offer promising avenues for its management [8-10]. The therapeutic sequence began with *Deepana-Pachana* using *Trikatu Churna* to enhance *Agni* and mitigate *Pitta* aggravation, followed by *Snehapana* and *Snehan* for tissue lubrication and toxin mobilisation. *Swedana* therapy alleviated burning sensations and prepared the body for *Virechana*, which effectively eliminated systemic toxins and reduced flare-ups. Post-purification dietary regimen (*Sansarjana Krama*) ensured metabolic stability and digestive recovery.

The three-month *Shamana Chikitsa* regimen incorporated classical Ayurvedic formulations such as *Sutshekhar Rasa*, *Mahamanjishthadi Kwath*, and *Avipattikar Churna*, along with topical applications including *Shatdhaut Ghrita* and *Paadchira Ointment*. These interventions collectively contributed to dosha equilibrium, blood detoxification, anti-inflammatory effects, and enhanced skin repair.

A case study by Saraf S et al., (2022) demonstrated the clinical efficacy of *Virechana Karma* as a *Shodhana* procedure, followed by *Panchatikta Ghrita Guggul* and *Mahamanjishthadi Kwath* as *Shamana* therapy, in a 21-year-old male with plaque psoriasis. The patient, presenting with erythematous scaly lesions and pruritus across limbs and trunk, underwent Ayurvedic intervention over four months. Notably, the *PASI* score reduced from 29.9 to 3.5, indicating substantial symptomatic relief and lesion regression. This case reinforces the therapeutic value of integrating *Sanshodhana* and *Sanshamana* approaches in managing *Ek Kushtha*, aligning with classical Ayurvedic principles and offering a safe, effective alternative for chronic dermatological conditions [11].

Aggarwal K et al., (2014) presented a case of a 27-year-old female with chronic palmoplantar dermatitis (*Vicharchika*), successfully managed through a combination of *Shodhana* and *Shamana Chikitsa* over four months. The study aimed to evaluate the therapeutic efficacy of Ayurvedic interventions and examine the influence of etiological factors on disease progression and remission. The patient showed marked clinical improvement, underscoring the relevance of individualised, causative-factor-oriented treatment strategies in chronic inflammatory skin conditions [12].

Collectively, these studies underscore the value of Ayurvedic interventions in managing chronic and refractory skin disorders. When integrated with modern diagnostic frameworks and outcome measures, personalised, dosha-specific regimens, as demonstrated in the present case, offer promising avenues for effective and holistic management of HHD.

CONCLUSION(S)

After a treatment period of three months, there was a marked improvement in clinical outcomes. The assessment parameters utilised, including the DLQI, PGA, and VAS, demonstrated significant positive changes. These findings indicate enhanced skin health, reduced symptomatology, and an improved quality of life for the patients involved. This case study underscores the potential efficacy of Ayurvedic interventions in the integrative management of HHD. Furthermore, it highlights the necessity for larger controlled studies to substantiate these outcomes and advocates for collaborative research between Ayurveda and modern dermatology to develop comprehensive care models for chronic and rare skin disorders.

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