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Case Series

Pathology Section

Clinico-pathological Spectrum of Rare Skin Syndromes and Diseases: A Series of Five Cases

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ABSTRACT

A significant threat to patients' well-being, mental health, capacity to function, and social participation- a measure of disability- is posed by skin diseases, resulting in significant mortality and morbidity worldwide. Despite the fact that the majority of rare diseases are complex, disabling, and life-threatening, little knowledge has been gained in this area. The diagnosis and classification of these rare skin syndromes with pre-determined sets of symptoms present a challenge. To diagnose a rare skin syndrome, one frequently has to correlate histologic findings with clinical symptoms, as there is a vast range of skin disorders, many of whose histologic traits overlap with just slight variances. However, histologic knowledge alone makes it difficult to diagnose these; proper diagnosis demands appropriate clinical knowledge. In the present study institute, 675 skin biopsies were performed over the course of five years, from 2018 to 2022. Based on clinico-pathological analysis, various rare skin syndromes were diagnosed, as depicted in present case series of five cases (10 years old boy, 40 years old male, 27 years old male, 44 years old male, 52 years old male patients) for: a) Griscelli Syndrome; b) Gougerot-Carteaud Syndrome, considered rare as it is one of the underdiagnosed and misdiagnosed syndromes due to its similarity with Pityriasis versicolour; c) Kyrle's disease, a rare perforating dermatosis occurring in 10% of chronic renal failure patients who are on dialysis; d) Nekam's disease; e) Sweet syndrome, an uncommon syndrome occurring in about 10-20% of malignancies.

Keywords: Griscelli syndrome type 3, Kyrle's disease, Rare disease, Sweet syndrome

INTRODUCTION

When determining health priorities, skin-related illnesses are often neglected in comparison to conditions that have a substantial mortality rate [1,2]. Skin conditions are frequently visible indications of far more serious systemic diseases, which can be made clear from comprehensive history-taking, general physical examination, and systemic examination [3]. Combining all of these improves diagnosis accuracy, which in turn improves treatment effectiveness [3]. Between 2010 and 2013, skin-subcutaneous diseases were the fourth-leading cause of non-fatal disease burden across the globe [4].

The Department of Pathology has received a total of 675 skin biopsies from the Department of Dermatology over five years (2018-2022). With excellent clinical and histological correlation, the diagnosis of five rare syndromes among 675 patients was made. The following case series reports exceptional cases of five rare skin syndromes. This could add crucial evidence and aid future researchers.

CASE SERIES

Case 1

A 10-year-old boy presented with discolouration of hair that had been present from birth, as well as dark lesions over sun-exposed areas of the face, the extensor aspect of the forearms, and the V region of the neck for four years. This patient was born out of a consanguineous marriage, and no one in the family had a history of similar complaints. Growth indicators were normal during the general physical examination, and a cutaneous examination revealed silvery grey hair throughout the body, including the scalp, brows, and eyelashes. Defined hyper-pigmentation with a few hypo-pigmented macules was present over the cheeks, the V area of the neck, the extensor aspect of the legs, and forearms. [Table/Fig-1a,b] depicts the features of the 10-year-old boy. Palms, soles, mucosa, and nails were normal. Xeroderma pigmentosa, Chediak-Higashi syndrome, and Griscelli syndrome - Type 3 were all considered in the clinical differential diagnoses.

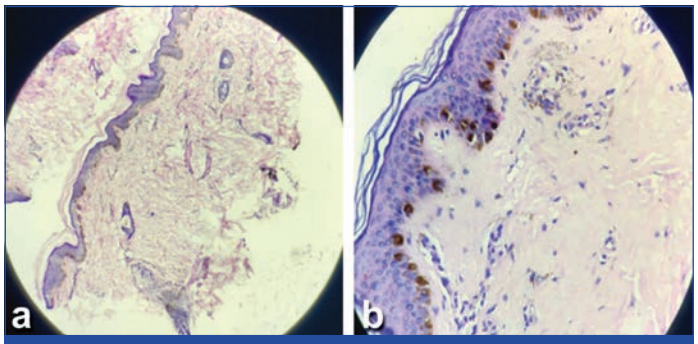
[Table/Fig-1]: a) Silver grey hair over the face and eyebrows and eyelashes. Defined hyper-pigmentation with few hypo-pigmented macules present over cheek; b) Hyperpigmentation present over V area of neck of a 10-year-old boy.

Anaemia (Hb-11.3 g/dL) was identified by laboratory testing, and the total leucocyte count and platelet count were both within normal ranges. The Erythrocyte Sedimentation Rate (ESR) was 100 mm at the end of 1 hour, and C-reactive Protein (CRP) was negative. The pelvic ultrasonography was normal.

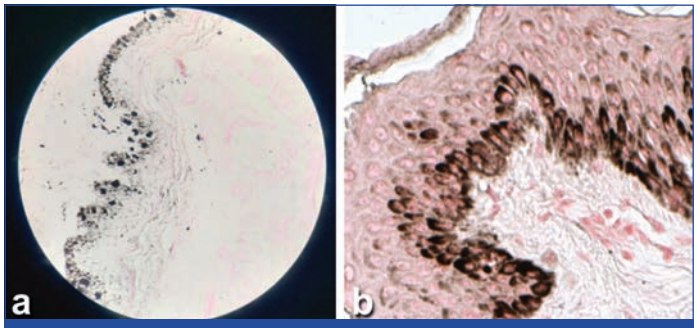
For microscopic analysis, skin biopsies were obtained from hyper- and hypo-pigmented lesions over the forearm. The microscopic section under study revealed stratified squamous epithelium with loss of rete ridges and hyperkeratosis. The Stratum Basale showed abnormal accumulation of melanin pigment. The dermis displayed basophilic degeneration of collagen fibers with entrapped adnexal structure. [Table/Fig-2a,b] represents the histopathological slides of the patient.

Xeroderma pigmentosa was initially identified as a possible diagnosis by histological analysis. However, a clinical history and general physical examination indicated hair discolouration, dark-coloured lesions across sun-exposed parts of the face, the extensor aspect of the forearm and legs, and the V area of the neck. This necessitated further Fontana-Masson special staining [Table/Fig-3a,b] and hair

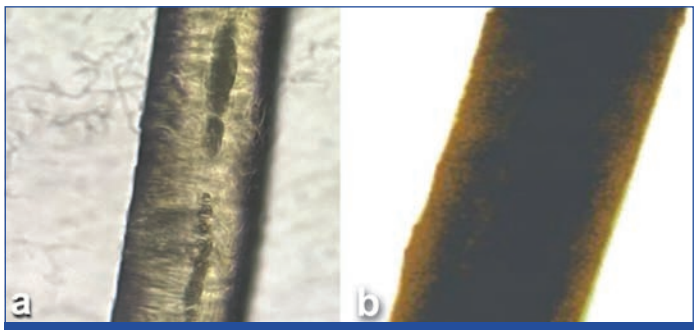
shaft examinations (depicted in [Table/Fig-4a,b]) in order to confirm the diagnosis of Griscelli syndrome - Type 3.



[Table/Fig-2]: a) Hyperkeratosis, abnormal accumulation of melanin pigment in Stratum Basale. Dermis shows basophilic degeneration of collagen on 10x magnification (H&E stain); b) Abnormal accumulation of melanin pigment in Stratum Basale on 20x magnification (H&E stain).



[Table/Fig-3]: a) Irregularly stained melanin in basal layer on Fontana Masson Stain (10x magnification); b) Irregularly stained melanin in basal layer on Fontana Masson Stain (40x magnification).



[Table/Fig-4]: a) Microscopy of hair shaft showing irregular clumps of pigment on 40x magnification; b) Microscopy of normal hair shaft showing small homogenous pigmented granules on 40x magnification.

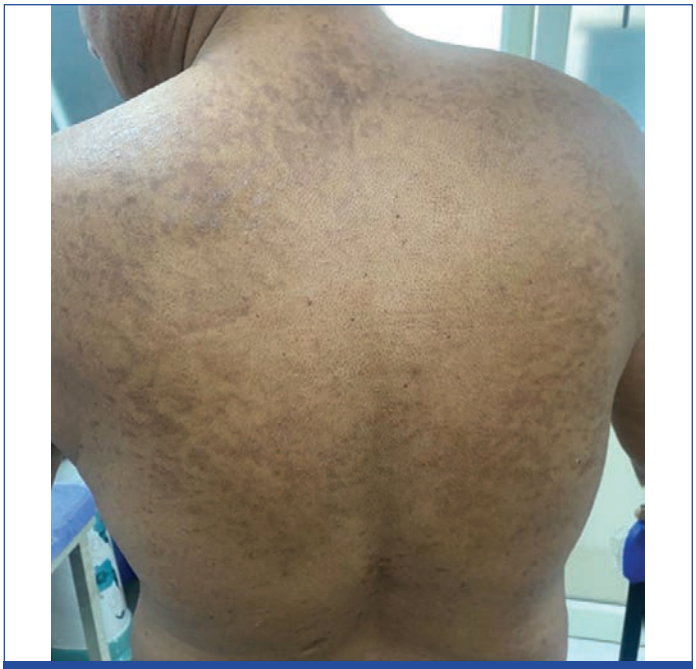
The diagnosis of Griscelli syndrome - Type 3 was confirmed by clinical manifestations (visible changes to the skin and hair) and hair shaft microscopic evaluation in the 10-year-old boy. After reassurance to the parents and the patient, he was discharged with no further follow-up.

Case 2

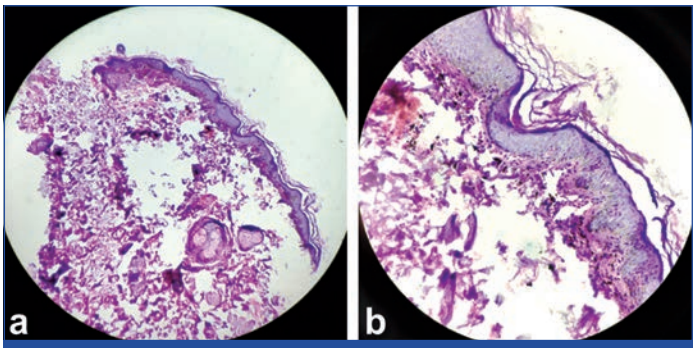
A 40-year-old male patient presented with a one-month history of itchy, scaling lesions on his back and torso. No notable family or medical history of such symptoms was found.

General physical examination revealed well-defined, multiple hyperpigmented plaques and macules with scaling over the face, chest, trunk, bilateral upper limbs and back as depicted in [Table/Fig-5]. Pityriasis versicolour was clinically diagnosed, and the patient was prescribed anti-fungal medication. However, a skin biopsy was advised when anti-fungal therapy failed to relieve symptoms. This mandated a thorough histological examination of the skin biopsy obtained from the lesion over the trunk, which demonstrated orthokeratosis with focal papillomatosis, acanthosis, and melanin incontinence of the epidermis as depicted in [Table/Fig-6a,b].

Based on the clinical presentations, in dermatological consultations, the diagnosis of tinea versicolour pigmented form was frequently



[Table/Fig-5]: Well-defined, multiple hyperpigmented plaques and macules over back in reticular pattern.



[Table/Fig-6]: a) Focal area of papillomatosis on 10x magnification (H&E stain); b) Illustrates an acanthosis and melanin pigment incontinence on 40x magnification (H&E stain).

confirmed. However, an appropriate diagnosis of Gougerot-Carteaud syndrome (Confluent and reticulate papillomatosis), based on the histological presentation and physical results, was made.

Based on previous case studies which demonstrated that Tablet Doxycycline at a dose of 100 mg/day resulted in symptom alleviation, a similar prescription was made for the patient. The patient was requested to return to the Outpatient Department (OPD) for follow-up. However, he failed to do so, and additional follow-up was missed.

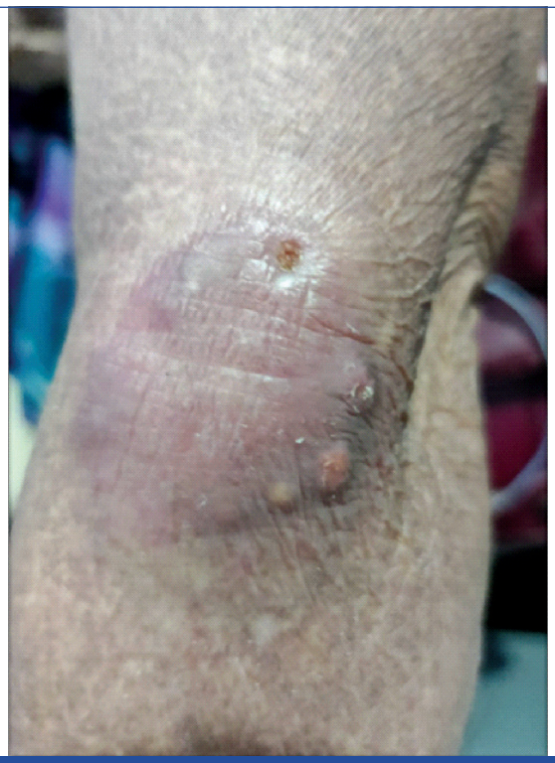
Case 3

A case of 27 years old male patient presented with the facility of fever, pedal oedema, and rashes all over his body for the previous 15 days. Itchy rashes that started on the right hand later spread to the upper body, trunk, and thighs. No notable family or medical history of such symptoms existed. On a general physical examination by dermatologists, there were multiple discrete erythematous plaques with central crusting, and a few satellite lesions were also seen. [Table/Fig-7] depicts the clinical presentation of the patient.

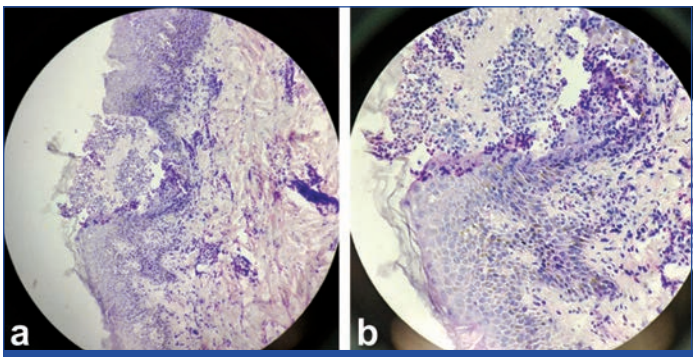
Peripheral smear examinations highlighted normocytic normochromic anaemia with neutrophilic leukocytosis. Laboratory investigations showed an increase of C-Reactive Protein (CRP 22.6 mg/L).

A skin biopsy was obtained from the lesion over the left forearm for microscopic evaluation. Histopathologic sections studied showed the epidermis having irregular acanthosis with a focal area showing spongiosis and a pustular lesion consisting of neutrophils, eosinophils, a few lymphocytes, and histiocytes. The papillary dermis was oedematous, and the reticular dermis showed peri-vascular infiltration

by lymphocytes, histiocytes, neutrophils, and a few eosinophils. [Table/Fig-8a,b] has depicted the histological results of the patient.



[Table/Fig-7]: Patient presenting with plaques with central crusting over the upper limb.



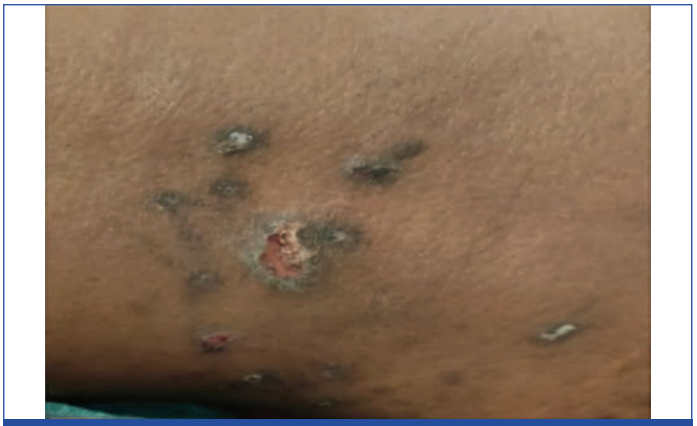
[Table/Fig-8]: a) Microscopic image showing irregular acanthosis with focal area of spongiosis and pustular lesion consisting of neutrophils on 10x magnification (H&E stain); b) Focal area of spongiosis and pustular lesion consisting of neutrophils, eosinophils, few lymphocytes and histiocyte on 40x magnification (H&E stain).

One must consider erythema multiforme, erythema nodosum, allergic dermatitis, Hansen's disease with type I lepra response, and other infectious disorders associated with rashes as differential diagnoses based on the clinical picture. However, the diagnosis of Sweet syndrome (Acute febrile neutrophilic dermatosis) was obtained using microscopic evidence. The patient was administered with Tablet (Tab.) Dapsone 100 mg OD for three months with regular monthly follow-up and fully recovered.

Case 4

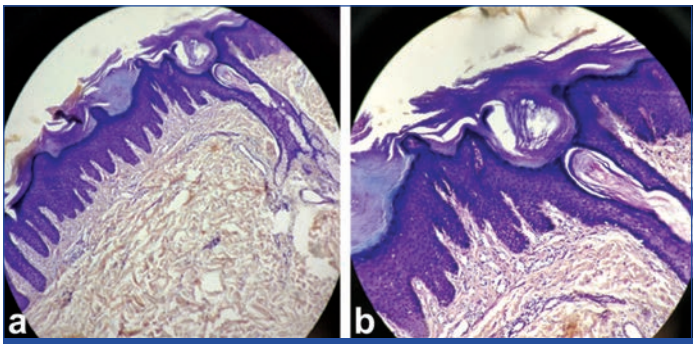
A 44-year-old male patient was seen with a six-month history of reddish, itchy lesions over the neck, face, trunk, bilateral upper limbs, and lower limbs. This patient was a known case of chronic renal failure for the past two years and had been undergoing dialysis. He was also a chronic alcoholic but had abstained for the past two years. No significant family history was provided.

General physical examination revealed multiple well-defined reddish to violaceous, hyperpigmented papules over the neck, face, trunk, bilateral upper limbs, and lower limbs. [Table/Fig-9] shows the clinical presentation of the patient. Based on the results of the clinical examination, Prurigonodularis and Kyrle's disease were the differential diagnoses.



[Table/Fig-9]: Well-defined hyperpigmented papules with focal area of ulceration seen over trunk.

A skin biopsy was obtained from the lesion on the thigh for histological examination. It revealed an epidermis with extensive hyperkeratosis, parakeratosis, hypergranulosis, and papillomatosis. A focus of extra follicular cornified plug was noted. The upper papillary dermis showed an increased microvasculature density with perivascular and peri-adnexal lymphoplasmacytic infiltration. [Table/Fig-10a,b] depicts the histological examinations.



[Table/Fig-10]: a) Microscopic image showing hyperkeratosis, parakeratosis, hypergranulosis and extrafollicular cornified plug on 10x magnification (H&E stain); b) Microscopic image showing hyperkeratosis, parakeratosis, hyper granulosis and extrafollicular cornified plug on 40x magnification (H&E stain).

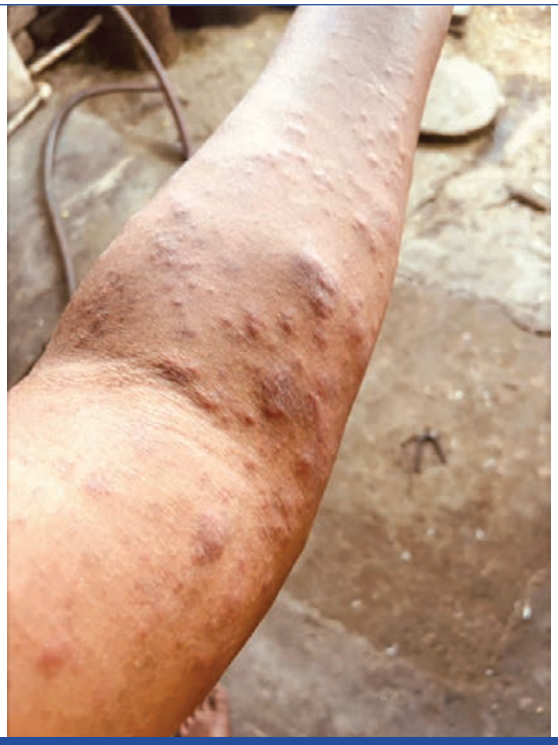
According to clinical and histological results, the patient was diagnosed with Kyrle's disease. He was then prescribed with topical corticosteroids and anti-histamines. Unfortunately, due to the patient's death, the clinicians could not follow-up with the patient further.

Case 5

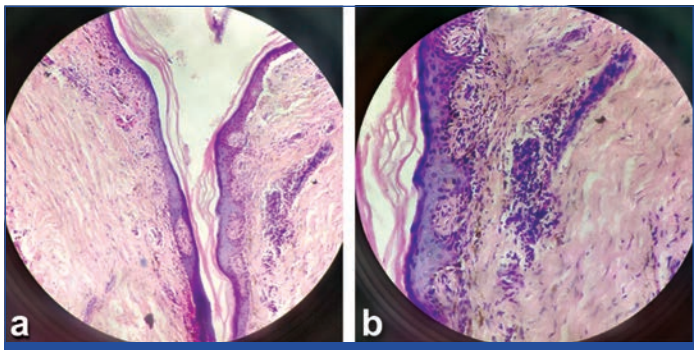
A 52-year-old male patient, presented with reddish skin lesions over the flexor aspect of both upper and lower limbs for three months. The patient did not provide any remarkable medical or family history. Clinical examination findings showed reddish to violaceous irregular, discrete to diffuse macules on both the upper and lower limbs [Table/Fig-11]. In order to distinguish between lichen planus and psoriasis, Nekam's disease and lichen planus-psoriasis overlap were taken into consideration.

A skin biopsy was obtained from the lesion over the right lower limb for microscopic evaluation. The microscopic section studied showed the epidermis having hyperkeratosis, parakeratosis, hypergranulosis, and necrotic keratinocytes seen at places. Vacuolar degeneration of the basal cell layer was seen with melanin incontinence. At places, the epidermis was atrophic. The dermis showed peri-adnexal and peri-vascular dense chronic inflammatory cell infiltration consisting of lymphocytes, histiocytes, and plasma cells [Table/Fig-12a,b]. Microscopic features provided a definitive diagnosis of Nekam's disease (Keratosis lichenoides chronica).

The patient was started on systemic corticosteroids, and simultaneously Psoralen Plus Ultraviolet-A (PUVA) radiation therapy was advised, but the patient took discharge against medical advice.



[Table/Fig-11]: Reddish violaceous irregular, discrete to diffuse macules over flexor aspect of upper limb.



[Table/Fig-12]: a) Hyperkeratosis and focal area of epidermal atrophy on 10x magnification (H&E stain); b) Shows evident Vacuolar degeneration of basal cell layer is seen with melanin incontinence on 40x magnification (H&E stain).

The clinical and diagnostic details of all five cases altogether are depicted in [Table/Fig-13].

DISCUSSION

The clinical and histological characteristics of a wide range of skin diseases and conditions commonly overlap, making diagnosis difficult and ultimately hampering therapy. The clinical and microscopic features of the above-discussed cases are summarised in [Table/Fig-13], which further helps in comparing and distinguishing them from other similar entities.

Griscelli Syndrome (GS): GS is an autosomal recessive multisystem genetic condition involving partial albinism as well as neurological and/or immunological abnormalities, caused by mutations in 15q21. Based on the point of mutation, three subtypes of GS have been identified as GS type 1, 2, and 3 [5]. GS type 1 and 3 have a very good prognosis, but GS type 2 has a poor prognosis, and it needs emergent diagnosis and treatment [6,7]. A delay in the diagnosis of these individuals may hinder treatment success. Additionally, as each form of the disease requires a particular course of therapy, it is critical to identify the patient's disease type as early as possible. Patients with GS1 are recommended palliative and supportive therapy, whereas GS3 patients do not need treatment and have an excellent prognosis [6,7]. In a case report by Mansouri Nejad SE et al., based on clinical signs and symptoms, three differential diagnoses were considered, namely: Elejalde syndrome, Chediak-Higashi, and Griscelli syndrome type-2. Griscelli syndrome diagnosis was confirmed by clinical manifestations and hair shaft microscopic evaluation [6]. Similar to the previously mentioned cases [6-8], the present patient's case presents clinical history-based differential diagnoses of Griscelli syndrome, Chediak-Higashi syndrome, and Xeroderma pigmentosa. Later, by using clinico-histologic correlation, the diagnosis of Griscelli syndrome type-3 was confirmed. Additionally, proving this disorder's autosomal recessive inheritance pattern is the fact that the patient's parents disclosed their history of consanguineous marriage. Due to the high rate of consanguineous marriage within our country, microscopic analysis of the skin and hair is advised in patients with immunodeficiency, organomegaly, and pancytopenia to rule out autosomal-recessive illnesses like GS.

Gougerot-Carteaud syndrome: Is a rare and benign disorder. The clinical presentation of Confluent and Reticulated Papillomatosis (CARP) is that of hyperkeratotic or verrucous brown papules that coalesce into plaques with a reticulated periphery and is most often clinically mistaken for tinea versicolour [9], and usually does not respond to therapy with antifungals. The diagnostic criteria for CARP proposed by Davis MD et al., require [10]:

1. Clinical findings of scaling brown macules and patches, some reticulated and papillomatous.
2. Location on the upper trunk and neck.
3. Section studied negative for fungal elements (fungal staining included).
4. Lack of response to antifungals.
5. Excellent response to minocycline.

Syndromes/ diseases	Age/sex	Clinical features	Microscopic features
Griscelli syndrome-Type 3	10 year/Male	Slivery grey hair all over body. Well defined hyper and hypopigmented macules noted (Features of partial albinism). No systemic or neurological involvement.	Hyperkeratosis with melanin accumulation in stratum basale and dermis displays entrapped adnexal structures with collagen degeneration. Hair microscopy shows irregular clumping of melanin.
Gougerot-Carteaud syndrome	40 year/Male	Hyperpigmented or verrucous papules that are coalesced into plaques with a reticulated periphery are noted. Often confused with tinea versicolour.	Orthokeratosis with focal papillomatosis. Acanthosis and melanin pigment incontinence.
Sweet syndrome	27 year/Male	Multiple discrete erythematous plaques with central crusting and satellite lesions. Presented with fever and CRP was elevated (22.6 mg/L).	Irregular acanthosis with focal area showing spongiosis and pustular lesion consisting of mainly neutrophils, eosinophils. Papillary dermis oedematous. Reticular dermis shows perivascular inflammation.
Kyrels disease	44 year/Male	Multiple red-violaceous papules with a central keratotic plug. (Most common site is lower extremities and trunk). Associated with chronic renal failure.	Hyperkeratosis, parakeratosis, hypergranulosis and papillomatosis. Extrafollicularcornified plug is characteristic feature. Inflammation is composed of Lymphoplasmacytic cells.
Nekams disease	52 year/Male	Reddish to violaceous papules and macules present over upper and lower limbs.	Hyperkeratosis, parakeratosis, hypergranulosis and necrotic keratinocytes with Vacuolar degeneration of basal cell layer and melanin incontinenceare seen. Epidermis shows focal atrophic changes.

[Table/Fig-13]: Summary of clinical and microscopic features.

The present case 2 discussed here fulfilled the first 3 criteria of 5. The patient was not followed-up after diagnosis and starting treatment so the response to treatment could not be commented on.

Ahogo KC et al., observed two cases of 38-year-old and 29-year-old patients. One of the patients presented with a six-month history of 1 to 5 mm-diameter flat papular lesions that were grayish-pigmented in colour and had verrucose skin [11]. These lesions started in the interscapular region and spread to the upper back and whole chest. Then they came together in places by large losangic placards at the thoracic region. Like the case-study described in this paper, the patient found no remission with local and systemic anti-fungal therapy and was misdiagnosed as tinea versicolour. The histological examination revealed hyperkeratosis, acanthosis, and papillomatosis [12].

Sweet syndrome: Also, called acute febrile neutrophilic dermatosis, was first described by Dr. Robert Sweet in 1964 [13]. The characteristic presentation includes the sudden onset of well-defined tender plaques or nodules with fever and, on occasion, oral or genital lesions. Arthralgia and ocular inflammations are frequently encountered [14]. Most cases of Sweet syndrome are idiopathic; however, they may be associated with several other conditions, namely malignancies (myelodysplasia, acute myelogenous leukemia, chronic myelogenous leukemia, multiple myeloma, monoclonal gammopathy, etc.), inflammatory and autoimmune diseases (inflammatory bowel disease, rheumatoid arthritis, Hashimoto's thyroiditis, etc.), and infections (viral hepatitis, HIV infection, tuberculosis, etc.). Cases of drug-induced Sweet syndrome have been documented with G-CSF (most common), antibiotics (minocycline, certain fluoroquinolones, nitrofurantoin, etc.), and several other drug classes, including NSAIDs, cancer chemotherapy agents, immunosuppressants, etc. Though several contributory factors induce the disease, the exact pathogenesis is yet unknown. However, cases in Japan have observed an HLA-B54 association, and some other cases have reported MEFV gene mutation in patients with Sweet syndrome [15].

Sweet syndrome is diagnosed based on conventional clinical, laboratory, and histology findings. The diagnostic criteria have been listed below. Major criteria include an abrupt onset of painful erythematous plaques or nodules and dense neutrophilic infiltrate without evidence of leukocytoclastic vasculitis by histopathology. The minor criteria include the presence of fever >38°C, the presence of an underlying haematologic or visceral malignancy, inflammatory disease, or pregnancy, OR preceded by an upper respiratory or gastrointestinal infection or vaccination. The patients usually respond to treatment with systemic corticosteroids or potassium iodide excellently, and abnormal laboratory values at presentation (three of four): Erythrocyte Sedimentation Rate (ESR) >20 mm/hour; positive CRP; >8000 leukocytes; >70% neutrophils [16].

A case report of a 36-year-old female patient who presented with itchy, tender erythematous-based pustules and papules arranged in a vesicle-like plaque on the chest, upper limbs, and back. Histological examination revealed oedema and a diffuse infiltration of neutrophils in the papillary dermis with swollen endothelial cells. The history revealed a post-infectious sequela [17,18]. The case reported in the present paper also presented with an acute infectious history and cutaneous manifestations. Microscopy of the cutaneous lesion revealed a pustular lesion and predominant neutrophilic infiltration. The diagnosis was confirmed as Sweet syndrome by clinicopathological correlation [18].

Kyrle's Disease (KD): KD is a rare acquired perforating dermatosis characterised by the trans-epidermal elimination of abnormal keratin and is associated with an underlying disorder such as diabetes mellitus or chronic renal failure. This condition was first described by Dr. Josef Kyrle in 1916 [19]. The most important feature is the multiple, discrete, eruptive papules with a central keratotic plug on the lower extremities. The exact aetiology of the disease is still unknown; however, some knowledge of the disease's association with genetic predisposition has been brought to light. The exact management of Kyrle's disease is yet unknown, and the strategy is mostly evidence-based [20].

In a case report done by Nair PA et al., the diagnosis of Kyrle's disease was made in a 64-year-old with a history of multiple, large, discrete hyperpigmented, hyperkeratotic papules with central crusted keratotic plugs over bilateral extremities, back, neck, scalp, etc., [21]. Similar to the above-mentioned patient, a history of chronic renal failure on hemodialysis was noted. A biopsy taken from one of the papules from the right leg showed irregular epithelial hyperplasia with a follicular cornified plug and focal parakeratosis. The plug contained basophilic degenerated material [20,21].

Keratosis Lichenoides Chronica (KLC): Also, known as Nekam's disease, is a rare mucocutaneous disorder. When a patient presents with a constellation of features consistent with lichen planus, seborrheic dermatitis, and aphthous ulcers, Nekam's disease should be considered, and further evaluation of such cases can aid in the detection of a greater number of cases [22]. According to an article published by Aruna C et al., in 2016, only 128 cases have been documented worldwide to date [23]. With a slight male preponderance, the fourth decade indicates a peak in occurrence. KLC can be identified by asymptomatic, violaceous, scaly papules that are organised in a reticular pattern across the body, most frequently on the limbs, and that have facial characteristics similar to rosacea or seborrheic dermatitis. KLC exhibits a poor response to therapy and runs a chronic, progressive course. The exact pathogenesis was not elucidated; however, an immune-mediated theory has been suggested [24].

The patient in case 5 presented with features mimicking lesions seen in lichen planus without the presence of aphthous ulcers or features of seborrheic dermatitis. However, microscopic study of the lesion showed parakeratosis, hyperkeratosis, alternate areas of acanthosis and atrophy, vacuolar degeneration of the basal cell layer with melanin incontinence, dermo-epidermal junction showed plasma cell infiltrates and dilated dermal capillaries, confirming the diagnosis of Nekam's disease.

CONCLUSION(S)

This case series aids in generating crucial evidence of the five rare skin syndromes, including Griscelli Syndrome, Gougerot-Carteaud Syndrome, Kyrle's illness, Nekam's disease, and Sweet syndrome. Most of the rare skin syndromes present with clinical and histologic characteristics that overlap with minute distinctions, making it frequently necessary to link histologic results with clinical characteristics in order to diagnose a case. Hence, it is of utmost importance to diagnose these syndromes to prevent the burden on the patient and plan an appropriate management strategy.

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