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Case Series

Exploring reticulate pigmentary disorders - A case series highlighting 5 distinctive types

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Abstract

The term "reticulate pigmentary disorders" is used in a broad sense to refer to a heterogenous group of acquired and congenital dermatosis because of their clinical pattern of presentation. Presentations range from the "freckle-like" hyper- and hypopigmented macules that are typically only seen in the true genetic "reticulate" pigmentary disorders to the reticular or net-like pattern. Clinically, the reticulate pigmentary disorders can be classified based on the extent and distribution into acral, flexural, and generalized. They can also be divided on the basis of age of presentation (infancy, childhood, adolescent, adult). In this case series, five distinct types of Reticulate pigmentary disorders, are reported. The first two cases are that of father and daughter diagnosed as dyschromatosis universalis hereditarian. The father was a 47-year-old man who presented with mottled hypo and hyperpigmented macules over face, trunks and limbs present since birth. The second case was his daughter who had similar lesions over dorsum of hands and dorsum of left foot. The third case was a 13-year-old girl who presented with a 2-month history of mottled hyperpigmented macules and few hyperpigmented papules over the axilla, groin along with few pits over forearms and was diagnosed as Dowling Degos disease. The fourth and fifth cases were that of young male patients with reticulated hyperpigmented macules over the neck, upper chest and arms, occurring on and off for 2 years. They were clinically diagnosed with Confluent and reticulated papillomatosis. The sixth case was a 65-year-old woman who presented with multiple flat lesions over left upper limb and clinically diagnosed with macular amyloidosis. The final case is of a 29-year-old male diagnosed with Darier's disease. This case series is being reported to highlight different reticulate pigmentary dermatosis of varied aetiologies.

Keywords: Reticulate pigmentation, Darier's disease, Dowling- Degos disease

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1. Introduction

The word 'Reticulate' is a common clinical term for skin lesions that have a "net-like," "sieve-like," or chicken wire" configuration.[1] This pattern of skin lesions is associated with a number of congenital and acquired dermatoses and other conditions.[2] In this case series we are reporting five distinct types of Reticulate pigmentary dermatoses of varied etiologies, namely Dyschromatosis universalis hereditaria, Confluent and reticulated papillomatosis, Dowling-Degos disease, Macular Amyloidosis and Darier's Disease.

Dyschromatosis universalis hereditaria **(**DUH) is a rare genodermatosis called characterized by hyper- and hypopigmented macules that can vary in size and shape. This disease spectrum includes unilateral dermatomal pigmentary dermatosis (segmental form), dyschromatosis universalis hereditaria (generalized form) and dyschromatosis symmetrica hereditaria (localized form).[3] Confluent and reticulated papillomatosis (CRP) of Gougerot and Carteaud is a relatively uncommon dermatosis which typically affects the neck, interscapular area, inframammary area, and abdomen, and is characterized by persistent papules and plaques that are confluent in the centre and reticulated at the periphery.[4] Dowling-Degos disease (DDD) is a rare autosomal dominant trait that is characterized by sporadic comedo-like lesions (dark dot, follicles), pitted acneiform scars, and numerous, symmetrical, progressive, and pigmented macules over the axillae, groins, face, neck, arms, and trunk.[5] Darier's disease (DD) is an uncommon

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keratinisation disorder which affects skin and oral mucosal. It is distinguished by seborrheic hyperkeratotic papules and nail abnormalities.[6] Macular amyloidosis (MA) is a unique type of skin amyloidosis, with amyloid deposited exclusively in the dermal layer. It is characterized by hyperpigmented patches, presenting as rippling, greyish-brown macules where amyloid accumulates in the papillary dermis. Lesions commonly appear on the upper back, arms, chest, and legs.[7]

2. Case Series

2.1. Case 1

A 47-year-old male patient presented with flat hyperpigmented and hypopigmented lesions over the face since birth. At age of 2 years, similar hyperpigmented macules started to develop over dorsum of bilateral hands and gradually involved other areas of the body including trunk and limbs. There was no history of photosensitivity, no history of handling chemicals and no history of intake of any drugs prior to the onset of lesions. There is no history suggestive of developmental delay. He was born of non-consanguineous marriage. There was history of similar lesions present in his grandmother and his daughter (Case 2). Systemic examination was normal. On examination, there were multiple mottled hyperpigmented and hypopigmented macules and patches present over face, neck, bilateral forearms, arms, dorsum of feet, legs, thighs, groin and genitalia. (**Figure 1**). There were few hypopigmented macules on the abdomen and palms. There were no lesions on soles and oral mucosa. Nails were normal. Based on the classical clinical presentation, he was diagnosed with Dyschromatosis Universalis Hereditaria. Biopsy was advised for histopathological examination, but the patient refused it. Patient was counselled about the condition. He was made aware of the limited response to treatment. He was given topical hydroquinone 4% cream and broad-spectrum sunscreen to be applied over sun exposed areas. He was advised to follow up after 1 month. However, he was lost to follow up.

2.2. Case 2

A 21-year-old girl (daughter of Case 1) presented with complaints of multiple flat hyperpigmented lesions over dorsum of both hands and dorsum of left foot since birth. Lesions did not progress in number or size. Lesions did not develop on other parts of the body. There was no history of photosensitivity, no history of handling chemicals and no history of intake of any significant drugs. There is no history suggestive of developmental delay. She was born of non-consanguineous marriage. History of similar illness was present in the father (Case 1) and her paternal great grandmother. Systemic examination was normal. On examination, there were multiple mottled hyper and hypopigmented macules on dorsum of both hands and on dorsum of left foot (**Figure 2**). Palms, soles, oral mucosa and nails were normal. She was diagnosed with DUH and was counselled regarding the condition. She was prescribed hydroquinone cream 4% cream to be applied over the hyperpigmented lesions and broad-spectrum sunscreen lotion to be applied over sun exposed areas. She was advised to follow up after 1 month. However, she was lost to follow up.

2.3. Case 3

A 13-year old girl presented with complaints of multiple flat and raised hyperpigmented lesions over axilla, groin, trunk and limbs since 2 months. There was no history of inflammatory skin diseases prior to the onset of the lesion. There was no significant past medical, surgical and gynaecological history. There was no history of similar complaints in family. The patient was born of a non-consanguineous marriage. Systemic examination was normal. On examination, there were multiple hyperpigmented macules over bilateral axilla, chest, anterior aspect of neck, dorsum of bilateral hands and feet, abdomen, back, flexor aspects of both forearms and lower one thirds of legs (**Figure 3**). Multiple pits of varying sizes were distributed over the medial aspect of bilateral thighs. The patient was diagnosed with Dowling Degos disease and was treated with topical steroids and retinoids over papules. There was mild clearance of hyperkeratotic papules but not complete resolution.

2.4. Case 4

A 28-year-old male with complaints of multiple asymptomatic flat and raised brownish lesions on neck, chest and back since the last 2 years. There was history of similar episodes on and off in the past which would resolve with treatment. On examination, there were multiple scaly brown macules, patches and plaques with some appearing reticulated and papillomatous over neck, arms, chest, abdomen and back. (**Figure 4**) Systemic examination was normal. Blood sugar levels and thyroid function test were normal. Potassium hydroxide staining of the scale and Periodic acid Schiff stain were negative for fungal elements. He was diagnosed with Confluent and Reticulated Papillomatosis. Patient was initially treated with oral Doxycycline 100mg OD for 1 month, Tretinoin 0.025% cream to be applied on alternate nights and broad-spectrum Sunscreen lotion in morning and afternoon. Patient was advised to keep the skin moisturized and avoid prolonged exposure to the sun. At 1 month follow up, there was significant improvement in the lesions. He was then given Doxycycline 100mg alternate days for 1 month along with the topical agents. At 2 months follow up, there was almost complete clearance of lesions.

2.5. Case 5

A 21-year-old male patient presented with complaints of multiple asymptomatic flat brownish lesions on abdomen, back, thighs, legs and bilateral axilla since the last one and a half months. There was history of similar episodes on and off for the past 2 years which would resolve with treatment. On

examination, there were multiple scaly brown macules and patches with some appearing reticulated on abdomen, back, thighs, legs and bilateral axilla (**Figure 5**) Systemic examination was normal. Blood sugar levels and thyroid function test were normal. Potassium hydroxide staining of the scale and Periodic acid Schiff stain were negative for fungal elements. He was diagnosed with Confluent and Reticulated Papillomatosis. Patient was treated with oral Doxycycline 100mg OD for 1 month & Tretinoin 0.025% cream to be applied on alternate nights. Patient was advised to keep the skin moisturized and avoid prolonged exposure to the sun.

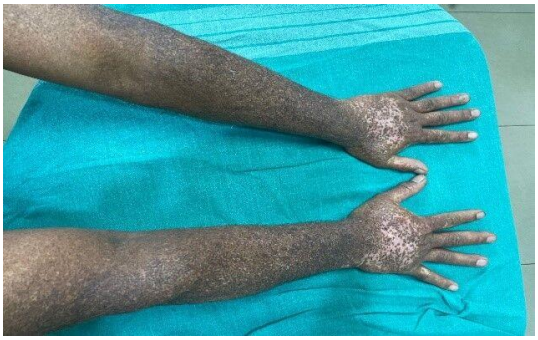

Figure 1: Multiple mottled hyperpigmented and hypopigmented macules and patches over extensor aspects of bilateral forearms and arms

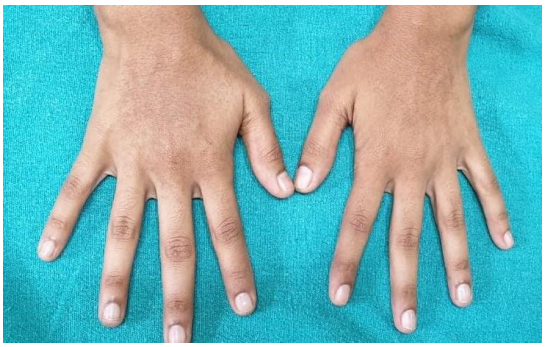

Figure 2: Multiple mottled hyper and hypopigmented macules on dorsum of both hands

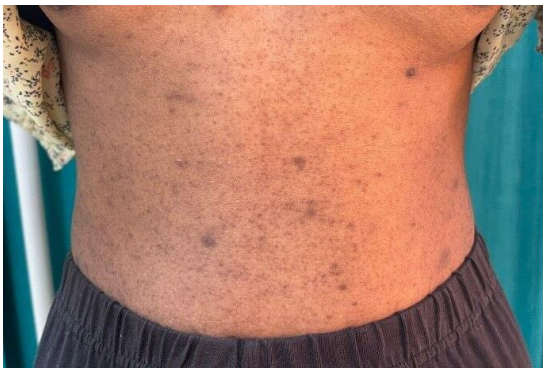

Figure 3: Multiple hyperpigmented macules over chest & abdomen

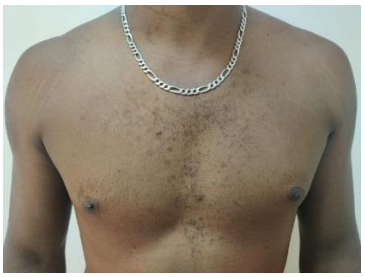

Figure 4: Multiple scaly brown macules and patches with some appearing reticulated and papillomatous on neck, upper chest.

Figure 5: Multiple scaly brown macules and patches with some appearing reticulated on back

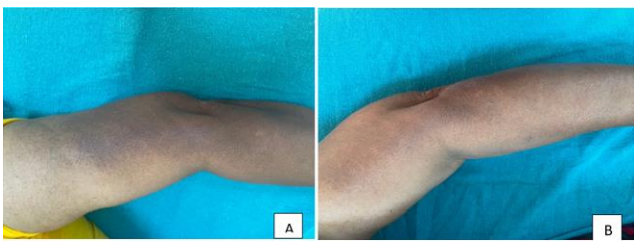

Figure 6: a: Multiple hyperpigmented patches of rippling, greyish-brown macules over extensor aspect of left upper limb**, b:** Marked reduction in hyperpigmented patches over extensor aspect of left upper limb after 4 sessions of FrCO2 laser.

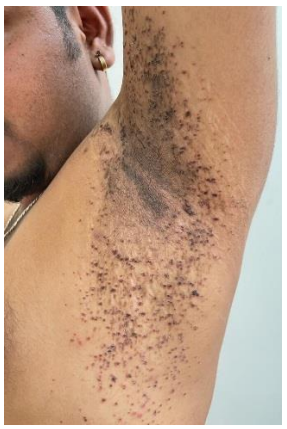

Figure 7: Multiple hyperpigmented, hyperkeratotic papules and plaques with few scales present over dorsum of left axillae

Table 1: The details of patients in this case series are summarized

Patient	Diagnosis	Age/ Sex	Onset of disease	Treatment	Follow up
1	DUH	47/M	Since birth	Hydroquinone 4% cream + Sunscreen	Lost to follow up
2	DUH	21/F	Since birth	Hydroquinone 4% cream + Sunscreen	Lost to follow up
3	Dowling Degos disease	13/F	2 months	Topical steroids, Retinoids and Hydroquinone 4% cream	Mild clearance
4	Confluent and reticulated papillomatosis	28/M	2 years on and off	Doxycycline 100mg and topical Retinoid	Almost Complete clearance
5	Confluent and reticulated papillomatosis	21/M	1.5 months	Doxycycline 100mg and topical Retinoid	Lost to follow up
6	Macular Amyloidosis	65/F	10 years	Fractional CO2 Laser	Almost clearance
7	Darier's disease	29/M	2 months	Oral and topical Retinoid	Lost to follow up

2.6. Case 6

A 65-year-old female presented with complaints of multiple flat lesions over left upper limb for 10 years. There was history of scrubbing the body with pumice stone. There was history of itching sensation. There was no history of inflammatory skin diseases prior to the onset of the lesion. There was no significant past medical and surgical history. There was no history of similar complaints in family. Systemic examination was normal. On examination, there were multiple hyperpigmented patches of rippling, greyish-brown macules over extensor aspect of left upper limb (**Figure 6**a). The patient was diagnosed with Macular Amyloidosis. Patient received four sequential fractional carbon dioxide laser (DERMA INDIA FUTURA RF30) treatment sessions with an interval of four weeks between each session with power- 35% and distance 1.1mm and wavelength-10,600nm. According to the global improvement scale, results were graded to be 4, (**Figure 6**b) which is 76-100% improvement and the patient satisfaction score was graded to be 4, which indicates Excellent satisfaction (>75%).

2.7. Case 7

A 29-year-old male patient presented with complaints of multiple asymptomatic flat and raised brownish lesions on thighs, bilateral axillae, dorsum of legs and gluteal region since 2 years. Lesions resolved with treatment and recurred on stopping treatment. There was history of exacerbation of lesions on sweating. On cutaneous examination, there were multiple hyperpigmented, hyperkeratotic papules and plaques with few scales present over dorsum of both hands, elbows, axillae, lower back, gluteal region and inner aspect of both thighs (**Figure 7**). Systemic examination was normal. Other members of his family were not affected. Patient was diagnosed with Darier's disease and was treated with oral Isotretinoin 30mg OD for 1 month & Tretinoin 0.025% cream to be applied at night. Patient was advised to keep the skin moisturized and avoid prolonged exposure to sun.

3. Discussion

Reticulate pigmentary disorders are a group of diseases characterized by mottled hyperpigmented and/or hypopigmented macules with distinct distribution and varying sizes and amounts of pigment.[1] Various congenital and acquired dermatoses, as well as few systemic diseases present with this pattern of skin lesions. Adya et al have proposed a classification of the reticulate disorders where they divided the disorders on the basis of the etiology.[2] In this case series, seven cases of five distinctive conditions i.e. DUH, Dowling Degos disease, Confluent and Reticulate papillomatosis, macular amyloidosis and Darier's disease; all of which fall under the umbrella of reticulate pigmentary disorders have been described.

3.1. Dyschromatosis universalis hereditarian (DUH)

DUH is an autosomal dominant rare disorder characterized by an early onset of multiple reticulate hyper- and hypopigmented macules which originates from the hands and can progress to involve the trunk, extremities and the face.[3] The condition is more commonly seen amongst Japanese population.[8] Dyschromia first manifests on the trunk before spreading to other parts of the body, including the palms and soles. The general health of the affected individuals is most often normal.[3] The nails are hyperpigmented dystrophic with pterygium formation being the classic finding. Ocular and auditory anomalies, photosensitivity, developmental delay, coxa valga, nerve compression and short stature are less frequently observed, as has been the autosomal recessive mode of inheritance.[1]

Patients have to be reassured that the condition is benign and non-progressive. The pigmented lesion can be targeted with the Q-switched alexandrite laser, but recurrence is inevitable.[1]

3.2. Dowling-degos disease (DDD)

This condition, also known as Dark Dot Disease or Reticular Pigment Anomaly of Flexures, is a progressive pigmentation disorder which start as discrete macules, gradually forming a lace-like or reticulate pattern, initially affecting groins and

axillae and progressing symmetrically to the neck, inframammary creases, trunk, proximal arms, and antecubital fossae. It is inherited as an autosomal dominant trait, with the gene defect localized to kertain.[5] Onset usually occurs in early adulthood (30–40 years). The disease's hallmark features include acneiform perioral pits and comedones. Histologically, the condition is characterized by an atrophic epidermis with "antler-like" intertwining rete ridges, increased melanin in the basal membrane, and dilated follicles with cysts.[1] The patient in this case series presented with hyperpigmented macules, hyperkeratotic papules on various body areas, and characteristic pits on the medial aspect of thighs.

Variations of DDD conditions like Haber's syndrome, Galli-Galli disease, and Pigmentatio Reticularis Faciei et Colli. Haber's syndrome is characterized by pigmented keratotic papules on the axilla, neck, and torso, accompanied by pitted scars on the face and persistent facial redness. Galli-Galli disease is essentially an acantholytic variant of DDD.[9] Pigmentatio Reticularis Faciei et Colli manifests with facial and neck hyperpigmentation along with multiple epidermoid cysts.

As DDD is genetically determined, there is currently no curative treatment. Various topical medications, including azelaic acid, retinoic acid, hydroquinone, and corticosteroids, as well as systemic retinoids, have been attempted with limited success. However, there is a singular report describing the use of Er: YAG laser resulted with positive outcomes. Fractional lasers might be effective for atrophic lesions.[1]

3.3. Confluent and reticulate papillomatosis (CRP)

This condition, named after the French dermatologists Gougerot and Carteaud, is characterized by a diverse clinical presentation. It is more frequently observed in females and begins around the age of 20. The lesions manifest as red, verrucous, and minimally scaly papules, predominantly found in the inframammary, interscapular, and epigastric areas, gradually merging to create brown plaques. Accentuated lesions may also be noticeable in the neck and axillae. As the condition advances, the lesions develop the distinctive reticulate appearance.

3.3.1. Davis et al introduced five criteria for diagnosing CRP

1. Clinical identification of scaling brown macules and patches, some of which exhibit a reticulated and papillomatous appearance.
2. Involvement of the upper trunk and neck.
3. Negative fungal staining, indicating the absence of fungus.
4. Lack of response to antifungal treatment.
5. Remarkable response to minocycline.

3.3.2. Jo et al. in 2014 proposed a change to the criteria

1. Scaly brown macules and patches with some appearing reticulated and papillomatous
2. Involvement of the upper trunk, neck, or flexural areas
3. Negative fungal staining or no response to antifungal treatment
4. Excellent response to antibiotics

In the present case series, patient 4 fulfilled Jo et al criterion for the diagnosis of CRP. The treatment for CRP includes the application of topical tacalcitol and topical calcipotriol, along with oral Minocycline at a dosage of 100–200 mg per day over weeks to months. Other antibiotics that have shown efficacy include clarithromycin (500 mg daily for five weeks), erythromycin (1000 mg daily), and azithromycin (500 mg daily). Additionally, drugs like Isotretinoin and etretinate have proven effective.[10]

3.4. Darier's disease (DD)

Darier disease is an uncommon keratinisation disorder that affects skin and oral mucosa. It is distinguished by seborrheic hyperkeratotic papules and nail abnormalities. DD typically first shows symptoms in childhood or adolescence and has an equal gender distribution. It is associated with a mutated ATP2A2 gene, which encodes for the sarcoplasmic/endoplasmic calcium adenosine triphosphatase (SERCA2). SERCA2 is crucial for regulating cytosolic Ca2+ concentration, which, in turn, governs the assembly of desmosomes. The mutation in ATP2A2 can be inherited in an autosomal dominant manner or occurs sporadically, with two-thirds of cases reported as sporadic. The widely expressed isoform SERCA2b in the epidermis becomes disrupted due to the mutation, leading to the deregulation of intercellular keratinocyte attachment. This results in acantholysis, a characteristic histopathologic feature that includes dyskeratoses, the presence of "corps ronds" in the malpighian layer, and "grains" in the stratum corneum. Corps rods and grains represent apoptotic keratinocytes harboring a mutant SERCA2 that fails to upregulate the p21 WAF1 gene, preventing them from exiting the cell cycle under stress. The persistence of lesions in this case is attributed to this failure to exit the cell cycle.

The mainstay of treatment is oral retinoids. Other modalities like Dermabrasion, electrosurgery, laser ablations of recalcitrant plaques with CO_2, ER: YAG laser have been successfully used.[10]

3.5. Macular amyloidosis (MA)

Macular amyloidosis is a subtype of cutaneous amyloidosis. It has a female preponderance and is identified by hyperpigmented patches, presenting as rippling, greyish-brown macules where amyloid material accumulates in the papillary dermis. Typically, MA emerges on the upper back, arms, chest, and legs. While its precise cause is unknown, certain risk factors have been identified, including the Epstein

Barr virus, ultraviolet Blight exposure, genetic predisposition, atopy, and the use of nylon towels or brushes.[12] Although it's quite common, this condition can lead to both physical discomfort and mental distress due to itching.[13]

Various treatment modalities are available, such as systemic retinoids, potent topical corticosteroids, topical calcineurin inhibitors, PUVA (Psoralen ultraviolet A) and UVB (Ultraviolet B) phototherapy, topical dimethyl sulfoxide (DMSO), cyclosporine, and cyclophosphamide. However, none of these treatments currently achieve complete elimination of amyloidosis lesions, and overall results have not been entirely satisfactory.[12]

Recent evidence suggests that chemical peel using TCA (Trichloroacetic acid) at concentrations of 10%, 20%, and 30%, Glycolic acid 20% along with fractional CO2 laser therapies, prove to be more effective.[14,15]

In this case series we have highlighted, five different types of cases with a common clinical feature of reticulate pigmentation and their management. Although there are several conditions that fall under the umbrella of reticulate pigmentation, some of them are common and easy to diagnose. Few conditions are uncommon and may require certain investigations for the diagnosis. One of the limitations of this case series is that we did not perform skin biopsy for these patients as they were not willing for the procedure and all the cases were diagnosed clinically. Another limitation is that some of the patients were lost to follow up and we were not able to assess the improvement following treatment.

4. Conclusion

In summary, this case series delves into the complexity of reticulate pigmentary disorders, encompassing a spectrum of conditions such as DUH, Dowling-Degos disease, Confluent and Reticulate Papillomatosis, Darier's disease, and Macular Amyloidosis. The diverse clinical manifestations and genetic underpinnings of these disorders highlight the need for a finer approach to diagnosis and management. Conducting molecular genetic testing on the specific causal genes associated with these conditions can aid in establishing a conclusive and precise diagnosis. Despite the challenges posed by limited treatment options for some conditions, the study underscores the importance of understanding and addressing these disorders to enhance patient outcomes.

5. Patients Consent

All patients were informed and written consent was taken from the patient. For any images presented appropriate consent has been obtained from the patients.

6. Source of Funding

None

7. Conflict of Interests

None

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