

Dermatoscopic Evaluation of Nail Patterns in Rare Papulosquamous Skin Disorders: A Diagnostic Bonanza

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ABSTRACT

Aim and objective: To dermatoscopically evaluate the specific nail patterns in rare papulosquamous disorders.

Materials and methods: The present descriptive and observational study was conducted on seven patients with skin biopsy-proven Darier's disease, lichen striatus (LS), and pityriasis rubra pilaris (PRP) selected by stobe randomization. All the patients were first evaluated clinically for nail pattern changes and then dermatoscopy of nails was performed using both wet and dry techniques. The difference in both observations was then analyzed.

Results: The specific nail patterns in Darier's disease were found to be longitudinal and transverse ridging and V-shaped nicking with onycholysis and alternating red and white lines. The nail patterns of LS include nail plate thinning fragmentation with splinter hemorrhages. The PRP patient showed nail plate thickening and distal brown discoloration of the nail plate.

Conclusion: The dermatoscope has come out as a noninvasive useful tool for subsurface, nail bed changes, and chromonychia where clinical examination alone is not sufficient.

Clinical significance: As these rare disorders present with specific nail patterns, knowledge of these nail patterns can aid in diagnosis.

Keywords: Dermatoscopy, Nail, Papulosquamous disorders.

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INTRODUCTION

Nails are the vital organs of the body acting as a window to various acute and chronic systemic diseases. Nail disorder comprises approximately 10% of all dermatological conditions.¹ Papulosquamous skin disorders are group of disorders, which can involve all three parts of the human cutaneous system, namely the skin, the hair, and the nail. These are the disorders that histopathologically show similar features of hyperkeratosis, hypergranulosis, acanthosis and elongation of rete ridges, papillomatosis, and inflammatory infiltrate. The papulosquamous skin disorders are characterized by scaly papules and plaques.² As all are characterized by similar scaly papules and plaques, confusion may result in their diagnosis. Darier's disease, lichen striatus (LS), and pityriasis rubra pilaris (PRP) are the part of the papulosquamous disorder spectrum, which are very rare diseases epidemiologically but present with very typical nail changes that can clear the diagnostic dilemma of these morphologically similar entities. These nail patterns can form a diagnostic algorithm when it comes to overlapping clinical features and can be great aide in establishing the diagnosis. But the irony of the situation is that the nail is capable of mounting only a limited number of reaction patterns to the large number of disorders affecting it and there occurs a great extent of overlap in the nail patterns as well, making specific diagnosis difficult. Although the nail histopathological study is considered the gold standard in diagnosing dermatological lesions, it has its limitations due to invasiveness and quite often a definite "specific" diagnosis is not possible in case of papulosquamous disorders.³ Nail biopsy and diagnostic histopathology are not commonly used for nail diseases; various reasons being apprehensions in the mind of patients and clinicians alike.⁴

Dermatoscopy is a beneficial interface between macroscopic dermatology (i.e., clinical features) and microscopic dermatology

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(i.e., histopathological features).⁵ Dermatoscopy of nails acts as a potential link between naked eye examination (clinical onychology) and nail histopathology, opening up a second front with a potential to prevent an invasive procedure like biopsy.⁶

We carried out the present study to demonstrate the beneficial role of nail dermatoscopy in these three rare conditions: Darier's disease, LS, and PRP, which present with clinically similar-looking morphology and hence specific findings in nail patterns can help to better evaluate the disease, thereby contributing toward the confirmation of diagnosis, assessment of prognosis, and decisions on treatment.

MATERIALS AND METHODS

The present descriptive and observational study was conducted on seven patients with skin biopsy-proven Darier's disease, LS, and PRP selected by stobe randomization, during the period of January 2018–September 2019. An approval from the institutional ethics committee was taken beforehand. An informed consent was

taken from each patient. All skin biopsy-proven cases of specific papulosquamous disorder including Darier's disease, LS, and PRP with clinically appreciable nail changes between the age group of 18 and 70 years were enrolled in the study. A detailed history of all the patients was taken in terms of demographic details. Local skin and nail clinical examination was done to see morphology of nail patterns. It was followed by dermatoscopic examination of all the nails to see morphological changes in nail patterns and capillary examination to examine all 20 nails. The DermLite DL4 Dermatoscope with polarized mode and magnification of 10x was used for both wet and dry dermatoscopy. The nonpolarized mode was used for nail plate examination to see for the nail plate surface changes. In few cases, Giemsa stain was used to stain the edges of the pits. The polarized mode was used for nail bed examination and color changes. The linkage fluid used was gel-based hand sanitizer. All the nails were examined in a sequence of dermatoscopic examination of proximal nail folds, lateral nail folds, lunula, nail plate surface changes, and nail bed changes. The photographs of the findings were taken and recorded. The results were analyzed statistically using the SPSS Software 19.0 version. The chi-square test, the Wilcoxon signed-rank test, and one-way ANOVA stats were used for comparing different variables and making correlations.

RESULTS

The mean age group was 21.25 years with SD of 12.6. Majority of patients (75%) were of the age group between 18 years and 25 years. Out of seven patients, two were males and four females. Mean duration of symptoms was 5 years with SD 2.1 years. About 75% of patients had cutaneous symptoms for more than 4 years.

Out of total four patients of Darier's disease, nail matrix changes included longitudinal ridging, transverse ridges, and V-shaped nicking. The comparison of clinical and dermatoscopic findings is shown in Figure 1. In the nail bed changes, onycholysis, subungual hyperkeratosis, splinter hemorrhages, and alternating red and white lines were seen. These patterns were observed first

clinically and then with dermatoscope and the observations were compared (Fig. 2).

Out of two patients of LS, nail matrix changes observed were longitudinal ridging, transverse ridging, nail plate thinning and fragmentation, leukonychia, and longitudinal melanonychia. Splinter hemorrhages were seen only dermatoscopically in both the patients. The comparison of both clinical and dermatoscopic findings is shown in Figure 3.

A single patient of PRP showed nail plate thickening and transverse ridges but pitting was appreciated only dermatoscopically. Onycholysis, subungual hyperkeratosis, and brown discolored nail plate were appreciable both clinically and dermatoscopically as shown in Figure 4.

DISCUSSION

Not much work has been done on dermatoscopy of nail changes in these rare papulosquamous disorders. The nail dermatoscopic characteristics have been described in only few studies till now and there have been no attempts to correlate them with clinical features.

The first rare papulosquamous disorder observed was Darier's disease (Fig. 5). Only four biopsy-proven patients could be included in the study. The mean age group was 21.25 years with SD of 12.6. Majority of patients (75%) were of the age group between 18 years and 25 years. Out of four patients, three were males and one female. Mean duration of symptoms was 5 years with SD 2.1 years. About 75% patients had cutaneous symptoms for more than 4 years. In our study, we found that finger nails were involved more often than toe nails. Puri et al. conducted a study on 20 patients of Darier's disease and found that the commonest age group of patients was 11–20 years (50% patients); 33.3% patients were between 21 years and 30 years, which is in accordance with our study.⁷

In the nail matrix changes, longitudinal ridging was seen in all the patients (100%) both clinically and dermatoscopically, and V-shaped nicking (Fig. 6) was also seen in 100% patients, which is highly pathognomic for Darier's disease. Presence of

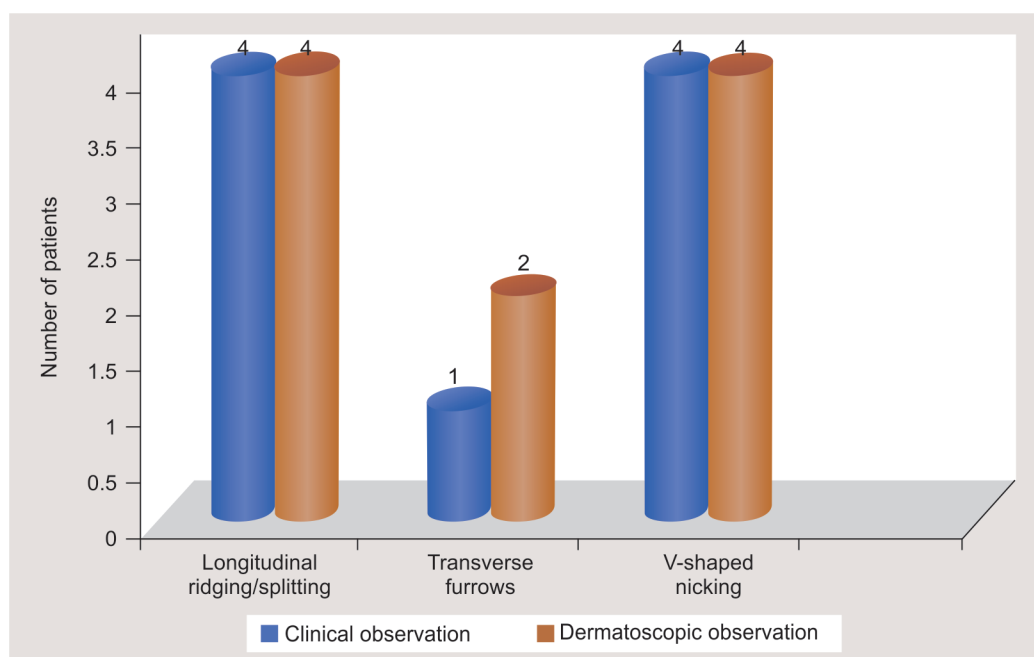


Fig. 1: Comparison of clinical and dermatoscopic observations of nail matrix changes in Darier's disease

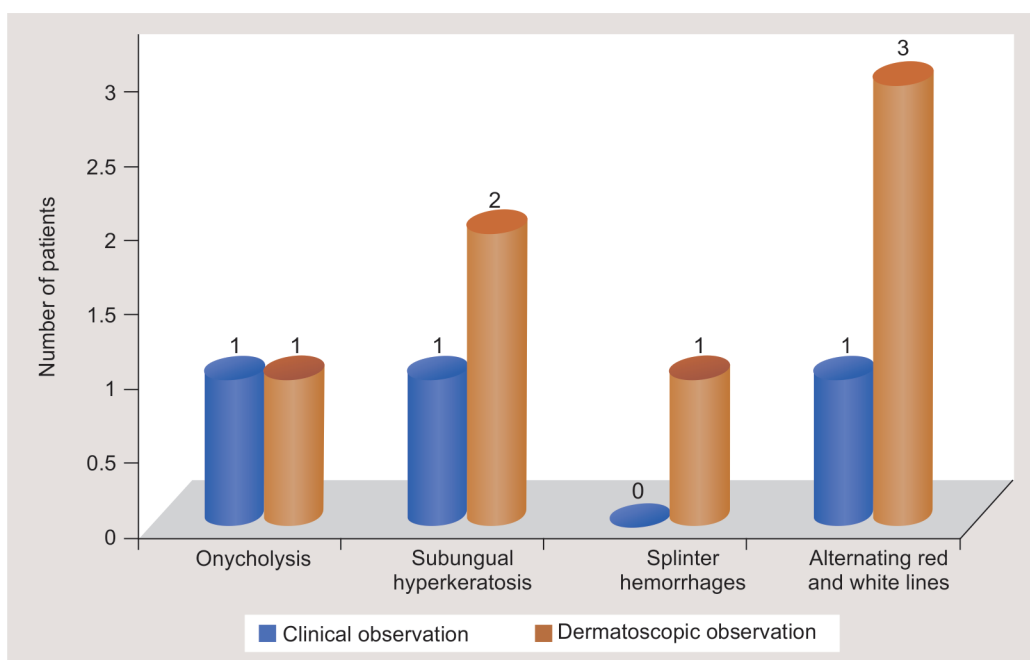


Fig. 2: Comparison of clinical and dermatoscopic observations in nail bed changes in Darier's disease

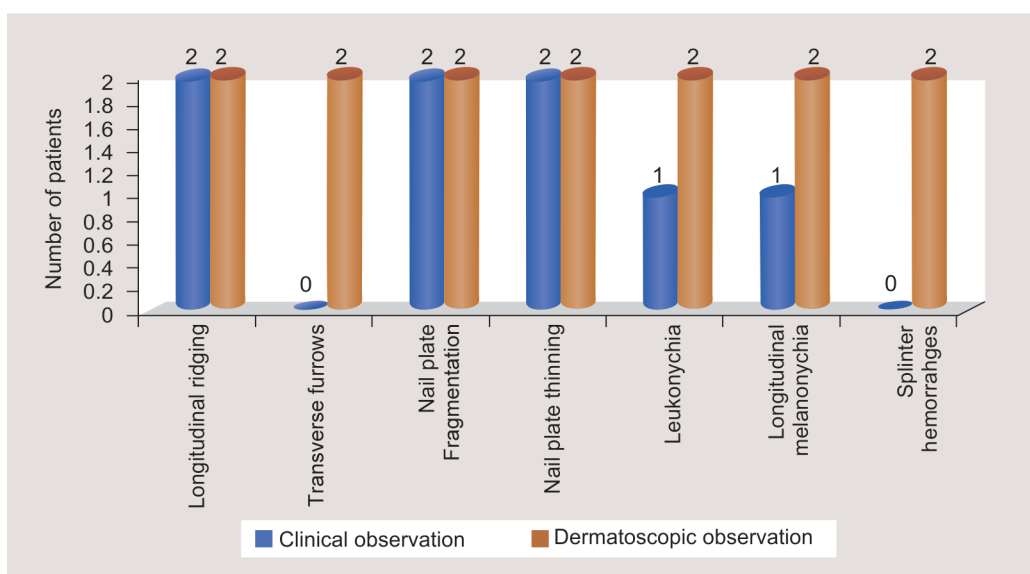


Fig. 3: Comparison of clinical and dermatoscopic observations in nail bed changes in lichen striatus

V-shaped nicking was only seen in Darier's disease among various papulosquamous disorders, thus being statistically significant. This signifies that V-shaped nicking can be a deciding factor for making a diagnosis of Darier's disease in case of diagnostic dilemma. Transverse ridges were appreciated in 25% patients clinically and 50% patients dermatoscopically. Pitting was seen in few patients as well (Fig. 7).

In the nail bed changes, onycholysis was appreciated in 25% patients both clinically and dermatoscopically. Subungual hyperkeratosis was seen in 25% patients clinically but later with dermatoscopy it was appreciated in 50% patients. Splinter hemorrhages were seen only with dermatoscope in 25% patients. Alternating red and white lines in nail bed (Fig. 8) were seen clinically

in 25% patients but with dermatoscope they could be appreciated in 75% patients. Hence, V-shaped nicking and alternating red and white lines are two most pathognomonic features of nail changes in Darier's disease whose observation is statistically better with dermatoscope than clinical examination. In the study conducted by Puri et al. on 20 patients of Darier's disease, they found alternating red and white bands in all the patients (100%) and V-shaped nicking at the free margins of nails in 93.3% patients, which is consistent with our study.⁷ In a case report published by Mahboob et al., they found that in their patient, all nails showed light and dark longitudinal bands, distal nail plate splitting, scalloping, and V-shaped notch was present in one of the nails.⁸ In another study conducted by Zaias et al., they concluded that longitudinal ridges,

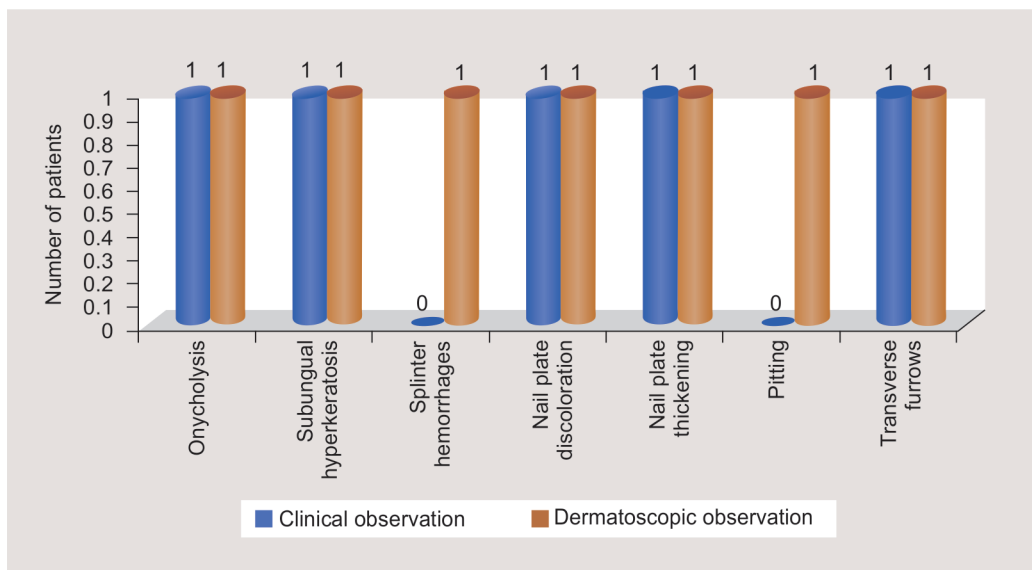


Fig. 4: Comparison of clinical and dermatoscopic observations in nail bed changes in pityriasis rubra pilaris



Fig. 5: Greasy papular eruptions in Darier's disease

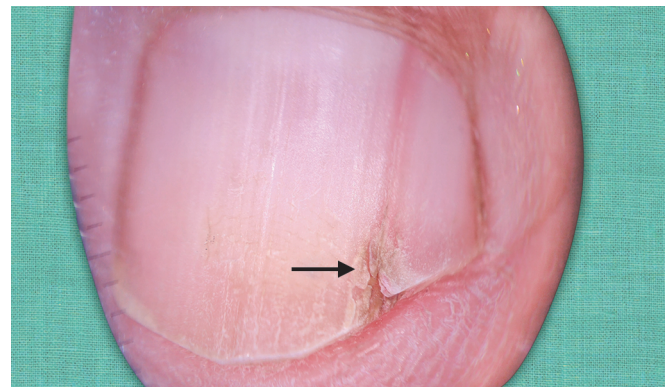


Fig. 6: V-shaped nicking in Darier's disease

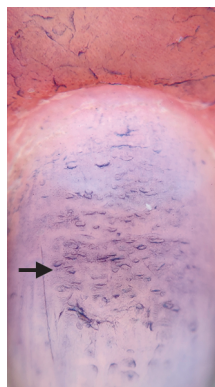


Fig. 7: Pitting in Darier's disease

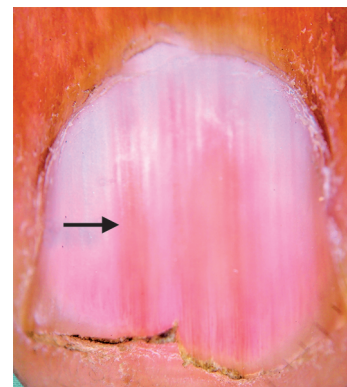


Fig. 8: Alternating red and white lines in Darier's disease

subungual hyperkeratosis, red or white streaks, or both, associated with distal wedge-shaped subungual keratoses, are the nail signs diagnostic for Darier-White disease.³ These findings were consistent with our findings.

The next rare disease among papulosquamous disorders with nail changes encountered was LS (Fig. 9). Two patients were enrolled

in the study. One patient was of 18 years, but interestingly the second patient was 52-years-old. Both were female patients (100%). Patrizi et al. in their study reported a female predominance with the male to female ratio of 0.5:1.⁹ The similar female preponderance was recorded in the study by Taniguchi et al.¹⁰ Finger nails were involved more than toe nails in 100% of patients.



Fig. 9: Linear scaly plaques over leg in lichen striatus



Fig. 10: Longitudinal ridges in lichen striatus

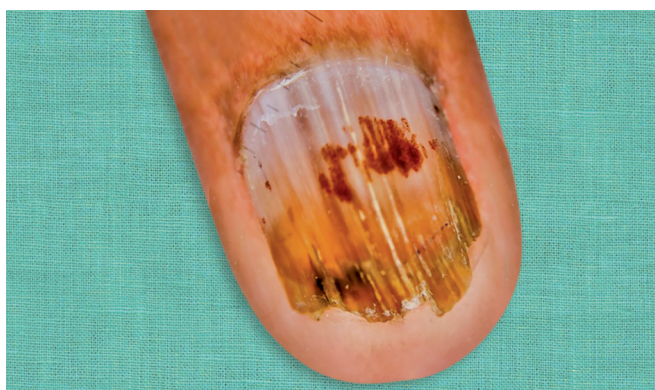


Fig. 11: Nail plate thinning and fragmentation in lichen striatus



Fig. 12: Hand involvement in pityriasis rubra pilaris

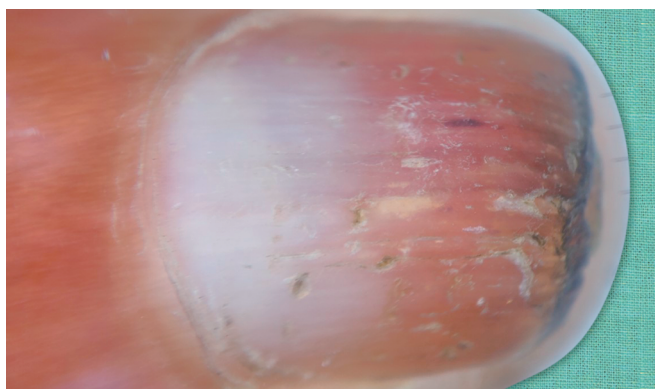


Fig. 13: Distal brown discoloration and thickening of nail plate in pityriasis rubra pilaris

Longitudinal ridging (Fig. 10) was seen clinically and dermatoscopically in 100% of the patients, whereas transverse ridging was seen only dermatoscopically in 100% of the patients but not appreciated clinically in any of the patients. Nail plate thinning and fragmentation was an important feature observed in 100% of the patients (Fig. 11). Leukonychia and longitudinal melanonychia were seen clinically in one patient and only dermatoscopically in the other patient. Splinter hemorrhages were seen in 100% patients but only dermatoscopically not appreciated clinically.

Nail changes in LS have been reported in various case reports only. No case series study has ever been done on LS involving nails.

In various case reports, it has been mentioned that nail changes can appear after the skin lesions, which finding is consistent with our study. In a case report by Palleschi et al. on a 27-year-old female patient, the nail changes appreciated were longitudinal ridging and thinning of nail plate.¹¹ In a study published by Krishnegowda et al., four cases of LS with nail involvement were studied. There were several types of nail dystrophy reported: fraying, longitudinal ridging, splitting and shredding, onycholysis, and total nail loss.¹² In a case report of seven patients of nail LS, Kim et al. reported the most common nail changes in LS were fissuring, longitudinal ridging, splitting, and shedding, which was consistent with our study.¹³

In our study, one patient of PRP was also recruited (Fig. 12). The incidence of PRP in India is 1 in 50,000 in an outpatient setting. Sehgal et al. in their study described that both the sexes were affected equally at all ages.¹⁴ A bimodal or trimodal age distribution has been recorded with peak incidence in the first, second, and sixth decades of life. Our patient was 25 years' old female, housewife by occupation. The duration of illness in our patient was 2.5 years, which is in accordance with the above study.

In our study, the nail changes observed were nail plate thickening (Fig. 13) with subungual hyperkeratosis (Fig. 14). Onycholysis was present along with nail plate pitting. Few transverse ridges were also seen. The nails showed brown discoloration of nail plate with splinter hemorrhages. Out of these, all findings were seen clinically as well as dermatoscopically except pitting and splinter hemorrhages, which were only appreciated dermatoscopically. Sehgal et al. described nail changes in PRP as nail thickening and yellow-brown discoloration of the nail plate,

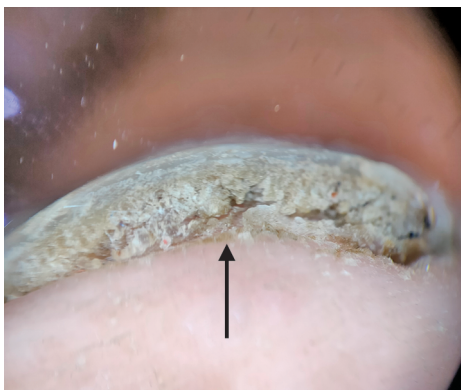


Fig. 14: Subungual hyperkeratosis in pityriasis rubra pilaris

subungual hyperkeratosis, and splinter hemorrhages, which were also appreciated in our study.¹⁴

In a case report by Lambert et al. on PRP in a 20-year-old girl, they found the nails were thickened, opaque, and painful, with distal onycholysis and yellow-brown discoloration and subungual hyperkeratosis, which was consistent with our study.¹⁵

In the article by Sonnex et al.,¹⁶ the authors stated that a change in the morphology of the fingernails may be helpful in distinguishing PRP from psoriasis. Distal yellow-brown discoloration, subungual hyperkeratosis, nail plate thickening, and splinter hemorrhage, which were indicative of nail plate disease, suggest the diagnosis of PRP, which was consistent with our findings.^{7,8}

As we could just recruit one patient of PRP, it would not be possible to fully describe the nail patterns found in all types of PRP. A large data of patients are required for the same, which is not possible as the disease is quite rare in India.

CONCLUSION

The three rare conditions, Darier's disease, LS, and PRP, described in this study have similar morphological appearances to other papulosquamous disorders like psoriasis, lichen planus, and seborrheic dermatitis, which often leads to misdiagnosing these entities. In these situations, the nails can open the diagnostic window in the patients helping the clinician to arrive at a specific diagnosis. The knowledge of specific nail patterns in these disorders gives a diagnostic clue. But as per our observations, mere naked eye examination of the nails can miss the finer details of nail plate and pathognomonic nail bed signs where the use of dermatoscope has been proved to be most significant as a noninvasive tool that helps in better visualization of surface and subsurface. Histopathology is a gold standard technique to confirm a diagnosis, but is an invasive procedure. Though dermatoscope is not a confirmatory tool, it definitely aids in establishing a diagnosis and avoiding the invasive procedures.

CLINICAL SIGNIFICANCE

These specific nail patterns in the clinically similar conditions can guide the clinician in timely diagnosis without the need of skin and nail biopsy and can help in early management, thus avoiding sequelae. Dermatoscope helps to visualize subsurface changes like splinter hemorrhages, leukonychia, melanonychia, and alternating red lines better than clinical examination alone.

REFERENCES

- De Berker DA, Baran R, Dawber RPR. Disorders of nails. In: Burn T, Breathnach S, Cox N, et al., ed. *Rook's Textbook of Dermatology*. 7th ed., Blackwell: John Wiley & Sons; 2004. p. 62.
- Hosamane S, Pai M, Philipose TR, et al. Clinicopathological study of non-infectious erythematous papulosquamous skin diseases. *J Clin Diagn Res* 2016;10(6):19–22. DOI: 10.7860/JCDR/2016/18607.8029.
- Zaias N, Ackerman AB. The nail in Darier-white disease. *Arch Dermatol* 1973;107(2):193–199. DOI: 10.1001/archderm.1973.01620170005001.
- Grover C, Chaturvedi UK, Reddy BS. Role of nail biopsy as a diagnostic tool. *Indian J Dermatol Venereol Leprol* 2012;78(3):290–298. DOI: 10.4103/0378-6323.95443.
- Grover C, Jakhar D. Onychoscopy: a practical guide. *Indian J Dermatol Venereol Leprol* 2017;83(5):536–549.
- Nakamura RC, Costa MC. Dermatoscopic findings in the most frequent onychopathies: descriptive analysis of 500 cases. *Int J Dermatol* 2012;51(4):483–485. DOI: 10.1111/j.1365-4632.2010.04720.x.
- Puri N. A clinical and histopathological study of Darier's disease. *J Pak Assoc of Derm* 2016;21(4):230–234.
- Mahboob A, Yaqub F, Shahzad Z, et al. Classical presentation of Darier's disease: a rare disorder of keratinisation. *J Ayub Med Coll Abbottabad* 2010;22(3):230–233.
- Patrizi A, Neri I, Fiorentini C, et al. Lichen striatus: clinical and laboratory features of 115 children. *Pediatr Dermatol* 2004;21(3):197–204. DOI: 10.1111/j.0736-8046.2004.21302.x.
- Taniguchi Abagge K, Parolin Marinoni L, Giraldo S, et al. Lichen striatus: description of 89 cases in children. *Pediatr Dermatol* 2004;21(4):440–443. DOI: 10.1111/j.0736-8046.2004.21403.x.
- Palleschi GM, D'Erme AM, Lotti T. Lichen striatus and nail involvement: truly rare or question of time? *Int J Dermatol* 2012;51(6):749–750. DOI: 10.1111/j.1365-4632.2010.04614.x.
- Krishnegowda SY, Reddy SK, Vasudevan P. Lichen striatus with onychodystrophy in an infant. *Indian Dermatol Online J* 2015;6(5):333–335. DOI: 10.4103/2229-5178.164489.
- Kim M, Jung HY, Eun YS, et al. Nail lichen striatus: report of seven cases and review of the literature. *Int J Dermatol* 2015;54(11):1255–1260. DOI: 10.1111/ijd.12643.
- Sehgal VN, Srivastava G, Dogra S. Adult onset pityriasis rubra pilaris. *Indian J Dermatol Venereol Leprol* 2008;74(4):311–321. DOI: 10.4103/0378-6323.42889.
- Lambert DG, Dalac S. Nail changes in type V pityriasis rubra pilaris. *J Am Acad Dermatol* 1989;21(4):811–812. DOI: 10.1016/S0190-9622(89)80284-1.
- Sonnex TS, Dawber RP, Zachary CB, et al. The nails in adult type I pityriasis rubra pilaris: a comparison with Sézary syndrome and psoriasis. *J Am Acad Dermatol* 1986;15(5):956–960. DOI: 10.1016/S0190-9622(86)70256-9.