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CASE REPORT

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Mimicry unveiled: The challenging diagnosis of pigmented purpura-like mycosis fungoides initially misdiagnosed as pigmented purpura

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Key Clinical Message

Unlike most cases, the lesions were localized to the dorsum of the hand, lacked pruritus (itching), and did not exhibit "sperm-like blood vessels," which are typically pathognomonic to classical MF.

Abstract

The study presents a rare case involving a 44-year-old woman who developed a skin condition on the base of her left thumb. Initially misdiagnosed as pigmented purpura, the need for further investigation arose to determine the nature of the condition accurately. The medical evaluation encompassed a comprehensive analysis of the patient's skin ailment. A series of diagnostic examinations were conducted to ascertain the underlying cause. Although routine blood tests yielded unremarkable results, the distinct characteristics of the rash prompted a more thorough investigation. Subsequent assessment revealed that the skin condition was not pigmented purpura, as initially presumed, but rather a manifestation of cutaneous T-cell lymphoma (CTCL) known as mycosis fungoides (MF). MF is an infrequent lymphoma predominantly affecting individuals aged 45–65, exhibiting a male-to-female sex ratio of 2:1. The annual incidence of MF ranges from 0.3 to 0.96 cases per 100,000 individuals. The woman's skin exhibited discrete patches adorned with colored dots, progressively thickening and pigmentation. Notably, the absence of pruritus did not dispel suspicion. This case underscores the significance of accurately diagnosing uncommon dermatological disorders to facilitate appropriate medical intervention. The unique appearance of the rash and its distinctive features, despite normal blood results, enabled the identification of MF. The patient's treatment encompassed a combination of steroids and narrowband UV therapy. Vigilance, continued research, and heightened awareness are paramount for early intervention and improved patient outcomes. Such efforts contribute to an enhanced understanding of the complexities of this condition.

KEYWORDS

mimicking dermatoses, mycology, mycosis fungoides, pigmented purpura, purpura

1 | INTRODUCTION

Among CTCLs, mycosis fungoides (MF) is the most common. It is a kind of cutaneous lymphoma that develops from memory T cells (CD45RO+), which are peripheral epidermotropic T cells that express the T-cell receptor (TCR) and the CD4+ immunophenotype.[1] The French dermatologist Jean-Louis-Marc Alibert first described it in 1806.[2] With a male to female sex ratio of 2:1, the annual incidence ranges between 0.3 and 0.96 instances per 100,000 people and mainly affects patients between the ages of 45 and 65.[3] Childhood and adolescent MF cases are more exceptional.[4]

Although the cause of MF is unknown, genetic, environmental, and viral variables, such as infection with the human T-cell leukemia virus 1 (HTLV-1), have been suggested as potential causes leading to lymphocyte activation or transformation.[5,6] Early identification and treatment of MF are crucial because research has shown that a delayed diagnosis is linked to disease progression and a worse long-term prognosis.[4]

MF is treated according to its stage. Therapies for the early stages include topical corticosteroids, phototherapy, topical bexarotene, irradiation, and preparations of nitrogen mustard. Bexarotene, histone deacetylase inhibitors, interferon, antibody treatments, systemic chemotherapy, and allogeneic hematopoietic cell transplantation (HCT) are alternatives for advanced stages.[7] Male gender, advanced age, and high LDH are all indicators of a poor prognosis.[8]

2 | CASE HISTORY

A 44-year-old female presented at our hospital on June 6, 2023. Her chief complaint was a rash at the base of the thumb of her left hand, characterized by petechial patches with pigmentation and lichenification (Figure 1A). The eruption showed no involvement of the trunk or proximal extremities. She neither experienced pain nor itching. The patient had no history of systemic illness or lymphadenopathy. She has no history of smoking, alcohol, or illicit drug abuse. She did not give any history of exposure to environmental factors like pesticides and toxins. Vital signs were within normal limits, and the physical examination revealed the aforementioned rash only confined to the fingers (Figure 1C).

3 | METHOD

Dermoscopy revealed fine, short linear vessels, dotted vessels, and orange-yellow patchy areas within the eczematous rash (Figure 1B,D). The patient had been misdiagnosed for 2 years, visiting different hospitals and using various medications. The initial misdiagnosis was lymphoproliferative disease. A comprehensive blood investigation, including CBC, RBC, WBC, platelets, RFT, LFT, LDH, and serology for HTLV, HBC, and HBsAg, all yielded normal results.

Biopsy and immunohistochemistry were performed. The biopsy findings revealed irregular epidermal hyperplasia, extensive lymphocyte infiltration in the superficial dermis, focal basal cell edema and degeneration, Pautrier's microabscesses, liquefaction of the basal layer and visible red blood cell overflow (Figure 2A,B). T-lymphocyte proliferation was evident in histochemistry, with CD3+ T cells (Figure 2C) and CD4+ T cells (Figure 2D), minor CD20+ B cells (Figure 2F), Ki-67+ cells (10%), CD8+ T cells (Figure 2E), TlA1 (Figure 2G), and EBER-negative cells. EVG staining showed broken

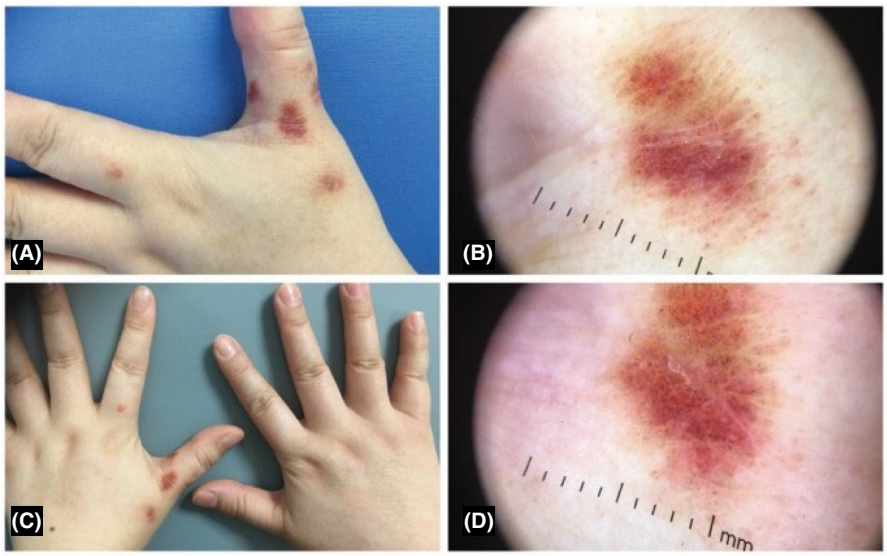

FIGURE 1 (A, C) Gross photograph showing a close-up view of the eczema-like lesion located in the left thumb; (B, D) Dermoscopic examination displaying delicate, abbreviated linear vessels, dotted vascular structures, and areas of orange-yellow patchiness within the context of the rash.

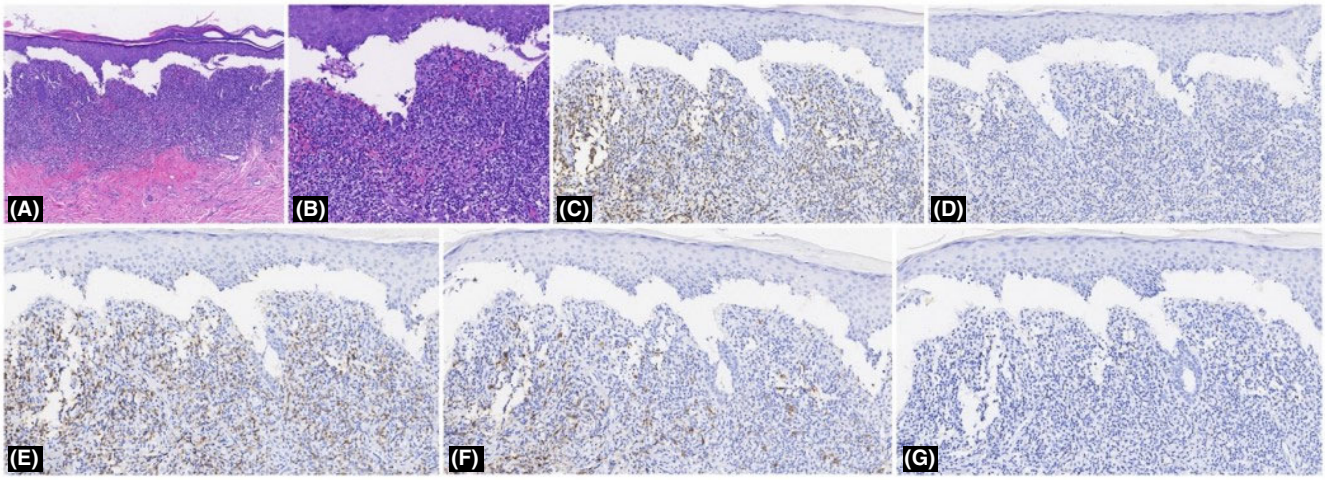

FIGURE 2 Comparison of immunohistochemical and special stain findings reveals T-lymphocyte proliferation and cellular composition in pigmented purpura-like mycosis fungoides lesion on (A) HE Staining, (B) CD3+T cells, (C) CD4+T cells, (D) CD8+ T cells, (E) CD20+ B Cells, (F) TIA1, and (G) TIA1-10X.

or missing elastin fibers in the dermis layers, and special staining for acid-fast was negative. The patient was treated with lincomycin lidocaine gel (20 g twice daily), and mometasone furfurate alosone (15 g once daily) through external application and narrow band UV therapy was also commenced. The patient's rash improved after treatment was followed up closely in the later period. There was not any recurrence seen as such.

4 | CONCLUSIONS

In conclusion, this case report underscores the challenges associated with diagnosing pigmented purpura-like MF due to its overlapping clinical and histological features with other dermatological conditions. The presented case deviates from conventional presentations, with lesions localized to the dorsum of the hand and lacking pruritus, a common symptom inMF. Moreover, in the dermoscopy findings, lesions did not have "sperm-like blood vessels," which is pathognomonic to classical MF. Continued research and clinical awareness are essential for improving early detection, enabling timely intervention, and ultimately enhancing patient outcomes. This case serves as a valuable addition to the medical literature and contributes to the understanding of pigmented purpura-like MF as a complex variant of cutaneous T-cell lymphoma (CTCL).

In contrast to most cases of MF, our patient (a) presented with the lesion on the dorsum of the hand at the base of the thumb at the metacarpophalangeal joint and interphalangeal joint and (b) had lesions that were not associated with pruritus, unlike other cases and (c) does not have sperm-like blood vessels which are pathognomonic to classical MF.

5 | DISCUSSION

Pigmented purpura, like MF, is a rare condition characterized by an overgrowth of cancerous T cells in the skin. It frequently manifests as a range of skin lesions that might mirror those of other dermatological disorders, leading to challenges in diagnosis. Pigmented purpura, like MF, is difficult to diagnose in its early stages because the symptoms and skin biopsy findings are similar to those of other skin conditions.[5,9] This case report highlights a Chinese female who presented with a rash on the dorsum of her left hand, which was initially misdiagnosed as pigmented purpura.

Although the patches and plaques of MF can mimic many other dermatoses (such as psoriasis, dermatitis, and dermatophytosis), there have been few instances of patients presenting with lesions that closely resemble the clinical appearance of pigmented purpuric dermatosis (PPD).[10] The pigmented purpura-like variation of MF initially may have the histologic appearance of pigmented purpura and generally progresses to the histological appearance of MF with time, similar to the numerous cases of early MF that are histologically indistinguishable from dermatitis.

According to recent research, pigmented purpuric eruption may be a precursor to MF, proceed to CTCLover time, or, very rarely, the two disorders may coexist.[11] Many experts support the view that PPD falls under the category of CTCL and is a type of T-cell dyscrasia.[12] Considerable overlap is observed between the features of CTCL and PPD.

Similar to our case, there have been examples of PPD being clinically and histologically diagnosed before MF was histologically diagnosed. Guitart and Magro[13] pointed

out a number of chronic illnesses, including PPD, that are connected to T-cell clones. These ailments frequently did not fit the diagnostic criteria for MF, manifested without a clear cause, and were resistant to topical treatments. Barnhill and Braverman 5 initially showed the transition of PPD to MF over 12 years in a small cohort study of three young males. With a mean age of 24.3 years, the age of onset ranged from 14 to 30 years. With an average latency to diagnosis of 8.4 years, biopsies in all three individuals were consistent with PPD for many years prior to the diagnosis of MF.[14]

Atypical of most cases of pigmented purpura like MF, the patients in this study demonstrated lesions that were confined to the dorsum of the hand, with no involvement of the trunk or proximal extremities. Moreover, it is not associated with pruritus as it is evident that pruritus is the most often reported symptom of patients with MF, with up to 88% of people describing it in varied degrees of severity and typically getting worse as the condition worsens.[15] This unique presentation posed a diagnostic challenge due to its similarity to pigmented purpura.

Lesions frequently start on the body's trunk in areas like the buttocks that are rarely exposed to sunlight.[10] These lesions first appear as small patches and remain undiagnosed for up to 10 years. MF should be differentiated from benign and malignant conditions that report similar clinicopathologic features. Several T-cell lymphomas and rarely B-cell lymphomas can display epidermotropic infiltrates. Still, MF should also be distinguished from various benign inflammatory conditions, including lymphomatoid drug eruptions, lichenoid keratosis, lymphomatoid eczematous dermatitis, and the inflammatory stage of lichen sclerosus.[16] Very rarely, other lymphoproliferative disorders such as Castleman's disease or Castleman-like conditions must be considered in the differential diagnosis of MF, especially when multicentric or an anomalous pattern is manifested.

The diagnosis is difficult, especially in the early stages and is confirmed by a skin biopsy as it mimics the pigmented purpura. The histopathological features observed in the biopsy and the dermoscopic findings described do not support the diagnosis of typical MF. Pautrier's micro abscesses[5] and basal alignment of neoplastic lymphocytes is a significant differentiating factor between MF-positive and non-MF cases. This feature was present in 86.8% of the studied MF cases.[17] Contrary to this, the dermoscopic findings of this case are not typical because it does not have “sperm-like blood vessels,” while sperm-like blood vessels are unique to classical MF. PP, like MF is characterized by hemosiderin laden histiocytes, brown dots and globules which may result from the spherical or elliptical arrangement of melanocytes or melanophages at the dermal–epidermal junction.[18]

The first-line treatment for MF that is most often advised is psoralen with ultraviolet A (PUVA) therapy. Light therapy, ultraviolet light (primarily NB-UVB 312 nm), topical and systemic chemotherapies, local superficial radiotherapy, the histone deacetylase inhibitor vorinostat, total skin electron radiation, photopheresis, systemic therapies (e.g., retinoids and rexinoids), and biological therapies (e.g., interferons) are among the recommended treatments. Combinations of treatments are frequently employed (valipour). It is advised to start treatment with topical and skin-directed treatments before moving on to more systemic medicines due to the potential side effects of treatment alternatives in early illness.[7] Similarly, our patient was initiated on topical treatment with the combination of lincomycin lidocaine gel and mometasone furfurate alosone. Along with this, narrowband UV therapy was also initiated for better outcomes. We believe that this case will provide valuable insights into the literature on early and accurate diagnosis of MF.

AUTHOR CONTRIBUTIONS

Sabita Aryal: Conceptualization; data curation; formal analysis; methodology; software; writing – original draft. **Jiang Zhu-qian:** Data curation; formal analysis; investigation; methodology; software; writing – review and editing. **Liu ye Qiang:** Funding acquisition; methodology; project administration; resources; supervision; validation; writing – review and editing. **Md Ariful Haque:** Data curation; formal analysis; investigation; methodology; software; validation; visualization; writing – original draft.

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CONFLICT OF INTEREST STATEMENT

All authors have completed the ICMJE uniform disclosure form. The authors have no conflicts of interest to declare.

DATA AVAILABILITY STATEMENT

Data sharing is not applicable to this article as no new data were created in this study.

ETHICS APPROVAL

Not applicable.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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REFERENCES

1. Hossain C, Jennings T, Duffy R, et al. The histological prevalence and clinical implications of folliculotropism and syringotropism in mycosis fungoides. *Chin Clin Oncol*. 2019;8(1):6.
2. Alibert JL. Description des Maladies de la Peau: Observees l'Hospital St. Louis et Exposition des Meilleurs Methods Suiview pour leur Traitement. Paris. In: Barrois l'aine et Fils. 1806.
3. Dobos G, Pohrt A, Ram-Wolff C, et al. Epidemiology of cutaneous T-cell lymphomas: a systematic review and meta-analysis of 16,953 patients. *Cancers*. 2020;12:E2921. doi:10.3390/cancers12102921
4. Wu JH, Cohen BA, Sweren RJ. Mycosis fungoides in pediatric patients: clinical features, diagnostic challenges, and advances in therapeutic management. *Pediatr Dermatol*. 2020;37:18-28. doi:10.1111/pde.14026
5. Akinbami AA, Osikomaiya BI, John-Olabode SO, et al. Mycosis fungoides: case report and literature review. *Clin Med Insights Case Rep*. 2014;7:95-98. doi:10.4137/CCRep.S15724
6. Girardi M, Heald PW, Wilson LD. The pathogenesis of mycosis fungoides. *N Engl J Med*. 2004;350(19):1978-1988.
7. Valipour A, Jäger M, Wu P, Schmitt J, Bunch C, Weberschock T. Interventions for mycosis fungoides. *Cochrane Database Syst Rev*. 2020;2020(7):CD008946. doi:10.1002/14651858.CD008946
8. Lebowitz E, Geller S, Flores E, et al. Survival, disease progression and prognostic factors in elderly patients with mycosis fungoides and Sézary syndrome: a retrospective analysis of 174 patients. *J Eur Acad Dermatol Venereol*. 2019;33(1):108-114.
9. Pope E, Weitzman S, Ngan B, et al. Mycosis fungoides in the pediatric population: report from an international childhood registry of cutaneous lymphoma. *J Cutan Med Surg*. 2010;14(1):1-6.
10. Hanna S, Walsh N, D'Intino Y, Langley RGB. Mycosis fungoides presenting as pigmented purpuric dermatitis. *Pediatr Dermatol*. 2006;23:350-354.
11. Georgala S, Katoulis AC, Symeonidou S, Georgala C, Vayopoulos G. Persistent pigmented purpuric eruption associated with mycosis fungoides: a case report and review of the literature. *J Eur Acad Dermatol Venereol*. 2001;15:62-64.
12. Sarantopoulos GP, Palla B, Said J, et al. Mimics of cutaneous lymphoma: report of the 2011 Society for Hematopathology/European Association for Haematopathology workshop. *Am J Clin Pathol*. 2013;139:536-551.
13. Guitart J, Magro C. Cutaneous T-cell lymphoid dyscrasia: a unifying term for idiopathic chronic dermatoses with persistent T-cell clones. *Arch Dermatol*. 2007;143:921-932.
14. Barnhill RL, Braverman IM. Progression of pigmented purpura-like eruptions to mycosis fungoides: report of three cases. *J Am Acad Dermatol*. 1988;19(1 Pt 1):25-31.
15. Ottevanger R, van Beugen S, Evers AW, Willemze R, Vermeer MH, Quint KD. Quality of life in patients with mycosis Fungoides and Sézary syndrome: a systematic review of the literature. *J Eur Acad Dermatol Venereol*. 2021;35(12):2377-2387. doi:10.1111/jdv.17570
16. Whittaker S, Hoppe R, Prince HM. How I treat mycosis fungoides and Sézary syndrome. *Blood*. 2016;127:3142-3153.
17. Naraghi ZS, Seirafi H, Valikhani M, Farnaghi F, Kavusi S, Dowlati Y. Assessment of histologic criteria in the diagnosis of mycosis fungoides. *Int J Dermatol*. 2003;42(1):45-52. doi:10.1046/j.1365-4362.2003.01566.x
18. Ozkaya DB, Emiroglu N, Su O, et al. Dermatoscopic findings of pigmented purpuric dermatosis. *An Bras Dermatol*. 2016;91(5):584-587. doi:10.1590/abd1806-4841.20165124

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