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ORIGINAL ARTICLE

Clinical management of pigmented purpuric dermatoses: evidence from a survey among the Study Group on Cutaneous Vascular Diseases of the Italian Society of Dermatology and Venereology

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ABSTRACT

BACKGROUND: Pigmented purpuric dermatoses (PPD) are a group of chronic, benign skin conditions with limited evidence-based data regarding their diagnosis and management. The aim of our work was to evaluate the clinical practices of dermatologists in managing PPD, focusing on patient presentation, treatment preferences, and the need for further diagnostic evaluations.

METHODS: We utilized a survey-based design for the present study. The survey was distributed to the Study Group on Cutaneous Vascular Diseases and Skin Ulcers of the Italian Society of Dermatology and Venereology. The survey collected data on the number of patients evaluated weekly, the proportion of patients requesting treatment, preferred therapeutic approaches, and indications for additional diagnostic tests.

RESULTS: Twenty-four clinicians from 11 centers participated. Respondents reported managing an average of 5 PPD cases per week, with treatment required in approximately 50% of cases, while the remainder were incidental findings during evaluations for other dermatological conditions. Skin moisturizers and topical corticosteroids (tCS) were universally recommended as first-line treatments. Additional therapies included zinc oxide cream, compression stockings, and flavonoid-based oral supplements. Further diagnostic workups, such as leg Doppler ultrasound, skin biopsy, and blood tests, were considered necessary only in selected patients based on clinical presentation and comorbidities.

CONCLUSIONS: Moisturizers and tCS are the cornerstone of PPD treatment, supplemented by tailored therapies and diagnostic evaluations. These findings highlight the need for standardized, evidence-based guidelines to optimize the management of PPD.

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KEY WORDS: Skin diseases; Surveys and questionnaires; Skin pigmentation.

Pigmented purpuric dermatoses (PPs) are a group of chronic, benign skin disorders characterized by mild inflammation of the skin capillaries.[1] These conditions are collectively known as "capillaritis" and are characterized by non-blanching purpura.[2] Clinically, PPDs are characterized by the presence of reddish-brown macules and patches resulting from capillary leakage and hemosiderin deposition within the skin (Figure 1A). Although these disorders are not life-threatening, their persistent and often aesthetically disturbing nature can cause patient discomfort and require medical attention. Despite its distinctive clinical presentation, the pathogenesis, diagnosis, and treatment of PPD remain areas of ongoing research with limited evidence-based guidelines.[2]

As already anticipated, blood leakage from the skin capillaries results in hemosiderin deposition, which is ultimately responsible for the characteristic reddish-brown pigmentation.[3] Clinical manifestations of PPD are limited to the skin and, unlike vasculitides, are not associated with systemic involvement.[4] PPD presents with non-blanching purpura, petechiae, and hyperpigmentation, most commonly affecting the lower extremities. The condition is often asymptomatic, but some patients may report mild pruritus or burning. PPD can affect individuals of all ages but is more commonly observed in adults and the elderly.[5, 6] Although often asymptomatic, PPD may be a cosmetic concern or cause mild discomfort such as itching.[1, 2]

PPD are classified into several clinical subtypes based on lesion morphology and distribution, with Schamberg's disease being the most common form.[7] Schamberg's disease, also known as progressive pigmentary purpura, is characterized by the typical "cayenne pepper spots" that are either clinically or dermoscopically apparent as pinpoint erythematous macules and/or teleangiectasias macules surrounded by brown discoloration (Figure 1B). Gougerot-Blum Syndrome, also known as pigmented purpuric lichenoid dermatosis, presents with purpuric macules and lichenoid papules. Eczematid-like PPD (or "Ducas and Kapetanakis pigmented purpura") includes features of both capillaritis and cutaneous eczema. It typically presents as a solitary, golden-brown patch, often on the lower extremities. Lichen aureus, another distinct sub-

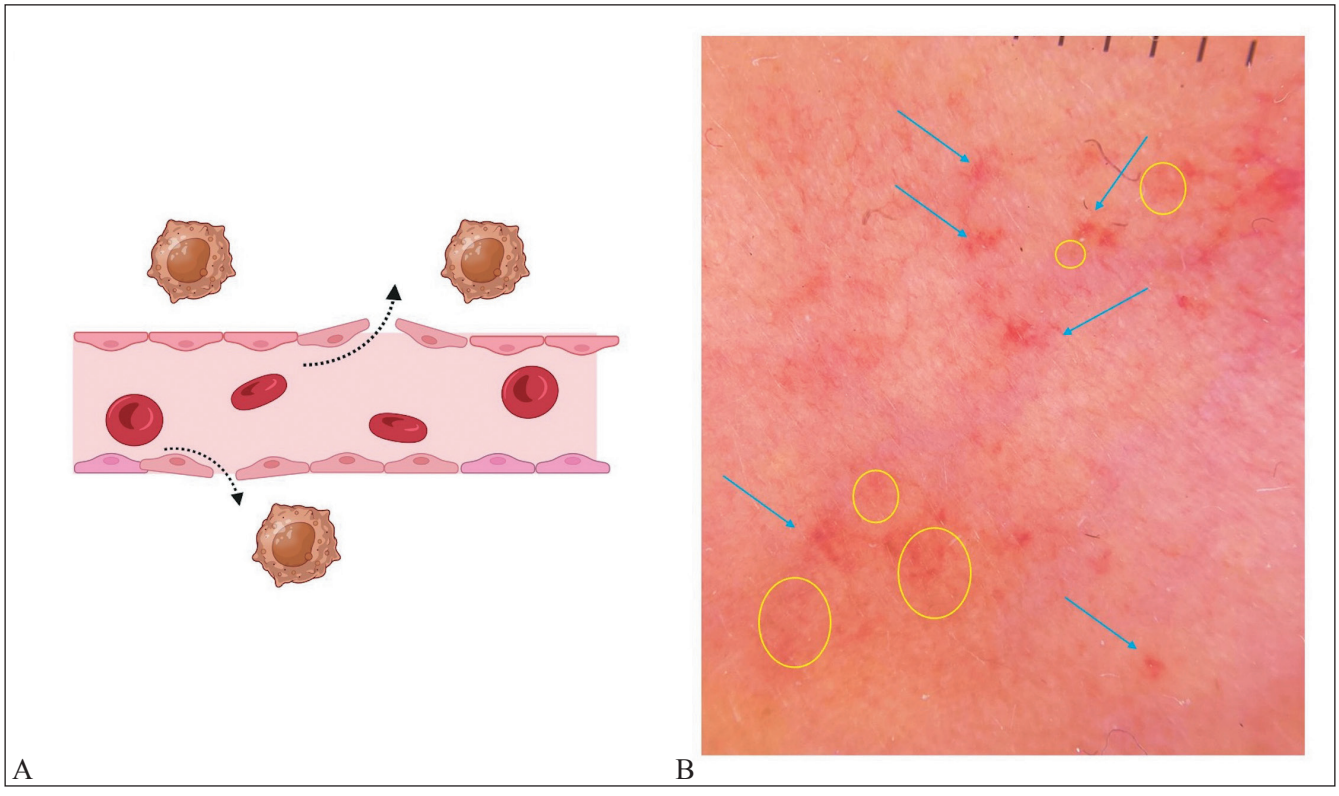

Figure 1.—A) Schematic representation of DPP pathogenetic mechanism: blood leakage from the skin capillaries resulting in hemosiderin deposition. Hemosiderin laden macrophages are represented in brown color (created with https://biorender.com); B) typical dermoscopic features in cutaneous capillaritides: teleangiectasia (arrows) and peri-capillary reddish-brown pigmentation (circles).

type, typically manifests as a unilateral, golden-brown to rust-colored patch or plaque.[8] It is most commonly seen on the lower extremities and is often described in younger individuals. The lesions are sharply demarcated and may occasionally be associated with mild pruritus.

Majocchi's disease, also known as "purpura annularis telangiectodes," is characterized by annular, ring-like purpuric lesions with a prominent telangiectatic component. This subtype is less common but can exhibit significant variability in lesion size and distribution.[9]

Rarer variants, including linear and granulomatous pigmented purpura, have also been described.[10] The complexity of PPD classification underscores the heterogeneity of the disease spectrum and highlights the need for a tailored diagnostic and therapeutic approach.

The exact pathophysiology of PPD remains poorly understood. Current hypotheses suggest that chronic inflammation of skin capillaries is primarily responsible for the extravasation of red blood cells, with subsequent hemosiderin deposition and characteristic pigmentation. Potential triggers include prolonged standing, venous insufficiency and/or hypertension, minor trauma, infection, aerobic exercise, contact allergens, and drug hypersensitivity.[11-14] Notably, immune-mediated mechanisms and systemic inflammatory factors have also been implicated, particularly in cases associated with systemic diseases such as diabetes or autoimmune diseases.[15] Interestingly, PPD has been associated with the use of certain medications, including immune checkpoint inhibitors such as pembrolizumab, which can trigger immune-mediated capillaritis.[16] In drug-related PPD cases, times of onset ranging from one week to years after initiating treatment have been reported.[13] These cases highlight the importance of evaluating medication history in patients presenting with new-onset PPD.

The diagnosis of PPD is primarily clinical, based on the characteristic morphology and distribution of the lesions. Dermatoscopy can aid in the evaluation, showing a diffuse coppery-red to brown pigmentation often characterized by perifollicular accentuation.[17] However, the absence of pathognomonic features sometimes requires histopathologic confirmation. In fact, a skin biopsy is sometimes performed to rule out vasculitis or other rarer differential diagnoses.[18-20] Differential diagnoses include vasculitis, stasis dermatitis, contact dermatitis, mycosis fungoides, and cutaneous sarcoidosis.[21] Histopathologic specimens of PPD typically show capillaritis, hemosiderin-laden macrophages, and extravasated erythrocytes without evidence of vasculitis.[3]

PPD have no definitive cure, and treatment focuses on symptom management and addressing patient concerns. Educating patients about the benign and chronic nature of PPD is critical to setting realistic expectations for treatment and outcomes. There are currently no standardized treatment guidelines for PPD, so management is largely based on clinical experience and case reports.[22-24] Commonly used treatments include lifestyle modifications (leg elevation, avoidance of prolonged standing), emollients, compression stockings, and topical corticosteroids (tCS).[25]

Emollients and tCS are often prescribed for symptomatic relief. Compression therapy and leg elevation are recommended for cases associated with venous stasis or edema.[2] Adjunctive treatments such as flavonoid-based oral supplements and topical depigmenting agents such as vitamin C may provide additional benefit, especially in chronic cases.[15] Other anecdotal therapeutic options are phototherapy and PDT.[26-28]

Despite its clinical relevance, PPD remains understudied, with management largely guided by expert opinion and anecdotal evidence. The lack of standardized diagnostic criteria and therapeutic protocols presents a challenge in ensuring consistent patient care. Studies such as those investigating granulomatous variants of PPD or its overlap with other dermatologic conditions provide valuable insights but highlight the need for more comprehensive, evidence-based investigations.[29, 30] This paucity of data underscores the importance of collaborative research efforts to establish comprehensive guidelines for the diagnosis and management of PPD.

The aim of this study was to raise awareness about a frequently under-represented condition and to provide a comprehensive overview of current therapeutic approaches to PPD in real-world clinical practice. Rather than comparing treatment efficacy, our goal was to highlight current management strategies, underscore existing knowledge gaps, and lay the groundwork for future research focused on treatment outcomes.

Materials and methods

This study used a survey-based design to gather insights from dermatology experts regarding their experiences and management practices for pigmented purpuric dermatoses (PPD). In order to target dermatologists with clinical experience in the management of PPD, all members of the Study Group on Cutaneous Vascular Diseases and Skin Ulcers of the Italian Society of Dermatology and Venereology were invited to participate in this study (N.=100).

The questionnaire was not validated, but all the questions had been agreed among the members of the Study Group (Supplementary Digital Material 1: Supplementary Text File 1).

The survey focused on collecting data on four key parameters: the number of PPD patients visited, the frequency of patient requests for therapy, the types of therapy recommended, and the need for further diagnostic testing. Both quantitative and qualitative questions were included in this study:

- number of patients: participating clinicians reported the approximate number of PPD patients they had seen in a given time period (one week), providing a quantitative measure of their clinical exposure;
- treatment seeking: respondents were asked how often patients sought treatment for PPD symptoms, expressed as a percentage of the total number of patients affected by PPD;
- type of therapy: participants chose from a list of therapies, including topical corticosteroids, emollients, compression therapy, systemic treatments, or no treatment, and could provide additional details about the rationale for their choice;
- diagnostic tests: experts indicated whether they ever considered further diagnostic tests, such as skin biopsies, blood tests, or imaging studies, to be necessary, and the situations that warranted such tests.

Statistical analysis

Responses were anonymized and aggregated for analysis. Descriptive statistics summarized data such as average number of patients treated, proportion of patients requesting therapy, and distribution of recommended treatments. The frequency and types of additional investigations were analyzed to identify common practices and potential consensus.

Results

Twenty-four clinicians from 11 national centers participated in this survey (24% response rate). According to the data collected, approximately five patients affected by PPD come to medical attention every week (mean: 5.08±1.98). However, only about half of the patients required treatment for PPD (mean 44.5%, median 50%), while the other cases of PPD were incidental findings in the context of dermatologic evaluation required for other skin problems. Main proposed treatments are included in Table I.

TABLE I.—*Commonly prescribed medications for pigmented purpuric dermatoses), with relative frequencies. Frequency is indicated as the percentage of clinicians that indicated such therapy in treated cases.*

Treatment	N.	%
Emollients/moisturizers	24	100%
Topical corticosteroids	24	100%
Zinc oxide cream	14	58.3%
Flavonoid-based oral supplements	8	33.3%
Vitamin C/ascorbic acid	4	16.7%
Compression stockings	13	54.2%
Draining treatments	3	12.5%
Rest, leg elevation	3	12.5%
Iron-chelating agents	3	12.5%
Topical calcineurin inhibitors	2	8.3%

All panel members (100%) stated that if treatment is required, the use of skin moisturizers and topical corticosteroids (tCS) is generally indicated. Four participants specified that the use of tCS was limited to forms of PPD other than Schamberg's disease. Whenever indicated (33.3% of cases), class III topical steroids, such as mometasone furoate or methylprednisolone aceponate, were considered the preferrable choice. Other commonly prescribed medications include zinc oxide cream (indicated by 14 dermatologists) as well as flavonoid-based oral supplements, which are considered a useful adjunctive treatment by several clinicians (N.=8) for their venotonic and capillary-protective properties. Topical vitamin C and topical iron chelating agents (mainly containing EDTA) were also sometimes used for their depigmenting effects in a smaller proportion of cases. Most of the experts surveyed prescribed compression stockings (N.=13). In terms of behavioral interventions, rest and leg unloading are more likely to be suggested in cases associated with peripheral edema/swelling. In selected cases, drainage devices and/or dressings may also be indicated, depending on the patient's clinical presentation. Two of the respondents also listed topical calcineurin inhibitors as possible maintenance treatment in selected cases.

All panel experts agreed that further diagnostic workup is required in only a proportion of patients, particularly when diagnoses other than PPD are suspected and should be ruled out, with the most important differentials including vasculitis, mycosis fungoides, stasis dermatitis and contact dermatitis. In these cases, additional testing may include vascular consultation and leg Doppler ultrasound, skin biopsy, blood tests (including inflammatory markers and autoantibodies), and patch testing. However, the type of workup is highly dependent on the clinical presentation and patient comorbidities.

Discussion

The present survey-based study provides valuable insights into the clinical management of pigmented purpuric dermatoses (PPD), a group of chronic, benign skin conditions with limited evidence-based guidance. By engaging 24 dermatology experts from 11 national centers, our research sheds light on the current diagnostic and therapeutic approaches to PPD while emphasizing the variability and challenges in managing this condition. The findings offer a snapshot of clinical practices, with several implications for improving patient care and guiding future research efforts.

First, our study underscores the high epidemiologic impact of PPDs in healthcare systems. In fact, PPD are frequently detected during dermatologic evaluation, although patients are often unaware of such a diagnosis or do not specifically seek medical care for this reason. On the other hand, the results of the study highlight another important aspect of PPD care: understanding and managing patient expectations. Although the condition is benign and often asymptomatic, the aesthetic implications can have a significant impact on quality of life. Approximately half of the patients seen for PPD actively sought treatment, suggesting a strong desire for therapeutic intervention even in the presence of minimal symptoms. This underscores the importance of patient-centered care and the need to educate patients about the chronic but non-threatening nature of PPD.

The therapeutic approaches reported in this study reaffirm the central role of moisturizers and topical corticosteroids (tCS) as first-line treatments for PPD. The use of zinc oxide cream and compression stockings also emerged as common practices, the latter especially for patients with associated venous insufficiency or peripheral edema. Adjunctive treatments, including flavonoid-based oral supplements and topical depigmenting agents such as vitamin C and DTA-based iron chelating agents, were less frequently employed but reflect efforts to address patient cosmetic concerns for chronic pigmentation, especially when residual inflammatory changes are absent or minimal. Notably, behavioral measures such as leg elevation and rest were recommended in selected cases, underscoring the need to address underlying circulatory factors. Such adjunctive therapies, while not universally adopted, demonstrate the diverse approaches clinicians employ to meet patient needs.[15] While some experts incorporate depigmenting agents to address aesthetic concerns, others focus solely on symptomatic relief, demonstrating a diverse range of practices that may lead to inconsistent patient outcomes.

Finally, the data presented also underscore that clinical evaluation remains the gold standard for the diagnosis of PPD, with further procedures required only in doubtful cases. In fact, experts agreed that further diagnostic workup is only necessary in a subset of patients, particularly when differential diagnoses such as vasculitis, stasis dermatitis, or contact dermatitis are suspected. The study identified a number of additional tests, including vascular consultation with Doppler ultrasound, skin biopsies, blood tests (*e.g.*, inflammatory markers and autoantibodies), and patch testing. These findings are consistent with the literature emphasizing the importance of a tailored approach based on clinical presentation and comorbidities.[3, 21] The variability in diagnostic practices reflects the lack of standardized criteria, further complicating the clinical management of PPD.

Although PPD is a well-recognized entity with characteristic clinical features, its management remains largely empirical due to a lack of high-quality research and standardized protocols. This study is among the first to systematically explore expert experience and practice, shedding light on the real-world challenges faced in diagnosing and treating this condition. The novelty of this research lies in its focus on the practical approaches taken by clinicians in the absence of definitive therapeutic or diagnostic frameworks. By analyzing data on patient volume, therapy requests, treatment choices, and use of further diagnostic testing, the study provides valuable insights into the variability and commonalities of current clinical strategies.

Limitations of the study

Several limitations of this study must be acknowledged. First, the reliance on self-reported data introduces the potential for recall bias, which may affect the accuracy of reported practices. Second, the sample size of 24 clinicians, while providing valuable insights, limits the generalizability of the findings. Third, the survey did not directly assess patient outcomes or satisfaction with the treatments provided, which would have added a critical dimension to understanding the effectiveness of current practices. Additionally, the lack of stratification by PPD subtype and the potential influence of seasonal variations on the epidemiology of PPD represent further sources of bias. Finally, the study is limited by the absence of advanced statistical analysis. Due to the low response rate and lack of comparative data, only descriptive statistics could be applied, precluding more in-depth quantitative interpretation.

Addressing the gaps identified in this study will require a multifaceted approach. The development of standardized diagnostic and therapeutic guidelines for PPD is a priority, as this would provide clinicians with an evidence-based

framework to improve consistency of care. Prospective studies evaluating the efficacy of commonly used treatments, including topical corticosteroids, compression therapy, and flavonoid-based supplements, are critical to determining the most effective management strategies. Such studies should also evaluate the role of adjunctive therapies, such as depigmenting agents, in improving patient outcomes. Equally important is patient-centered research. Investigating patient-reported outcomes and satisfaction with different treatment modalities will provide valuable insights into how these interventions affect quality of life. Understanding patient perspectives can also help clinicians tailor treatments to address both functional and aesthetic concerns. In addition, subgroup analyses examining variations in management practices and outcomes across PPD subtypes are needed to identify tailored approaches for specific clinical scenarios. Finally, the establishment of longitudinal registries to collect long-term data on the natural history, triggers, and treatment responses of PPD will provide a solid foundation for future guidelines. Such registries could facilitate the identification of trends and patterns, contributing to a deeper understanding of this condition.

Conclusions

The results of this survey-based study make a significant contribution to the understanding of clinical practices for the management of PPD, a condition for which evidence-based guidelines are notably lacking. At the same time, this survey underscores the heterogeneity of clinical practices for the management of PPD. While moisturizers and tCS remain universally recommended, the variability in adjunctive therapies and diagnostic workups highlights the need for individualized care tailored to patient presentation. In conclusion, the present study highlights the gap between clinical need and available evidence, underscoring the urgency for further research to establish consensus guidelines and evidence-based recommendations. Moving forward, collaborative research efforts are essential to address the gaps in understanding and develop comprehensive protocols that ensure consistent and effective management of PPD.

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Conflicts of interest
The authors certify that there is no conflict of interest with any financial organization regarding the material discussed in the manuscript.

Authors' contributions
Alessia Paganelli, Maria C. Collina, Maria A. Pilla, and Emanuele Scala conceived the original idea. Alessia Paganelli and Alberico Motolese supervised the project; Caterina Longo, Giovanni Pellacani, Andrea Conti, Annunziata Dattola and Antonio G. Richetta helped supervise the project. Marco Spadafora, Claudia Pezzini, Victor D. Mandel, Francesca Di Tullio, Barbara De Pace, Alfonso Motolese, Andrea Marani, Luca Ambrosio, and Camilla Vassallo were involved in data collection; Alessia Paganelli and Andrea Michelerio took the lead in writing the manuscript. Federico Garbarino contributed to writing of the manuscript. All authors provided critical feedback and helped shape the research, analysis and manuscript. Andrea Michelerio, Alfonso Motolese, Federico Garbarino contributed to the interpretation of the results. All authors read and approved the final version of the manuscript.

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