INTERNATIONAL JOURNAL
of PEDIATRIC DERMATOLOGY

Vol 1. Iss. 1 (January-June, 2024)
ISSN 3035-1006 online

Pigmented purpuric dermatosis in pediatric patients

M. Zavala[1], A. Chaaban[1], H. Fuenzalida[1]

[1]*Department of Dermatology, Hospital El Pino, Santiago, Chile*

KEYWORDS

pigmented purpuric dermatoses, purpura, pediatric, children

ABSTRACT

Pigmented purpuric dermatoses correspond to chronic, idiopathic and self limited diseases. They are rare, especially in the pediatric population. These conditions manifest as purpuric lesions with variable pigmentation as a result of erythrocyte extravasation and hemosiderin deposits in the superficial dermis. They may have a significant impact on patients' quality of life. There is no standardized treatment, so it must be evaluated on a case by case basis to define the management, considering risk and benefits involved.

CORRESPONDING AUTHOR

Mario Zavala MD
Department of Dermatology
Hospital El Pino
Santiago, Chile

mario.zavala.mena@gmail.com

1. Introduction

Pigmented purpuric dermatoses (PPD) are a group of uncommon benign disorders characterized by grouped petechiae, purpuric macules or patches, with pigmentation ranging from erythematous-violaceous to yellow-brown. They occur more frequently in adult men and are characteristically located on the lower extremities. They are classified based on their clinical and histopathological features into five classic subgroups (Schamberg disease, purpura annularis telangiectodes of Majocchi, lichen aureus, eczematid like purpura of Doucas and Kapetanakis, purpuric lichenoid dermatosis of Gougerot and Blum). The commonest is Schamberg disease. In pediatric patients, Schamberg disease and lichen aureus are the most frequent (1, 2, 3). As they manifest as purpura, pathologies associated with hemostasis disorders and vasculitis should be considered in their differential diagnosis (1). Being idiopathic and self limited conditions, they are usually not associated with systemic findings and may not require any treatment (1, 2).

There is still no standardized management; recommendations for treatment are based on case series (4). Several cases of PPD have been described in pediatric patients, however, being an uncommon pathology, even more in this age group, information is limited and remains as a therapeutic challenge (2, 5).

2. Case Report

A 14 year old female patient, with no medical history, consulted for a 4 month history of erythematous purpuric macule in the right scapular region, which progressively spread medially with the appearance of lesions with similar characteristics.

The patient emphasized that she had exacerbations in which the lesion became more erythematous, leaving purpuric lesions in the affected area.

Physical examination revealed erythematoviolaceous patches with fuzzy borders and negative diascopy (Fig. 1). Dermatoscopy showed red and violaceous dots on a patchy erythematous base, with heterogeneous distribution (Fig. 2). Laboratory testing revealed low inflammatory markers, negative autoantibodies, and no alterations in coagulation and blood biochemistry test. Skin biopsy reported “Orthokeratotic epidermis with generally preserved histoarchitecture. Focal perivascular lymphocytic inflammatory infiltrates are identified in the superficial dermis, with no evidence of parietal vascular damage. Erythrocyte extravasation in the dermis is noted in some areas”.

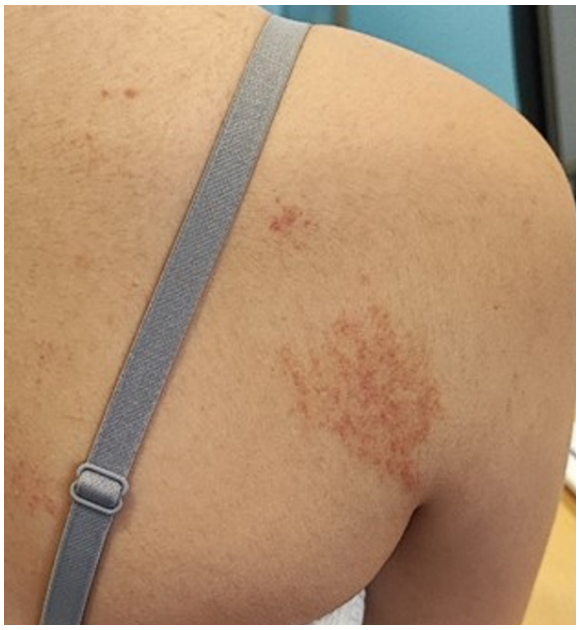

Fig 1. *Clinical picture showing erythematoviolaceous patches in the right scapular region.*

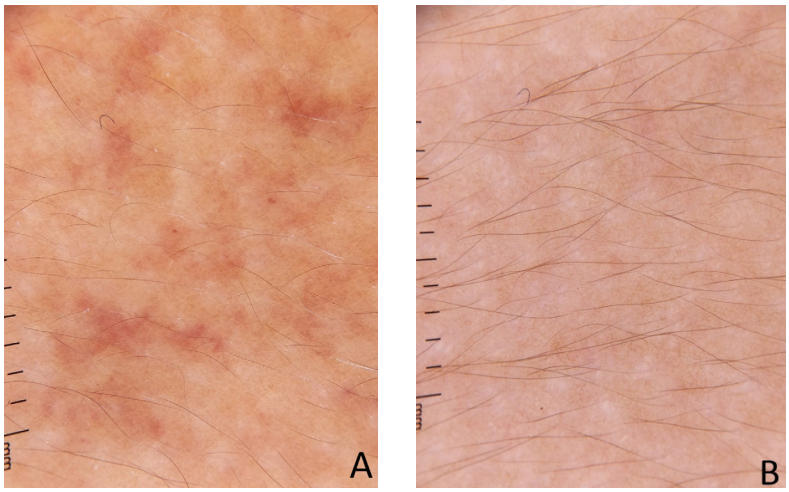

Fig 2. *Dermoscopic findings showing (a) red and purpuric dots on a patchy erythematous base and (b) adjacent normal skin.*

3. Discussion

PPD are idiopathic and benign conditions characterized by erythematous-violaceous lesions with variable pigmentation due to red blood cells extravasation and hemosiderin deposits. They are rare conditions that evolve in a chronic but self-limited manner. There isn't any characteristic that allows predicting the time of resolution. In the pediatric population, resolution may take years (1, 3). PPD are more common in adult men, between the fourth and fifth decade, although they can appear at any age. These pathologies infrequently occur in children, especially infants. The average age in this group varies between 8 and 15 years. In contrast, purpura annularis telangiectodes of Majocchi and lichen aureus occur predominantly in children and young adults, and Majocchi disease affects mostly women (3, 6, 7, 8). The most prevalent subtype of PPD is Schamberg disease. Lichen aureus also occurs with high frequency in children (1, 4).

Their etiology is still unknown, however, all of these entities show perivascular mononuclear infiltrate in the papillary dermis, erythrocyte extravasation and hemosiderin deposits in biopsies. The inflammation cause remains unclear (1, 2).

The pathophysiology of PPD is not completely understood, but it has been proposed to involve an altered cellular immunity and capillary fragility (1, 4, 5).

Several factors have been associated with PPD as triggers, like infections (hepatitis B and C), physical activity, capillary fragility, venous hypertension, diabetes and drugs, including vitamins (B1), vasodilators (nitroglycerin, hydralazine, sildenafil), analgesics (acetaminophen, acetylsalicylic acid) and retinoids (isotretinoin), among others (1, 4).

Clinically, they manifest as petechiae and purpuric macules or patches, on a base with colors ranging from red and purple to yellow and brown, typically on both lower extremities. They may spread to the trunk and upper extremities, particularly in young patients in whom a generalized distribution has also been observed. The distinctive color of the lesions is due to capillary hemorrhage and the reabsorption of hemosiderin (1, 2, 3, 5).

PPD are generally asymptomatic, but may be pruritic (2). They present exacerbation and remission periods before complete resolution of the disease (4). In children, it has been reported from 6 months to 9 years until the resolution of the condition (5).

They are classified into five classic subtypes according to their clinical and histopathological features, despite this, they all share common histological findings. Other variants have also been described but are less prevalent (granulomatous PPD, itching purpura of Loewenthal, familial, linear PPD, transitory PPD) (4).

Schamberg disease, also called progressive pigmented purpuric dermatosis, is the most frequent subtype of PPD. It manifests as non blanchable orange-red or red-brown macules with peripheral petechiae in a pattern of grains of cayenne pepper. It is often asymptomatic and located on both lower extremities, but they can spread to the trunk, upper extremities, thighs and buttocks (1, 4).

Purpura annularis telangiectodes of Majocchi or

Majocchi disease, affects predominantly young female patients. It is characterized by annular red-violaceous macules or patches, with peripheral dark red telangiectasias. These lesions exhibit central clearing giving them their distinctive annular pattern. Majocchi disease begins in lower extremities and may spread to the trunk and upper extremities; it usually resolves within months (4, 7).

Lichen aureus is a more localized presentation of PPD, characterized by single or isolated lesions which begin suddenly. Typically presents golden-brown macules or papules that tend to coalesce into lichenoid patches or plaques of 1 to 20 centimeters. The color of the lesion varies between yellow-orange, golden-brown, rust and purple-brown (2, 4, 9). They are usually asymptomatic and affect lower extremities unilaterally, but may present skin itching, pain and occur in atypical
locations, such as the trunk and upper extremities, especially in children (4, 7). A specific dermoscopy pattern has been described, characterized by a coppery-red background associated with red, brown and gray dots, red and brown globules, and a pseudo reticulum formed by interconnected pigmented lines. Rare clinical presentations have been described in children and adolescents, such as zosteriform and segmental variants along Blaschko's lines or related to the the trajectory of cephalic and saphenous veins (4). In pediatric population has spontaneous remission after 1 to 12 years (7, 9).

Eczematid-like purpura of Doucas and Kapetanakis, also called pruritic purpura, as the name suggests, is a pruritic presentation of PPD and more extensive than other subtypes. It manifests clinically similar to Schamberg disease but with scales on the surface. It has an acute onset in a period of 15 to 30 days and may persist for months or years, remitting spontaneously. Allergic contact dermatitis due to rubber or clothing have been associated with this subgroup (4, 8).

Purpuric lichenoid dermatosis of Gougerot and Blum is characterized by the appearance of confluent violaceous lichenoid papules forming plaques. Papules are polygonal or round. It is more frequent in adult men and located on the legs but may affect the trunk and thighs; it remits in months or years (1, 4, 7, 8).

Regardless of age, PPD have the same clinical features; however, as they are less common conditions in childhood, their diagnosis will be more complex (3).

PPD's diagnosis is based on clinical findings and may be confirmed by biopsy. Dermoscopy is a non-invasive method that allows visualization of morphologic features that may not be identified with the naked eye and will provide diagnostic clues. Findings in PPD's dermoscopy are diffuse coppery-red pigmentation (due to dermal infiltrate, erythrocyte extravasation and hemosiderin deposition) and red globules and dots (due to extravasation
of red blood cells and dilated blood vessels). Brown dots and pigmented pseudo reticulum may be observed in some cases.

Although there are no specific markers, it is recommended to perform tests to rule out other causes of purpura such as thrombocytopenia and coagulopathy through a complete blood count and coagulation tests, autoimmune disorders including rheumatoid factor and antinuclear antibodies, and the detection of chronic infections by hepatitis B and C viruses (4, 6).

Biopsy reveals lymphohistiocytic inflammatory infiltrate in small caliber vessels, dilated blood vessels, endothelial cell swelling, extravasation erythrocytes and hemosiderin deposition in macrophages (2, 4, 8).

Schamberg disease and purpura annularis telangiectodes of Majocchi exhibit the distinctive perivascular infiltrate. Lichen aureus is histologically characterized by an intact epidermis separate from a band-like dermal lymphohistiocytic infiltrate by healthy connective tissue (Grenz zone). It is the only variant that does not present epidermal spongiosis. The infiltrate pattern varies according to the time of evolution, in initial phases the biopsy presents patches, and later it exhibits a band (2, 4, 8, 9).

Purpuric lichenoid dermatosis of Gougerot and Blum also presents a band-like dermal infiltrate, in addition to epidermal spongiosis with irregular parakeratosis. Finally, histopathologic findings in eczematid-like purpura of Doucas and Kapetanakis are marked epidermal spongiosis and a infiltrate with increased numbers of neutrophils (2, 4).

Disorders associated with purpura should be ruled out. The most frequent that must be excluded are: thrombocytopenia, use of antiplatelet agents, anticoagulants or corticosteroids and venous stasis purpura.

Perls and Fontana Masson stains allow differentiating PPD from stasis dermatitis, the first ones present hemosiderin deposits in the superficial dermis, while in the second ones, these deposits are located in deeper layers (4). Another differential diagnosis is leukocytoclastic vasculitis, but unlike PPD, it presents with palpable purpuric lesions. Kaposi sarcoma may manifest similar to purpuric lichenoid dermatosis of Gougerot and Blum, but often affects elderly and immunosup-

pressed patients. More chronic and disseminated cases of PPD may correspond to mycosis fungoides, which in early stages may mimic these conditions. Its biopsy reveals monoclonal infiltrate and loss of CD7 (1, 4).
Regarding treatment, there is no standardized management. Given the benign nature of the condition, generally asymptomatic and self-limited, management could be limited to follow-up. However, presentations of PPD with extensive, symptomatics (pruritic) and long-standing lesions, may have a greater impact on the patients' quality of life and require treatment (4).
Most recommendations are based on small case series. Some systemic and local treatments have shown improvement, the most reported are topical corticosteroids, phototherapy, vitamin C, rutoside and pentoxifylline, among others (4, 5).

4. Conclusion

PPD are rare idiopathic pathologies, but with a significant impact on the quality of life of those affected. Treatments have been based on clinical case series, with no standardized management available to date. Although these conditions are infrequent in the pediatric population, the diagnosis should be considered when presenting with characteristic lesions. Biopsy provides useful information to differentiate the classic forms of PPD.

FUNDING
No funding was received.

DISCLOSURE
All authors report no conflict of interest.

References

1. Ollech A, Paller AS, Kruse L, et al. Pigmented purpuric dermatosis in children: a retrospective cohort with emphasis on treatment and outcomes. J Eur Acad Dermatol Venereol. 2020;34(10):2402-2408.

2. Spigariolo CB, Giacalone S & Nazzaro G. Pigmented purpuric dermatoses: A complete narrative review. J Clin Med. 2021;10(11):2283.

3. Martos-Cabrera L, López-Balboa P, Ramírez-Lluch M, Colmenero I, Mateos-Mayo A, Torrelo A & Hernández-Martin A. Pigmented purpuric dermatosis in an infant. Pediatr Dermatol. 2021;38:504-505.

4. Martínez Pallás I, Conejero del Mazo R, & Lezcano Biosca V. Dermatosis purpúricas pigmentadas. Revisión de la literatura científica. Actas Dermosifiliogr. 2020;111(3):196-204.

5. Coulombe J, Jean SE, Hatami A, Powell J, Marcoux D, Kokta V & McCuaig C. Pigmented purpuric dermatosis: Clinicopathologic characterization in a pediatric series. Pediatr Dermatol. 2015;32(3):358-362.

6. Ozkaya DB, Emiroglu N, Su O, Cengiz FP, et al. Dermatoscopic findings of pigmented purpuric dermatosis. An Bras Dermatol. 2016;91(5):584-587.

7. Chayed Z, Mose KF & Bygum A. Pigmented purpuric dermatoses. Ugeskr Laeger. 2020;182(21):V12190682.

8. Kim DH, Seo SH, Ahn HH, Kye YC & Choi JE. Characteristics and clinical manifestations of pigmented purpuric dermatosis. Ann Dermatol. 2015;27(4):404-410.

9. Yáñez S, Val-Bernal JF, Arce F & Navarro O. Liquen áureo o purpúrico. Estudio de seis casos, tres con presentación zosteriforme. Actas Dermosifiliogr. 2002;93(7):437-442.