

Pigmented Purpuric Dermatoses

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Citation: Jacob M, Wright R, Mazur L, Aly F (2018) Pigmented Purpuric Dermatoses. J Paediatr Neonatal Dis 3(2): 203

Received Date: June 29, 2018 **Accepted Date:** August 28, 2018 **Published Date:** August 30, 2018

Abstract

The pigmented purpuric dermatoses (PPD) are skin rashes that are benign but can often be mistaken for other purpura-causing diseases, which must be ruled out. Although they are more prevalent in adults, they can also be seen in children. Though these dermatoses rarely involve other organs, the rash can be distressing for the parents of an adolescent or child. We presented a case of a 15 year old girl with a pathological diagnosis of eczematid-like form of PPD, which clinically diagnosed as the Schamberg's form of PPD. Biopsy is frequently necessary to reach a final diagnosis.

Keywords: Pigmented Purpuric Dermatoses; Schamberg Disease; Eczematid-like Type; Rutoside; Ascorbic Acid

List of abbreviations: PPD: Pigmented Purpuric Dermatoses

Case Report

A 15 year old female presented to the clinic with a six month history of a 'rash' on her arms and legs. It started on the feet and spread to her upper legs and arms. She had some transient pruritus but denied fever and chills, weight loss, abnormal bruising or bleeding, abdominal pain, hematuria, menorrhagia, arthralgias, and edema. There were no known sick contacts, or history of travel. She was not on any medication but occasionally used acetaminophen for headaches.

On physical examination, the vital signs were normal and her body mass index was 28.49 kg/m² (96%). Her skin had multiple erythematous, discrete, purpuric, non-palpable, non-blanchable macules over the arms and legs, both anterior and posterior aspect. The face, trunk, palms and soles, and buttocks were spared. The rest of the physical examination was unremarkable except for acanthosis nigricans on the posterior neck (Figure 1 and 2).

Initially, she was diagnosed with Schamberg's disease and was prescribed triamcinolone cream. However, when the rash failed to improve she was referred to dermatology. Biopsy of one of the lesions revealed capillaritis with abundant extravasated erythrocytes with spongiosis without mounds of parakeratosis, eosinophils or vasculitis. She was diagnosed with eczematid-like type of PPD.

Follow up and Outcome

Our patient was started on Rutoside 50mg BID and ascorbic acid 500mg BID. A few weeks later, she happily noted that the rash began to fade.

Discussion

Pigmented purpuric dermatoses (PPD) or Pigmented Purpuric Eruptions (PPE) are a group of chronic benign skin conditions characterized by symmetrical petechiae and purpura that are usually localized to the lower extremities [1]. Five types, Schamberg's disease, Majocchi's disease (purpura annularis telangiectodes), Lichen aureus, Eczematid-like purpura of Doucas-Kapetanakis, and Gougerot-Blum are described in the Table 1. Although there are differences in the morphology between the conditions, their pathology is similarly characterized by perivascular lymphocytic infiltration, red blood cell extravasation, and hemosiderin deposition. Schamberg's disease is the most common type in both adults and children. It was first described in 1901 by Schamberg in a teen with reddish-brown oval patches bordered by red to brown macules "cayenne pepper" spots at the borders [2]. The

cayenne pepper spots represent the extravasated red blood cells or hemosiderin deposits. Lesions are typically asymptomatic but mild pruritus can occur [1,3]. The clinical course waxes and wanes over several months but spontaneous remission is not uncommon.



Figure 1: (A) Anterior Pigmented Purpuric Dermatoses; (B) Anterior Pigmented Purpuric Dermatoses



Figure 2: Posterior Pigmented Purpuric Dermatoses

Although the etiology is unknown, possible mechanisms include capillary fragility, humoral, and cell-mediated immune responses, and medication reactions [2]. In addition to the vascular abnormalities, immunohistology shows a perivascular infiltrate of CD3+, CD4+ and CD1a+ dendritic cells with close spatial contact between lymphocytes and dendritic cells. Perivascular immunoglobulin and complement deposits suggest a role for immune complexes in the pathogenesis [1]. Hypercholesterolemia, venous insufficiency, focal infections, chemical ingestions, exercise, and gravitational dependency may also play a role. Medications associated with PPDs include acetaminophen, aspirin, NSAIDs, glipizide, glybuzole, diuretics, dipyridamole, and bezafibrate (a lipid-lowering agent). They act as haptens that lead to the formation of antibody-antigen complexes. These deposit in the endothelium causing vascular disruptions and eventually the clinical presentation of PPD [1-4].

Although some cases resolve spontaneously treatment options include topical steroids, oral antihistamines, pentoxifylline (an agent that decreases blood viscosity), bioflavonoids and ultraviolet B (UVB) light therapy [2]. A study of 17 patients showed that PPD resolved without treatment in five and with topical corticosteroids or UVB in eight. The median duration to resolution was less than one year (range 6 months to 9 years) [5]. Topical corticosteroids are used for their vasoconstrictive, permeability reducing, and antipruritic effects. However, long-term use may lead to skin atrophy, increased vascular fragility and may worsen the course of the disease [6]. Narrow band UVB therapy acts to suppress T lymphocyte activity and interleukin 2 production [7].

Pentoxifylline may be more beneficial than topical steroids but its long-term efficacy is unknown [8]. Bioflavonoids including Rutoside and ascorbic acid are also beneficial. As antioxidative radical scavengers they reduce capillary permeability and fragility. Additionally, they modulate signaling in endothelial cells to reduce vascular inflammation. Ascorbic acid is also necessary for collagen synthesis and maintenance of the basal lamina and connective tissue of capillaries. One study showed that 70% of patients had resolution of the rash after 8 months and those outcomes were better with early treatment [5]. In cases associated with medications, resolution may occur with discontinuation of the offending agent.

There are reports of an association between pigmented purpura-like rashes and progression to cutaneous T-cell lymphoma (mycosis fungoides). These patients have a clinical presentation of PPD with histopathologic findings of mycosis fungoides (MF). The clinical presentation of MF progresses from patches to infiltrated plaques and eventually cutaneous tumors. Therefore, presentations of PPD that are persistent need close follow up [9].

Interestingly, our patient's clinical presentation was typical of Schamberg's disease while her biopsy results showed the eczematid-like type. Clinically, she did not have the scaling and pruritus of the eczematid-like type but her biopsy showed spongiosis. The typical presentation of eczematid like purpura of Doucas-Kapetanakis has a more extensive distribution with an eczematous appearance and associated pruritus. Histologically, there is spongiosis with inflammation of the epidermis with the perivascular lymphocytic infiltrate. Eczematid-like purpura may be a pruritic variant of Schamberg's disease which could explain the overlapping features in our patient [2]. In the prior cases, treatment strategies included triamcinolone cream, ascorbic acid and narrow band UVB therapy. There was improvement with these treatments, however, the clinical course of eczematid-like purpura can wax and wane with time [1,3].

Disease	Epidemiology	Appearance	Distribution	Pruritic	Prognosis
Progressive pigmented purpuric dermatosis (Schamberg's Disease)	More frequent in adults than children, but the segmental form is more common in children than adults.	Patches in absence of papules and scaling.	Commonly involves the legs, but can begin to involve the arms as well. It is usually bilateral and symmetrical and sometimes segmental.	Usually no associated pruritus.	Will likely devolve on its own with the average course being between one and two years.
Eczematid-like purpura of Doucas and Kapetanakis [10]	Presents with seasonal eruption and is seen more often in males	Presence of scaling	Involves legs but can extend toward the upper body	Pruritus is a characteristic feature.	Spontaneous improvement can be seen in a few months, but tends to recur
Lichen aureus [11,12]	More common in adults	Macules with evident micropapules, and often has golden color	More often segmental and be unilateral. Typically involves relatively less surface area	Can be pruritic.	Has a longer course in comparison to the other purpuric dermatoses. There have been reports of 4 to 8 year averages for the disease.
Purpura Annularis Telangiectodes [13,14]	More common in female adolescents	Patches with centrifugal evolution, central resolution and telangiectasias	Commonly involves the legs	Can be mildly pruritic	Can be chronic and progressive
PPD of Gougerot-Blum [15,16]	More common in adults	Combination of Schamberg-like and red-brown lichenoid papules	Usually involves lower legs	Can be pruritic.	Can be chronic

Table 1: Differential Diagnosis of Pigmented purpuric dermatosis

Conclusion

Our patient was a 15 year old female with an atypical presentation of eczematid-like purpura of Doucas and Kapetanakis. She underwent a treatment regimen of Rutoside and Ascorbic acid with improvement of symptoms. However, due to the likelihood of recurrence close follow-up is planned. Our case showed the importance for biopsy to reach a final diagnosis as prognosis may vary depending on the type of PPD.

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