

A Case of Majocchi Purpura: Clinical and Histopathological Approaches for Diagnosis

Fadhli Aulia Mughni, Sarah Mahri, Teffy Nuary, Rahadi Rihatmadja, Kusmarinah Bramono
*Department of Dermatology and Venereology, Faculty of Medicine University of Indonesia / Dr. Cipto Mangunkusumo
National General Hospital, Jakarta, Indonesia*

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Abstract: Majocchi Purpura is one of the morphological varieties of pigmented purpuric dermatoses. They are rare disorders with unknown etiology, and characteristically presents as nonblanching petechiae, pigmentation, and the presence or absence of telangiectasia with a predilection for the lower extremities. They are usually asymptomatic and self-limiting, but tend to have a chronic course and are highly recurrent. The diagnostic approach for patients suspected with pigmented purpuric dermatoses is quite straightforward, with clinical observation and histopathological examination as the mainstay diagnostic modalities. However, this disorder may initially show similar manifestations as other cutaneous diseases, such as contact dermatitis, vasculitis, or mycosis fungoides, making it often misdiagnosed and mistreated by dermatologists. We present a case of a 28-year-old woman initially referred with polyarteritis nodosum. However, the lesions were painless and consisted of several hyperpigmented annular patches that clinically resembled Majocchi Purpura. We submitted the case for biopsy with vasculitis as the differential diagnosis. Histopathological findings were consistent with pigmented purpuric dermatosis, that we commenced the treatment with symptomatic measures supplemented with antioxidants. Recognizing distinctive clinical patterns of disease was indispensable. Laboratory examinations had limitations, and their results needed to be interpreted within the clinical context. The correct diagnosis will prevent overtreatment and unnecessary healthcare visit by patient.

1 INTRODUCTION

Pigmented Purpuric Dermatoses (PPD) is a group of disorders characterized by nonblanching purpuric rash, leaving residual hyperpigmented patches mainly on lower extremities. (Sardana, 2004) The etiology is unknown, although several drugs and other conditions have been documented. (Devere, 2012) Most patients are asymptomatic or presenting with mild symptoms. PPD is usually chronic, highly recurrent, and difficult to treat, although self-limiting cases have been reported. (Devere, 2012) This disorder is morphologically categorized into: (1) Schamberg's disease (SD), (2) Purpura annularis telangiectodes of Majocchi (Majocchi Purpura), (3) Pigmented purpuric lichenoid dermatosis of Gougerot and Blum (PPLD), (4) Eczematid-like purpura of Doucas and Kapetanakis, (5) Itching purpura of Lowenthal, and (6) Lichen aureus (LA). (Kim, 2015)

PPD is considered uncommon, despite lack of sufficient data in Indonesia. A clinicoepidemiological study by Sharma and Gupta Sharma, 2012 in 2012 found 0.18% PPD cases from 55,323 patients. All races may be affected, with more occurrence seen in males except in Majocchi purpura. (Sardana, 2004, Hoesly, 2009) The majority have history of routine activity that put constant pressure to the limbs, such as prolonged standing during work, high-intensity sport, or repetitive tasks. Several factors is believed to contribute to its pathogenesis, including capillary fragility, humoral immunity, cell-mediated immunity, gravitational forces, venous hypertension, focal infection, and contact allergy. (Devere, 2012, Sharma, 2012)

Most cases were diagnosed clinically. The presentations of PPD are usually characteristic, with pigmented patches and petechiae as the most consistent findings, without palpable purpura. However, initial manifestations may easily be interpreted as other cutaneous diseases, mainly

because their rare occurrence entices clinicians to weigh other more common disorders showing similar features. Pigmented purpuric dermatoses need to be distinguished from contact dermatitis, stasis dermatitis, angioma serpiginosum, mycosis fungoides, and most importantly, vasculitis. (Sardana, 2004, Devere, 2012) Histopathology examination most commonly shows perivascular infiltrate of lymphocytes around superficial blood vessels, endothelial cell swelling, erythrocyte extravasation, and hemosiderin deposition. (Sardana, 2004-Kim, 2015) While crucial for confirming the clinical diagnosis of PPD, skin biopsy is also important to exclude cutaneous T-cell lymphoma, which in its early stages closely resembles PPD. (Sardana, 2004)

No satisfying therapy has been found for PPD. Some lesions have been reported to subside spontaneously, however numerous agents have been used to alleviate the symptoms and skin lesions, including topical corticosteroids, antihistamines, bioflavonoids, ascorbic acid, griseofulvin, pentoxifylline, cyclosporine, and phototherapy, with variable but inconclusive outcomes. (Sardana, 2004) Misdiagnosis may lead to overtreatment and unnecessary healthcare visits, therefore it is crucial for dermatologists to be able to recognize PPD lesions and manage the patients accordingly.

2 CASE

A 28-year-old woman was referred to the Dermatovenereology clinic, Cipto Mangunkusumo National Central General Hospital with the diagnosis of polyarteritis nodosum. She presented with multiple brownish-red lesions on the legs that have been present for four months. Initially, asymptomatic red spots appeared on both feet. They spread upward reaching up to the thighs. Approximately two weeks later, some lesions faded to brownish discoloration. The lesions were more noticeable in relatively cold environment, such as in air-conditioned room. She also occasionally complained of ankle joints pain since two months before presentation. She took oral B-complex vitamins, but to no avail.

Physical examination was unremarkable except the presence of multiple brownish-purpuric patches bilaterally on the arms, lower legs, and feet. Some of

the macules were annular and reticular. No palpable purpura was observed. (Figure 1).

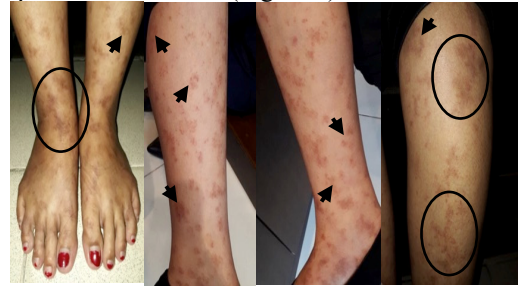


Figure 1. Multiple petechial and hyperpigmented patches in annular (arrow) and reticular (circle) configurations.

Past medical history was unremarkable. Previous use of medications was denied. The patient usually wore high-heeled shoes for work, which require repeated walking between offices during working hours. Initial laboratory examinations showed mild leukopenia, eosinophilia, and high erythrocyte sedimentation rate. The results of hemostasis, urinalysis, as well as blood chemistry examinations were within normal ranges. At the Internal Medicine clinic, she was also examined for anti-nuclear antibodies (ANA), Hepatitis B surface antigen (HbsAg), anti-hepatitis C virus (anti-HCV), human immunodeficiency virus (HIV), mixed activated partial thromboplastin time (APTT), and lupus anticoagulant to rule out other diagnostic possibilities. All those results were negative, except for ANA.

We diagnosed this patient with Majocchi purpura. Schamberg disease, another type of PPD, and vasculitis was also considered. The patient was then sent for biopsy. Histopathology from two different sites revealed superficial perivascular lymphocytic infiltrates, endothelial cell swelling, and erythrocytes extravasation, which are consistent with PPD. (Kim, 2015) (Figure 2)

The patient has been followed up for two months, and was treated symptomatically with emollients and antioxidants, including ascorbic acid 500 mg daily. Topical corticosteroid twice daily was started one month ago because the patient exhibited mild occasional pruritus. There was slight improvement of the patient's skin lesions, without worsening of symptoms or appearance of new purpuric patches.

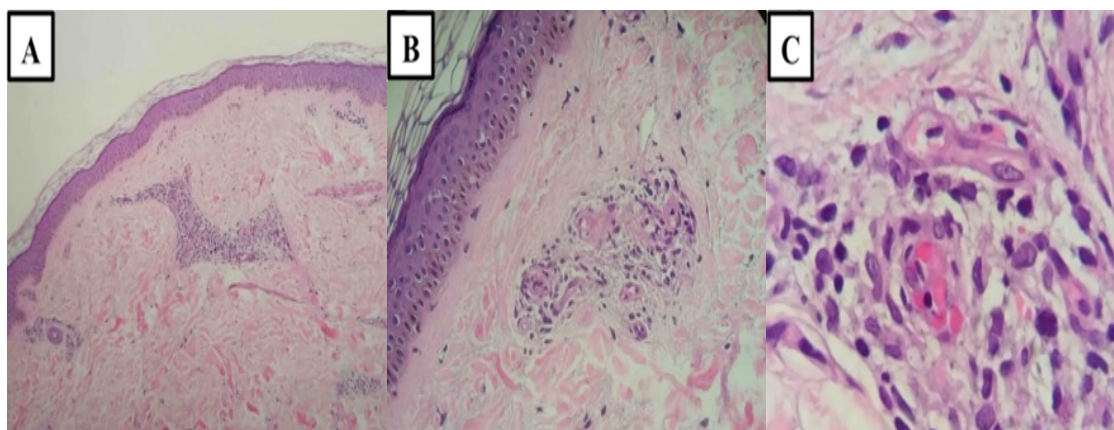


Figure 2. (A) Perivascular lymphocytic infiltrates in the upper dermis (H&E, 50X); (B) Endothelial cell swelling, superficial perivascular lymphocytes (H&E, 100X); (C) Erythrocytes extravasation around the superficial blood vessel (H&E 400X).

3 DISCUSSION

The clinical appearance of pigmented purpuric dermatoses may be similar to cutaneous vasculitis. However, the purpuric patches in cutaneous vasculitis are palpable, unlike in PPD. (Devere, 2012) Our patient was initially assessed as polyarteritis nodosa (PAN), a necrotizing vasculitis of medium-sized vessels that commonly affected the nerves, intestinal tract, heart, and joints. Indeed, the initial manifestations of some cases of PAN are skin lesions. (Kazandjieva, 2017) However, she did not complain of constitutional symptoms, such as fever, malaise, weight loss, or abdominal pain, and did not exhibit characteristic subcutaneous nodules. Therefore, it was important to rule out related systemic diseases, since the patient also complained of ankle pain. There were several case reports associating persistent PPD with CTCL and mycosis fungoides, which can all be excluded by histopathologic examination. (Toro, 1997, Martinez, 2001)

The reticular pattern of the macules and exacerbation by cold suggested the possibilities of disturbance of blood flow to the skin in response to

low temperature exposure, such as seen in livedo reticularis. Occlusions of vessels in livedo reticularis may be found in vasculopathy disorders, such as antiphospholipid syndrome (APS) and cryoglobulinemia. (Gibbs, 2005) The negative results of HBsAg, anti-HCV, HIV, mixed APTT, and lupus anticoagulant examinations of this patient ruled out those diseases. Anti-nuclear antibodies were positive, however it does not entirely confirm the presence of an autoimmune disease. (Pisetsky,

2011) Our patient's history and clinical appearance does not support an autoimmune disease, but continuous observation is needed.

Typical histopathologic findings of PPD mentioned in literatures are perivascular infiltrate of lymphocytes in superficial dermis, endothelial cell swelling, erythrocytes extravasation, and hemosiderin deposition. (Devere, 2012, Kim, 2015) Superficial perivascular lymphocytes were clearly observed in our patient, as well as endothelial swelling and extravasation of erythrocytes in the dermis. However, in two biopsy specimens we could not find deposition of hemosiderin. Although it is an important feature for distinguishing PPD from other disorder, hemosiderin deposition is not always found. In a retrograde analysis of 113 patients with PPD in Korea, hemosiderin deposition was found in only 26.3% subjects, while perivascular lymphocyte infiltration and erythrocyte extravasation were found in 79% and 50% patients, respectively. (Kim, 2015) It is advisable to do histochemical staining with Perls stain to identify hemosiderin more easily, as well as with Masson-Fontana stain to exclude melanin pigment. (Kazandjieva, 2017) Focal karyorrhectic nuclear dust may occasionally be found, especially in active pronounced lesions, alongside with narrowing of vessel lumens. (Kazandjieva, 2017, Weedon, 2010) Both features were found in this patient.

The patient was treated with antioxidants and symptomatic therapies, which consist of emollient, anti-inflammatory and antipruritic agents. Treatment of PPD is difficult, and results in literatures are inconsistent. (Hoesly, 2009) proposed a systematic approach for the management of Majocchi Purpura. Through review of available literatures, the scheme

suggested treatment of this disorder according to the symptoms severity and use of combination treatment if needed.

4 CONCLUSION

We reported a case of Majocchi purpura in a patient who was initially assessed as a polyarteritis nodosum. Although uncommon, it is important for dermatologists to be able to recognize the characteristic PPD lesions, and perform the necessary examinations to rule out other suspected disorders. Histopathology examination is until now the most important tool to confirm the diagnosis of PPD. Pigmented purpuric dermatoses is highly recurrent, although some lesions are self-limiting. Treatment is seldom satisfactory, and remains a challenge for clinicians.

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