

Extensive Purpura Annularis Telangiectodes of Majocchi in an Elderly Woman: Diagnostic Challenges in a Resource-Limited Hospital

Review began 10/05/2026

Review ended 10/25/2026

Published 10/27/2026

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DOI: 10.7759/cureus.118680

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Abstract

Purpura annularis telangiectodes of Majocchi is a rare variant of pigmented purpuric dermatoses that primarily affects children and young women. It is characterized by asymptomatic, reddish-brown annular macules and patches, predominantly on the lower extremities. Its pathogenesis is not fully understood. Diagnosis is primarily clinical, although biopsy may be useful to rule out differential diagnoses. There is no established curative treatment, and therapeutic options yield variable responses.

We report the case of an 88-year-old woman with a five-year history of progressively spreading, mildly pruritic annular purpuric lesions, initially affecting the lower extremities and subsequently involving the trunk and upper extremities. Treatment prior to our outpatient visit consisted of corticosteroids, phototherapy, and antibiotics, which proved ineffective. Examination revealed annular erythematous-violaceous, non-blanching macules and patches with central clearing and residual hyperpigmentation. Histopathology demonstrated a superficial perivascular lymphocytic infiltrate, vascular dilation, and erythrocyte extravasation, consistent with pigmented purpuric dermatosis. Dermoscopy revealed coppery pigmentation, supporting the diagnosis of purpura annularis telangiectodes of Majocchi. The atypical late-onset and near-generalized distribution prompted consideration of important mimickers, which were excluded histopathologically. The cornerstone of treatment was the initiation of colchicine; in addition, the use of adjuvant therapies, such as ascorbic acid, compression stockings, and emollients, led to a marked improvement in the lesions and pruritus after three months. This case highlights the importance of recognizing atypical presentations of this chronic, benign condition and avoiding unnecessary immunosuppressive therapy.

Categories: Pathology, Internal Medicine, Dermatology

Keywords: cutaneous histopathology, pigmented purpuric dermatosis, purpura annularis telangiectodes, resource-limited settings, treatment outcome

Introduction

Annular telangiectatic purpura is a rare subtype of pigmented purpuric dermatosis (PPD) [1]. It generally affects children and young women, with no recognized ethnic predilection [2,3]. It typically presents as asymptomatic, symmetrical annular reddish-brown macules and patches that primarily involve the lower extremities; however, atypical forms of extension to the trunk or unilateral presentation have been reported [3-5].

Although its pathogenesis remains poorly understood and most cases are considered idiopathic, several mechanisms have been proposed, including capillary fragility, gravitational forces, venous insufficiency, focal infections, and contact hypersensitivity. In addition, many patients report a history of activities that are associated with sustained lower limb pressure, such as prolonged standing at work, high-intensity sports, or repetitive physical tasks, suggesting that mechanical stress may contribute to disease development in susceptible individuals [6]. Other case reports have described the disease as being inducible by vaccination, sclerotherapy, ionized radiocontrast medium, and the initiation of biological therapy [7-12].

Although the diagnosis is primarily clinical, a skin biopsy may be useful for ruling out mimickers or malignancies such as annular elastolytic giant cell granuloma, trichophytic granuloma, or T-cell lymphoma. Histopathologic features of PPD include perivascular lymphocytic infiltrates, endothelial edema, erythrocyte extravasation, and hemosiderin deposition. No curative treatment has been established; management relies on symptomatic relief using corticosteroids, flavonoids, cyclosporine, and phototherapy, with variable responses [6].

Case Presentation

An 88-year-old woman presented to the dermatology outpatient clinic with a five-year history of progressively enlarging purpuric lesions. During this period, she was evaluated and treated by several dermatologists with corticosteroids and phototherapy, without clinical recovery. She also received empirical

How to cite this article

Viquez M M, Melgar Cossich V B, García-Mayorca G (October 27, 2026) Extensive Purpura Annularis Telangiectodes of Majocchi in an Elderly Woman: Diagnostic Challenges in a Resource-Limited Hospital. Cureus 18(10): e118680. DOI 10.7759/cureus.118680

treatment with second-generation cephalosporins in an outpatient setting, without improvement. The eruption initially affected the lower extremities and gradually spread to the lower trunk. The patient reported mild to moderate pruritus but denied pain, fever, and constitutional symptoms. The patient reported a medical history of hypertension, hypothyroidism, and osteoporosis, managed with irbesartan, hydrochlorothiazide, triamterene, levothyroxine, and ibandronate. She denied history of prolonged standing, repetitive walking, wearing high-heeled shoes, occupational activities involving prolonged weight-bearing, or recognized risk factors for chronic venous insufficiency or repeated lower-extremity trauma.

Physical examination was unremarkable except for the presence of multiple annular erythematous-violaceous non-blanchable macules and patches that extended centrifugally with a central clearing; some of the lesions left residual hyperpigmentation without significant atrophy (Figure 1). The lesions were predominantly distributed over the inferior extremities, trunk, and superior extremities. Dermoscopic examination revealed a diffuse yellowish-to-coppery background with numerous erythematous dots and sparse superficial telangiectasias (Figure 2).

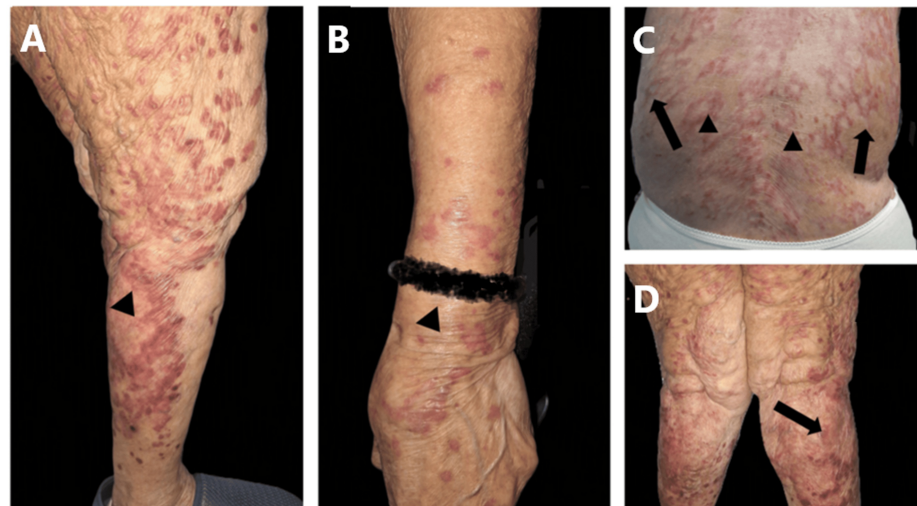


FIGURE 1: (A-D) Multiple petechial and purpuric patches with annular (arrowhead) and polycyclic (arrow) configurations.

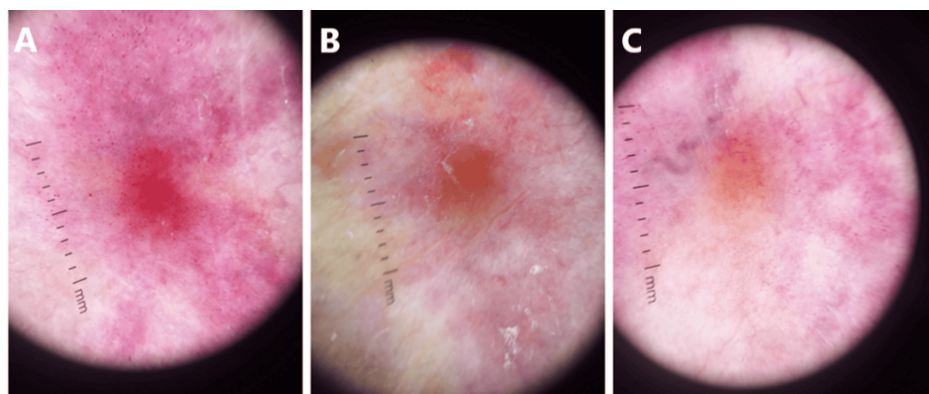


FIGURE 2: (A-C) Dermoscopic examination revealed a pinkish-to-yellow background with peripheral red dots, as well as superficial whitish areas and some telangiectasias.

Initial laboratory tests, including a complete blood count, revealed no abnormalities. Autoimmune testing, including antinuclear antibodies (ANA) and complement levels, was normal. Urinalysis was unremarkable, without evidence of urinary tract infection or proteinuria, and the metabolic panel was within normal limits. Potassium hydroxide (KOH) preparation and fungal cultures yielded negative results.

A punch biopsy was obtained from a lesion on the back. Histopathological examination demonstrated mild epidermal spongiosis and a mild superficial perivascular chronic inflammatory lymphocytic infiltrate with

vascular dilatation, congestion, and erythrocyte extravasation, findings consistent with purpura (Figure 3). Hemosiderin staining was not available.

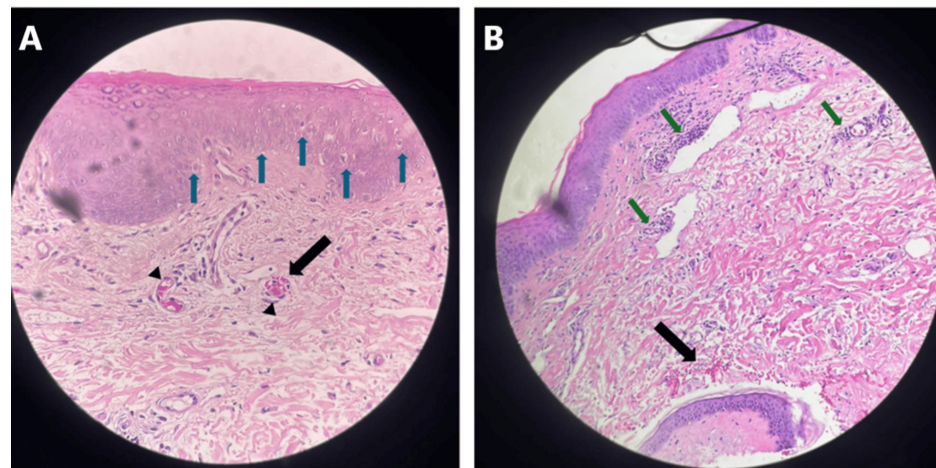


FIGURE 3: (A, B) Mild spongiosis (blue arrows); mild superficial perivascular chronic inflammatory lymphocytic infiltrate (green arrow) with dilation, congestion (arrowhead), and erythrocyte extravasation (black arrows). H&E (20x).

The patient was treated with oral colchicine (0.6 mg/day), ascorbic acid (1 g/day), graduated compression stockings, and regular application of barrier emollients. At the three-month follow-up, the patient demonstrated a marked reduction in the number of purpuric lesions, significant improvement in pruritus, and partial clearing of the plaques (Figure 4). Residual post-inflammatory pigmentary changes, without evidence of disease progression.

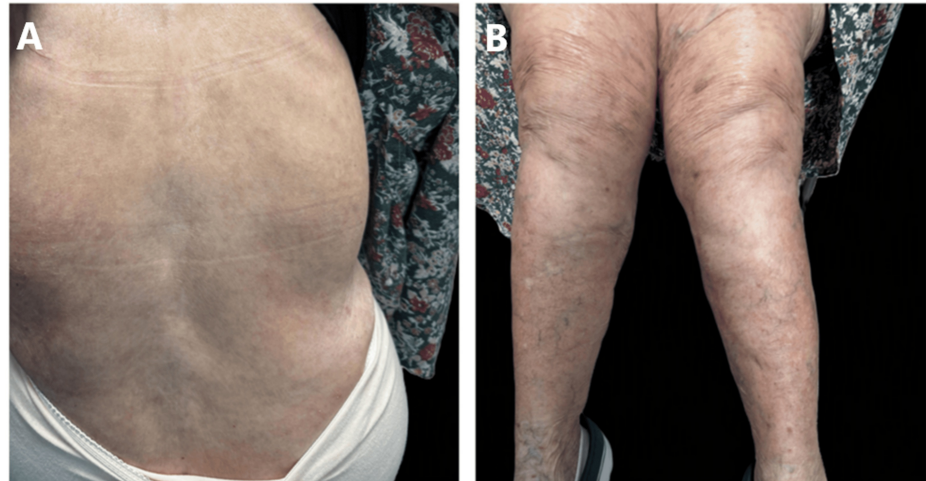


FIGURE 4: (A, B) Follow-up after treatment initiation.

Discussion

Purpura annularis telangiectodes of Majocchi (PATM) is one of five chronic, benign PPD variants distinguished by clinical morphology despite shared histopathologic features [1,13]. Although it predominantly affects the lower extremities of young women, our 88-year-old patient had near-generalized lesions involving the back, abdomen, breasts, and upper and lower extremities, an atypical late-onset presentation characterized by extensive cutaneous involvement that may be underrecognized. Although presentations in elderly patients are scarce, cases with atypical presentations have been described [3,4]. Widespread forms have mainly been described in association with triggers such as vaccination [7]. In our patient, no vaccination, radiocontrast exposure, sclerotherapy, or biological therapy preceded the eruption [7-12].

Although the diagnosis of Majocchi purpura is often based on its characteristic clinical presentation, a skin biopsy may be required when lesions are atypical or when important mimickers are considered. In our patient, the annular morphology prompted a differential diagnosis that included annular elastolytic giant cell granuloma, trichophytic granuloma, and cutaneous T-cell lymphoma. Histopathological examination was essential to exclude these entities [13]. The characteristic findings of a superficial perivascular lymphocytic infiltrate, erythrocyte extravasation, hemosiderin deposition, and dilated capillaries, in the absence of fibrinoid necrosis or neutrophilic nuclear debris, supported the diagnosis of Majocchi purpura and distinguished it from leukocytoclastic vasculitis. Likewise, the absence of granulomatous inflammation, fungal elements, and atypical lymphoid infiltrates effectively ruled out annular elastolytic giant cell granuloma, trichophytic granuloma, and cutaneous T-cell lymphoma, respectively. Additionally, there was no proteinuria, hypocomplementemia, or ANA suggestive of autoimmune processes.

Histopathological examination of all PPD shared blood vessel dilation, swelling of the endothelial cells, perivascular lymphocytic infiltration, erythrocyte extravasation, and hemosiderin deposition [1]. In our case, hemosiderin deposits were not reported because neither Prussian blue nor Fontana-Masson staining was available at our hospital. However, as reported by Kim et al. in their retrospective analysis, the presence of hemosiderin deposits was noted in only 26.3% of patients, despite the availability of the stains; in contrast, perivascular lymphocyte infiltration and erythrocyte extravasation were observed in 79% and 50% of cases, respectively [14]. Nevertheless, our dermatoscopic evaluation revealed coppery pigmentation supporting hemosiderin deposition [15,16].

Treatment is largely supportive, as no curative approach exists [1]. Our patient had a favorable clinical course after the initiation of colchicine, with marked improvement of the skin lesions, which may be related to its immunomodulatory effects [1,17,18]. Additionally, ascorbic acid may exert hemorheological effects by reducing reactive oxygen species and enhancing vascular wall strength through the promotion of collagen synthesis [19,20]. However, given the simultaneous use of pharmacological and non-pharmacological measures in this patient, we cannot determine whether the initiation of these two agents was responsible for the significant clinical improvement of the skin lesions. The chronic but benign course underscores the importance of avoiding unnecessary immunosuppressive therapy [1].

Conclusions

Although PATM typically affects younger individuals, its characteristic clinical features should prompt consideration of the diagnosis even outside the usual age group. This pathology should be considered in elderly patients presenting chronic annular purpuric lesions, particularly when systemic symptoms are absent. Accurate diagnosis depends on integrating clinical, dermoscopic, and histopathologic data. As a benign but chronic entity, PATM requires long-term follow-up to monitor its course and improve patients' quality of life. Supportive and microcirculatory therapies may reduce disease activity and improve symptoms. In our case, clinical improvement was observed following a combined therapeutic approach that included colchicine and ascorbic acid; however, because several treatments were administered simultaneously, the contribution of each agent cannot be determined. For this reason, the favorable tolerability and low-cost approach may be useful in refractory or extensive disease, while early recognition helps prevent misdiagnosis and unnecessary interventions.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

Concept and design: Marjorie M. Víquez, Verónica B. Melgar Cossich, Gabriel García-Mayorca

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Disclosures

Human subjects: Informed consent for treatment and open access publication was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have

declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

Acknowledgements

We sincerely thank the patient for providing informed consent for the publication of this case and for her valuable contribution to this report. We also acknowledge the Pathology Department of Saint Thomas Hospital for their contribution to the diagnostic confirmation.

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