

Extensive annular petechial eruption in an elderly man with Parkinson's disease: A clinicopathological challenge

Case Presentation

A man in his sixties presented with a mildly pruritic, annular-looking petechial rash that first appeared on his lower limbs 2 years ago. The rash progressed upwards to his anterior trunk and cheeks. He had Parkinson's disease and was started on co-formulated levodopa and benserazide, a few months before the rash appeared. He had no systemic symptoms, anorexia, or weight loss. His other medications include ropinirole and selegiline, both of which were started a year before rash onset.

Examination revealed extensive annular-looking petechial macules involving the lateral cheeks, neck, trunk, and both

upper and lower limbs [Figures 1a and 1b]. His back and buttocks were spared. Punch biopsies of the macules on both the patient's neck and abdomen showed superficial perivascular lymphohistiocytic infiltrates admixed with plasma cells, scattered eosinophils, and extravasated red blood cells [Figure 1c]. A few haemosiderin pigments were also identified on Perl's stain. No dysplasia or epidermotropism of lymphocytes was seen.

Question

What is your diagnosis?



Figure 1a: Annular petechial macules involving the anterior trunk.



Figure 1b: Close-up photograph of the neck and left lateral cheek, showing lesions of similar morphology.

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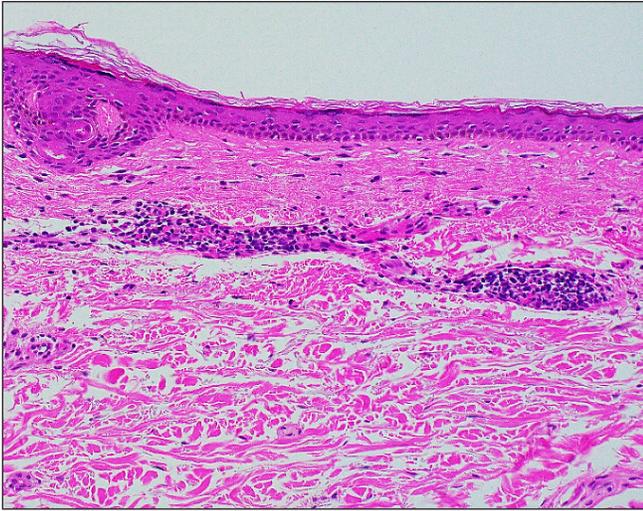


Figure 1c: Histology from the macules demonstrating superficial perivascular lymphohistiocytic infiltrates with plasma cells, scattered eosinophils, and extravasated red blood cells (Haematoxylin & Eosin, 200 \times).

Answer

Diagnosis: Purpura annularis telangiectoides of Majocchi

Discussion

Purpura annularis telangiectoides of Majocchi (PATM), also known as Majocchi's disease, is a subtype of pigmented purpuric dermatoses (PPD), a group of histopathologically similar capillaritis that differs by their clinical morphology. Our case illustrates a patient with an annular-looking petechial eruption but with atypical features of extensive involvement of the trunk and face after the initiation of co-formulated levodopa and benserazide. Given the temporal relationship and the presence of eosinophils, a drug reaction could be a possibility. This patient had a platelet count of $294 \times 10^9/L$.

PATM, first described in 1896, is an uncommon condition that tends to affect children and young females.¹ It presents as bluish-to-red macules with dark-red telangiectatic spots and the centrifugal extension of these macules gives it its characteristic annular configuration. Dermoscopy shows irregular round-to-oval red dots, globules, and structureless areas with diffuse ill-defined brownish-red homogenous areas [Figure 1d]. Like most PPDs, PATM predominantly involves the lower limbs with extension to the upper extremities symmetrically, although an atypical unilateral presentation of PATM has been reported.² Extensive involvement including the trunk and face, as in this case, has not been reported.

PATM shares similar histopathology features with the other PPD subtypes: dilated blood vessels, erythrocyte extravasation, haemosiderin deposition, and perivascular lymphohistiocytic infiltration. In a retrospective review by Huang *et al.*, all inflammatory reaction patterns (lichenoid, perivascular, interface, spongiotic, granulomatous) can be seen in PPD but only lichenoid and perivascular patterns were found in PATM.³

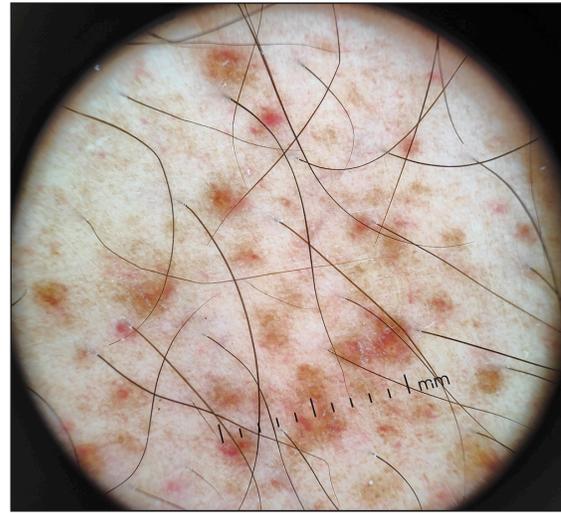


Figure 1d: Dermoscopy showing irregular round-to-oval red dots, globules, and structureless areas with ill-defined brownish-red homogenous areas.

The pathogenesis of PATM and other PPD remains unknown, although venous hypertension, gravity, capillary fragility, and focal infections have been proposed as contributing factors. Drug associations with analgesics, oral hypoglycaemic agents, and anti-hypertensive agents have been reported, in particular with Schamberg disease.⁴ While neuroactive agents (e.g. carbromal, chlordiazepoxide, lorazepam, meprobamate) have been reported, drugs used for Parkinson's disease, as in this case, have not been previously associated.

PATM should be differentiated from other causes of petechiae such as leukocytoclastic vasculitis with annular morphology, idiopathic thrombocytopenic purpura, PPD-like mycosis fungoides, and purpuric pityriasis rosea [Table 1]. There are also reports that PPD may mimic or progress to cutaneous T-cell lymphoma (CTCL).^{1,5} In 56 patients with persistent PPD, Toro *et al.* found 29 patients (52%) with histological features of CTCL.⁵ Hence, histological diagnosis should always be pursued in patients with atypical or persistent presentations of PPD.

While PATM is a benign condition, the absence of guidelines and the persistent nature of this condition make management challenging. Conservative or non-pharmacological management (leg elevation, compression stockings) can be considered for asymptomatic patients.¹ Suspected culprit drugs should be stopped where feasible. For symptomatic patients with PPD, treatment options include topical agents, such as corticosteroids and calcineurin inhibitors. Other treatment options have been described with variable success, including intralesional triamcinolone, oral agents (rutoside, ascorbic acid, pentoxifylline, venoactive agents, e.g., diosmin, calcium dobesilate, colchicine, griseofulvin, cyclosporine, methotrexate), phototherapy (narrow-band ultraviolet B phototherapy, psoralen plus ultraviolet A photochemotherapy) and laser therapy.^{1,4}

Table 1: Distinguishing features between selected conditions

Feature	Purpura annularis telangiectodes of Majocchi	Idiopathic thrombocytopenic purpura	Leukocytoclastic vasculitis with annular morphology	PPD-like mycosis fungoides	Pityriasis Rosea
Age	5 th decade	3 rd to 4 th decade	All ages	After 50 years of age	2 nd to 3 rd decade
Gender predilection	Male	Female	None	Male	None
Location	Lower limbs	Lower limbs	Lower limbs	Lower limbs (but usually more widespread)	Trunk with a herald patch Proximal limbs (Langer's lines)
Pruritus	None to mild	None to mild	None to mild	Yes	Yes
Palpable lesions	No	No	Yes	No	Yes (Papulosquamous)
Other bleeding manifestations	No	Yes (other mucocutaneous sites)	No	No	No
Associations	Venous insufficiency, exercise, RA, SLE, CLL, hyperlipidaemia, drugs (NSAID, statins, CCB, ACEi, beta-blockers)	Infections (HIV, hepatitis C, CMV, VZV), drugs, SLE, APS, HIV, CVID	Infections (<i>Streptococcus pyogenes</i> , bacterial endocarditis, HIV, hepatitis, tuberculosis), malignancy (paraneoplastic), drugs (antibiotics, thiazides, phenytoin, allopurinol, warfarin, NSAID), autoimmune (SLE, dermatomyositis, RA)	Progression from PPD	Infections (HHV-6, HHV-7), medications (vaccines, gold, captopril, barbiturates, penicillamine, clonidine)
Histology	Small vessel perivascular lymphohistiocytic inflammation with endothelial cell swelling, no leukocytoclasia, extravasated red blood cells (RBC) with variable haemosiderin deposition in macrophages	Erythrocyte extravasation in the absence of vascular inflammation	Perivascular neutrophils with fibrinoid necrosis, some extravasated RBC, leukocytoclasia	Epidermotropism with infiltration by atypical lymphocytes	Subacute spongiotic dermatitis with perivascular lymphocytic infiltrates, focal extravasation of RBC without leukocytoclasia

Legend: ACEi: Angiotensin-converting enzyme inhibitors, APS: Antiphospholipid syndrome, CCB: Calcium channel blockers, CLL: Chronic lymphocytic leukaemia, CMV: Cytomegalovirus, CVID: Common variable immunodeficiency disorders, HHV: Human herpesvirus, HIV: Human immunodeficiency virus, NSAID: Non-steroidal anti-inflammatory drugs, PPD: Pigmented purpuric dermatosis, RA: Rheumatoid arthritis, RBC: Red blood cells, SLE: Systemic lupus erythematosus, VZV: Varicella zoster virus

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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Net Quiz 1

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Summary: A man in his sixties with Parkinson's disease developed an annular, pruritic petechial rash on his lower limbs progressing to trunk for two years. Biopsies showed perivascular lymphohistiocytic infiltrates with eosinophils and extravasated red blood cells. A diagnosis of Purpura annularis telangiectodes of Majocchi (PATM) was made. PATM, a subtype of pigmented purpuric dermatoses, presents typically on lower limbs and rarely extends to trunk and face. Management includes conservative measures and topical agents. This case highlights an atypical PATM presentation and the importance of histological diagnosis for patients with atypical presentations.