



Correspondence

Purpura annularis telangiectodes: A rare entity in pediatric age group

Dear Editor,

Pigmented purpuric dermatosis (PPD) is a heterogeneous group of disorders characterized by petechiae and skin pigmentation. These dermatoses are presumably due to capillaritis and minimal bleeding from the dermal vessels.^[1]

PPD has various clinical patterns which can be classified as follows: progressive pigmentary dermatosis or Schamberg's disease, pigmented purpuric lichenoid dermatitis, eczematoid-like purpura, lichen aureus, disseminated pruriginous angiodermitis, unilateral linear capillaritis, granulomatous pigmented purpura, and purpura annularis telangiectodes (PAT) of Majocchi.^[2]

PAT is one of the rarest forms of PPD and presents clinically as macules that progress to annular lesions with the presence of petechiae and hyperpigmentation. PAT usually occurs in adolescence and particularly in women. However, it is extremely rare in childhood.^[1]

A 5-year-old boy presented to the emergency department with a 2-month history of asymptomatic macules showing a progressive increase in the number and size of lesions. The patient had no remarkable medical history and no drug intake before the appearance of the skin lesions. The last vaccination received was the mumps-measles-rubella vaccine 3 years previously. No other recent vaccines were identified as possible triggers.

Physical examination revealed annular lesions with well-defined edges and an erythematous-brownish color with a lightened center. The lesions were located on the trunk and upper right arm and the front and back of the legs. A polarized light dermoscopy showed the presence of globules and reddish spots, with areas of coppery-brownish pigmentation^[3] [Figure 1].

Analytical studies included biochemistry, blood count, coagulation study, and immunoglobulin, autoimmunity, and urine sediment analysis. All were without significant findings. Serological tests for hepatitis B, hepatitis C, and HIV were negative.

A skin biopsy taken for anatomopathological study from a lower left leg lesion showed an epidermis of normal thickness with mild hyperkeratosis and vacuolation of the basal layer. Similarly, the small-caliber vessels in the papillary dermis showed perivascular lymphocytic infiltration and extravasation of red blood cells. Perls' Prussian blue

stain showed hemosiderin pigment deposits in the dermal macrophages [Figure 2].

These clinical findings, dermoscopic features, and analytical and histological studies enabled the diagnosis of PAT.

The therapeutic recommendations included the application of topical corticoids (methylprednisolone aceponate 0.1%) twice a day. The lesions presented a complete resolution at 6 weeks.

The diagnosis of PPD is based on a correct clinical history that looks for possible triggers, the evolution of the clinical picture, and a physical examination identifying the different symptoms. However, the diagnosis can be supported with a few compatible dermoscopic features and certain analytical studies that rule out systemic characteristics, as well as histopathologic findings that confirm the proposed diagnosis.^[1,4]

Only three cases of childhood PAT have been reported in the medical literature.^[5-7] Of these three, one is a familiar case of PAT in a 2-month-old newborn male,^[5] another is that of a 10-year-old girl,^[6] and the last is a 10-year-old boy whose condition was treated with ascorbic acid and rutoside with good progress.^[7] In all cases, the lower extremities were affected, and no possible precipitating factor was identified^[5-7] [Table 1].

The differential diagnosis of PAT includes cutaneous vasculitis, nummular dermatitis, contact dermatitis, stasis dermatitis, Kaposi's sarcoma, and cutaneous lymphomas.^[4] The etiopathogenesis of this disease is unknown, but various triggers have been proposed such as exercise, venous stasis, capillary fragility, hypertension, infections, alcohol consumption, administration of various drugs or chemicals, and allergic contact dermatitis.^[2]

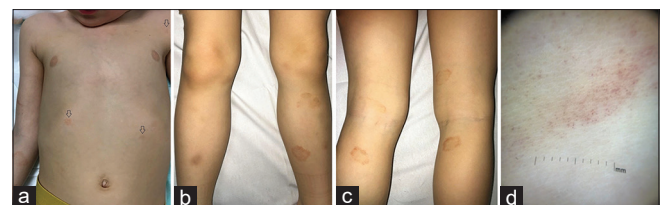


Figure 1: (a-c) Brownish erythematous annular lesions located on the trunk and limbs. (d) Globules and reddish spots are evident, with areas of brownish pigmentation (polarized light dermoscopy, $\times 10$).

Table 1: Clinical cases of PAT in the pediatric age group

Authors	Year	Age	Sex	Time evolution of disease	Triggers	Treatment	Resolution	HP
Our case	2019	5 years	Male	2 months	No triggers	No treatment	6 weeks	Yes
Goyal T <i>et al.</i>	2014	10 years	Male	4 years	No triggers	Vitamin C 500 mg/day and rutoside 50 mg/day orally	Significant regression 12 weeks	Yes
Ozturk P <i>et al.</i>	2006	10 years	Female	3 years	No triggers	-	-	Yes
Honda H <i>et al.</i>	1997	2 months	Male	1-2 months	No triggers	No treatment	6 months	Yes

HP: Histopathology, PAT: Purpura annularis telangiectodes

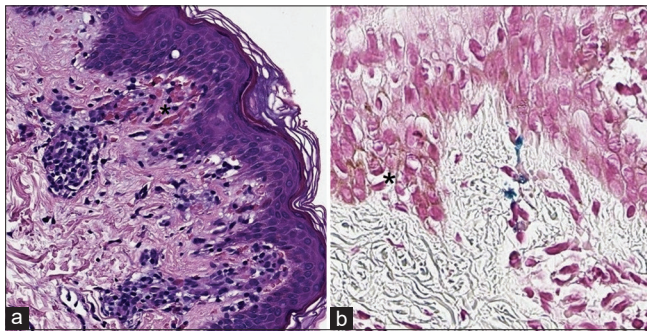


Figure 2: (a) Perivascular lymphocytic infiltration is evident in the small-caliber vessels of the papillary dermis, as well as an extravasation of red blood cells. (H and E staining $\times 20$). (b) Histochemical staining shows hemosiderin pigment deposits in dermal macrophages (Perls' Prussian blue $\times 20$).

The resolution may be spontaneous, but various treatments, such as topical corticosteroids, topical calcineurin inhibitors, antihistamines, ascorbic acid, rutoside, phototherapy, pentoxifylline, colchicine, griseofulvin, and cyclosporine, have been used, depending on the extent and refractoriness of the lesions.^[8] However, the current evidence as to the efficacy and safety of these treatments in infancy is limited, due to the scant number of published cases.

Pigmented purpuric eruptions are a group of dermatoses of idiopathic etiology with no systemic characteristics. In spite of this, sometimes, the rarest forms require a thorough patient evaluation to rule out vasculitic or thrombocytopenial processes. These dermatoses tend to be chronic, but they also spontaneously resolve over time in a variety of ways. Majocchi's disease, also known as PAT, is one of the rarest forms of PPD in children, and few therapeutic alternatives are available for patients of this age.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the parents have given their consent for images and other clinical information to be reported in the journal. The parents understand that names and initials will not be published, and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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