

Unusual Presentation of Purpura Annularis Telangiectodes of Majocchi Responding to PUVA Sol

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ABSTRACT

Purpura annularis telangiectodes of Majocchi (PATM) is characterized by punctate telangiectatic macules progressing to annular, hyperpigmented patches with central clearing and infrequent atrophy. It is most common in young females. Here we present a case of a 29 year old female, housewife who presented to our OPD with complaint of brownish black lesions on neck, shoulders, upper chest, back and legs from past 1 year and responded to oral PUVA Sol therapy.

Keywords: PUVA, Dermoscopy, Pigmented purpuric dermatosis

INTRODUCTION

In 1896 Domenico Majocchi observed the unusual hemorrhagic eruption which now bears his name. He called this disorder "purpura annularis telangiectodes".

¹ Pigmented purpuric dermatosis is a group of vascular disorders with varied clinical manifestations. They are chronic, progressive, cause concern, and are resistant to treatment.² The term pigmented purpuric dermatoses includes Schamberg's disease (i.e., progressive pigmentary dermatosis), purpura annularis telangiectodes (Majocchi disease), lichen aureus, itching purpura, eczematid like purpura of Doucas and Kapetanakis, and the pigmented purpuric lichenoid dermatosis of Gougerot and Blum.³

Majocchi disease is characterized by punctate telangiectatic macules progressing to annular, hyperpigmented patches with central clearing and infrequent atrophy. It is most common in young females. The lesions are usually symmetrically distributed on the legs and are occasionally symptomatic.⁴

We present a case of Majocchi disease with an unusual presentation who responded to oral PUVA Sol.

CASE REPORT

A 29 year old female, housewife presented to our OPD with complaint of brownish black lesions on neck, shoulders, upper chest, back and legs from past 1 year. The lesions were believed to begin as a bruise which gradually increased in size to show central clearing of the lesions, they were associated with mild itching.

Family history was insignificant.

The patient was averagely built and well nourished, conscious, alert and oriented to time, place and person. Systemic examination was within normal limits.

On cutaneous examination, the lesions were found to be brown in colour, irregularly shaped with typical 'cayenne paper' spots in the centre and over the edges. ([figure: 1](#))



Figure 1: multiple irregular brown colored macules.

Her complete blood count, routine biochemical tests, erythrocyte sedimentation rate, ANA, viral markers and RPR were normal or negative.

Dermoscopy revealed presence of Coppery red pigmentation, Red globules, Brown dots, Reticular network, Brown globules, linear vessels and prominent follicular openings. (fig: 2a,b)

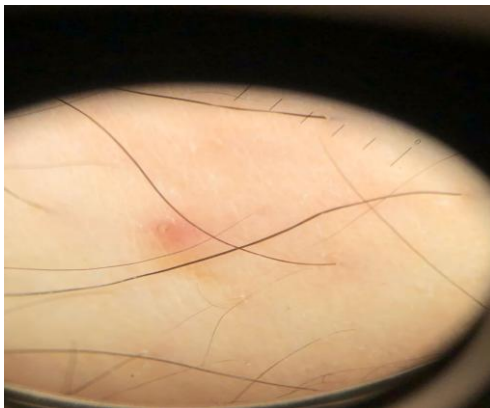


Fig: 2a) Dermoscopic picture showing red globules and reticular network.

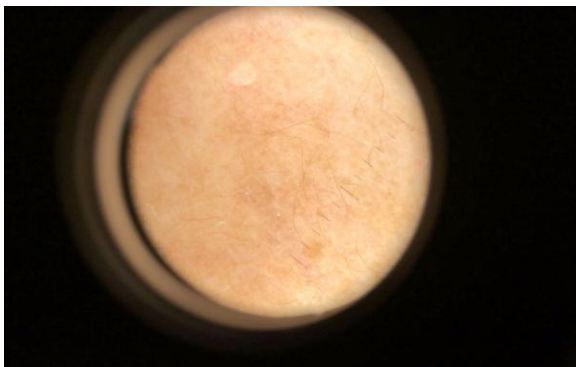


Fig: 2 (b) Dermoscopic picture showing brown globules, reticular network and prominent follicular openings.

With the above clinical, dermoscopic and histopathological findings the diagnosis of purpura annularis telangiectodes was confirmed.

Histopathological examination was consistent with Majocchi's disease and revealed hyperkeratosis, thinned out granular layer, follicular plugging, mild to moderate degree of lymphocytic infiltrate around dermal vessels along with occasional melanophage. Focal extravasation of RBC's was noted. (fig: 3 a,b)

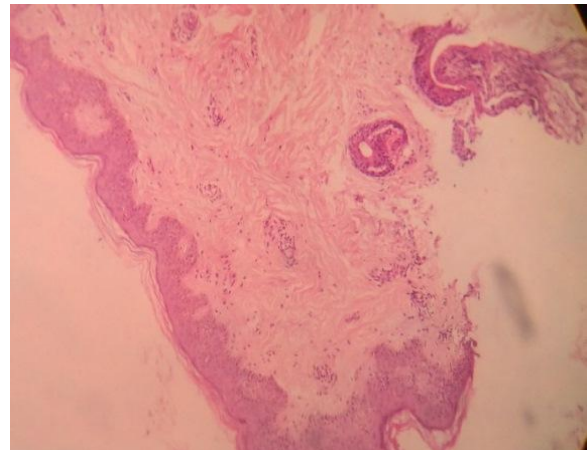


Fig: 3 (a) HPE examination shows hyperkeratosis, thinned out granular layer and mild lymphocytic infiltrate around dermal vessels.

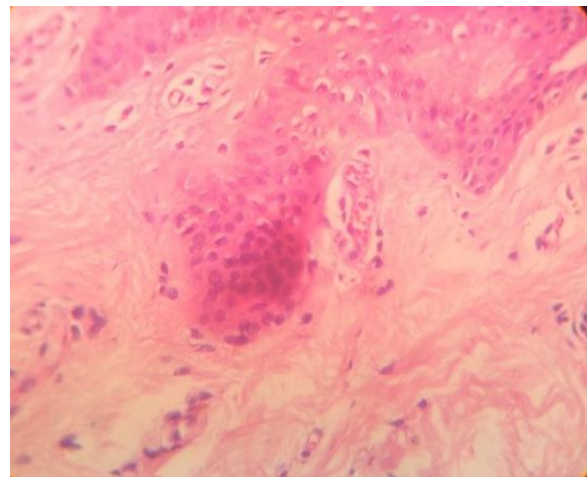


Fig: 3 (b) Focal extravasation of RBC's.

The patient was started on oral PUVA-sol in the dose of 0.8mg/kg three alternate days a week. The patient reported excellent relief in the follow up period after 1 month. (fig :4)



Fig: 4 follow up after 1 month.

DISCUSSION

PPD has five clinical types: Progressive pigmented purpuric dermatosis (Schamberg's disease), purpura annularis telangiectodes (Majocchi disease), pigmented purpuric lichenoid dermatosis (Gougerot-Blum disease), itching purpura (eczematid like purpura of Doucas and Kapetanakis) and lichen planus. Clinical overlap maybe observed between these types.⁵

Purpura annularis could be divided into 3 stages of evolution: (1) the telangiectatic, (2) the hemorrhagic pigmentary and (3) atrophic.¹

The etiology of the Majocchi disease is unknown. A hypothesis of the vascular pressure increase has been supposed because the lesions appear usually in the legs and buttocks.⁶

The eruption usually begins in the form of punctate capillary ectasias and hemorrhages. The color of the lesion varies from the usual bright red of the earliest stages to the darker red tint or rust color of the later stages. The eruptions usually appear first in the lower extremities, and are symmetrical in distribution.¹

Histological changes do not affect the epidermis but just beneath it the vessels are usually dilated, some showing aneurysmal distortion. Small cell infiltration, more abundant about the widely dilated vessels, is noted, and some of the

subdermal arterioles show an obliterating endarteritis.⁷

Dermoscopy shows irregular, round to oval red dots, globules and patches with a red brownish or red coppery diffuse homogeneous background pigmentation. This pattern histologically represents hemosiderin deposition within the papillary dermis.⁸

Most forms of PPD do not require treatment. Compression stockings and leg elevation may be effective via a reduction in venous stasis and edema.⁹ Topical steroids are recommended. They contribute to treatment through their anti inflammatory effects.¹⁰ Methotrexate and PUVA have well known anti inflammatory properties. PUVA has previously also been used in various forms of PPD with beneficial effects.⁴

This case report will add upon the evidences for managing this refractory disease besides providing dermoscopic findings

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