CASE REPORT

Purpura annularis telangiectodes of Majocchi associated to vitiligo

Claudia Procel,* Gilda Zurita**

* *General Physician*
** *Dermatologist*

Correspondence: revistaclinicaeup@hotmail.com

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ABSTRACT

Purpura annularis telangiectodes of Majocchi is a uncommon form of presentation of progressive pigmented purpura, it is caused by autoimmune capillaritis showing the participation of CD4 +; CD3 + lymphocytes and Langerhans cells, also the increased early expression of ICAM-1, ELAM-1 adhesion molecules in keratinocytes near the areas of lymphocytic infiltrate (CD11a) in the epidermis. Majocchi's telengiectasis annular purpura is characterized by a hyperpigmented purple reddish annular macula in its periphery accompanied by telengiectasias and an atrophic whitish plaque base. These lesions are grouped or disseminated, and they do not disappear under vitrepressure, usually appears in lower limbs as a place of origin and tends to ascend to the trunk and upper limbs having a greater predisposition for young adult women. Majocchis purpura may be accompanied by moderate pruritus.

Likewise, vitiligo is an autoimmune disease triggered by various stimuli, in which cellular oxidative stress leads to an autoimmune response against melanocytes causing depigmentation of the skin.

We present the medical case of a 38-year-old female with a two year diagnosis of vitiligo who presents mild pruritus on a violaceous hyperpigmented circular lesions in her lower limbs of a one month evolution. Biopsy of the lesion is compatible with progressive pigmented purpuric dermatosis. Through clinical and histopathological characteristics of the lesion, the diagnosis of purpura annularis telangiectodes of Majocchi was made. The patient started oral treatment with 1000mg of vitamin C (ascorbic acid) per day for 30 days plus phototherapy (UVB) twice a week with a great outcome.

INTRODUCTION

Progressive pigmented purpuric eruptions are chronic, benign dermatosis, localized mainly on the lower extremities and characterized by the presence of petechiae and brownish skin pigmentation, without any blood abnormality, nor venous stasis.

Histologically it is caused by autoimmune capillaritis with the participation of CD4 +; CD3 + lymphocytes and Langerhans cells interacting with endothelial cells. These infiltrate is well-organized and in a defined manner. Immunohistochemistry analysis

demonstrate a strong expression of ICAM-1, ELAM-1 adhesion molecules in keratinocytes near the inflammatory infiltrated areas (CD11a) in the epidermis. Traditionally these dermatosis have been divided into 5 clinical groups: Schamberg disease or progressive pigmentary purpura, purpura annularis telangiectoides or Majocchi disease, pigmented purpuric lichenoid dermatosis of Gougerot and Blum, eczematoid-like purpura of Doucas and Kapenakis, and lichen aureus.

Purpura annularis telangiectodes or Majocchi disease, is an uncommon form of presentation of these progressive pigmented purpuric eruptions, which have a preference for the feminine sex and the lower extremities, characterized by purple reddish irregular circular patches accompanied by pruritus. It is a skin condition of a difficult diagnosis since it is very uncommon and there are many differential diagnosis to be analyzed first. It is also a therapeutic challenge, there is no medical treatment that has been proven entirely effective; a definite cure has not been found.
It is well known that vitiligo as an autoimmune condition is associated to other immune diseases. These association between purpura annularis telangiectodes of Majocchi and vitiligo has not been described in the literature. We present a medical case of a 38 year old female with a 2 year diagnosis of vitiligo that shows lesions compatible to progressive pigmented purpura.

CASE REPORT

Clinical History

A 38 year old female with a childhood diagnosis of atopic dermatitis, diabetes mellitus diagnosed 3 years ago and vitiligo 2 years ago. She reported family history of diabetes mellitus and hypertension. She comes to the consult presenting asymmetrical achromic circular patches on lower extremities accompanied by moderate pruritic purple reddish annular macules and papules that started on the posterior side of the legs spreading to the anterior side of the thighs.

Physical Exploration

The examination reveals purple reddish macules that group forming patches, that do not disappear to the digital pressure, with an annular and semiannular pattern. Some of the patches are on top of the vitiligo lesions diagnosed years ago.

COMPLEMENTARY STUDIES

A biopsy punch of one of the lesions was performed on the posterior side of the leg for a histological analysis.

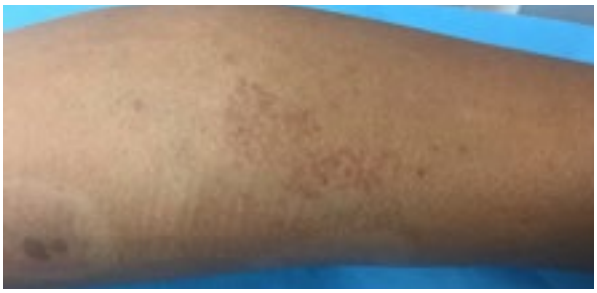

Figure 1. Macules on a circular pattern on the lower extremity of the patient.

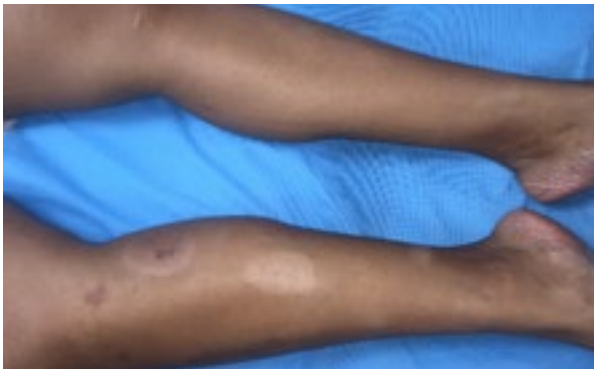

Figure 2. Purple reddish macules surrounded by achromic lesions compatible with vitiligo on lower extremities of the patient.

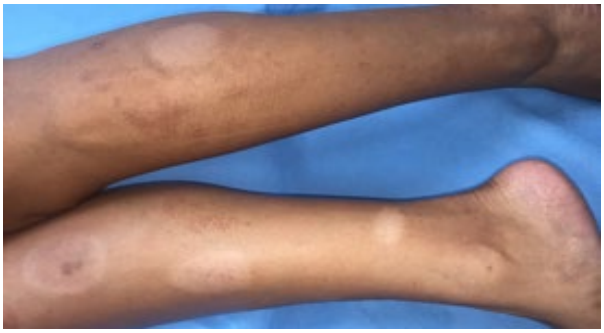

Figure 3. Purple reddish macules dispersed on both lower extremities.

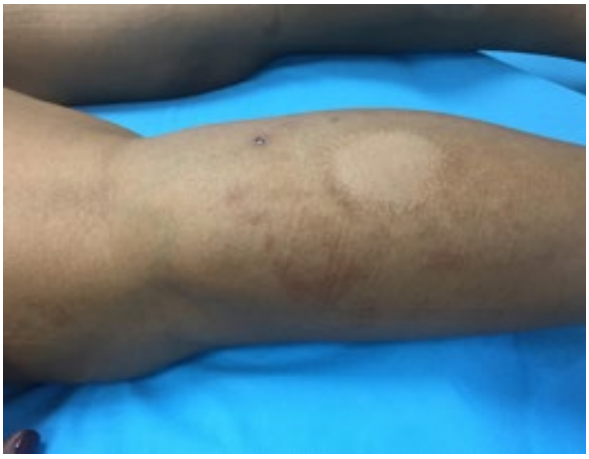

Figure 4. biopsy punch area for the skin sample.

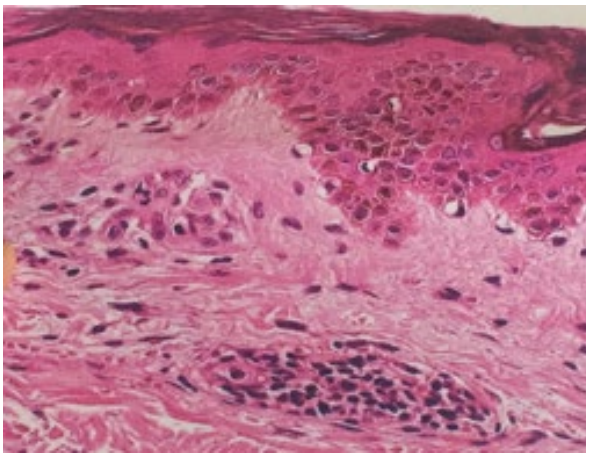

Figure 5. the microscopic examination demonstrate extravasated erythrocytes.

DIAGNOSIS

The histological alterations of the sample are compatible to pigmented purpuric dermatosis.

HISTOPATHOLOGY

The histopathological analysis demonstrate a normal skin structure. Mild irregular acanthosis and espongiosis on epidermis. Superficial perivascular lymphocytic infiltrate on dermis, as well as, extravasated erythrocytes.

EVOLUTION AND TREATMENT

We started with narrow-band UVB light therapy, daily intake of 1000mg of ascorbic acid and 50mg of rutoside, 25mg of hydroxyzine at night and topic application of 0.1% desonide.

The response to the treatment has been satisfactory, the macules on the lower extremities have decreased there pigmentation from purple reddish to brownish, as well a decrease in the intensity of the pruritus, showing some episodes of exacerbation of the pruritus on both extremities. The chronic evolution of this condition has been explained to the patient, as well as the diverse treatments that are available.

DISCUSSION

Both vitiligo and purpura annularis telangiectodes of Majocchi are skin conditions that develop due to an autoimmune response, specifically a hypersensitivity mediated by inflammatory cells.

Vitiligo develops on genetically susceptible individuals, triggered by various stimuli, from sun burns to mechanical direct trauma and exposure to chemical substances that they all initiate an autoimmune response against melanocytes causing depigmentation of the skin. The physiopathology of this condition involves two important processes: the first one is the excessive oxidative stress that works as the trigger and the second one is the autoimmune reaction as the main factor for the chronic evolution of this disease.

In vitiligo there is an abnormal reaction to the oxidative cellular changes. There are reports of an excess production of anti-oxidant enzymes like superoxide dismutase surrounding damaged tissues and in the serum, which incites the activation of the “heat-shock protein 70” (iHSP70) resulting in the progressive loss of melanocytes.

The sensor of cellular stress and development of the proteins takes place in the endoplasmic reticulum. Due to this oxidative stress the folding of the proteins is not done properly and this cluster of abnormal proteins activates the Unfolded Protein Response (UPR), which will induce apoptosis of the melanocytes and production if interleukins (IL 6 and 8) for the skin the immune response.

The UPR could be the link between the trigger and the activation of the autoimmune response in the progression of vitiligo. In fact, UPR also activates the immune response and plays a role in other autoimmune disorders like type I Diabetes and degenerative disorders.

In vitiligo the cytotoxic direct response of the melanocytes is produce by lymphocytes T CD8+, INF-y, that plays an important role in the dissemination of the lesions. There is evidence that proofs that IFN-y caused a higher expression of CXCL10, a chemokine that regulates the invasion of lymphocytes T CD8 into the epidermis and follicles, as well as the apoptosis of the melanocytes.

Some studies have indicate an increase in TNF-a associated to vitiligo. Webb et al observed that the inhibition of TNF-a was linked to the inhibition of the progression of vitiligo in three patients. He also explained that this effect could have been previously missed, since other studies only focused on the ability of the TNF-a promoting repigmentation.

The autoimmune response that has been seen in purpura annularis telangiectodes of Majocchi is produce by cells-mediated hypersensitivity since in the infiltrates many CD4+, CD3+ lymphocytes and dendritic cell have been identified interacting with the endothelial cells. These infiltrate is found in a well-organized and defined manner. Immunohistochemistry analysis demonstrate a strong expression of ICAM-1, ELAM-1 adhesion molecules in keratinocytes near the inflammatory infiltrated areas (CD11a) in the epidermis. These proofs the role of the T cells in the pathogenesis of progressive purpuric dermatosis. Other researches demonstrated that the early expression of adhesion molecules in the endothelial cells is what determines the perivascular infiltrate. These molecules regulate the path in or out of lymphocytes in the inflammatory tissue, and also regulate the interaction between lymphocyte and dendritic cells. At the same time the lymphocytes liberate cytokines, specially TNF-a that promotes the expression of adhesion molecules and causes the abnormal liberation of endothelial plasminogen activator factor and/or the excessive production of plasminogen activator inhibitor. This process would contribute to the decreased skin fibrinolysis and deposit of intraperivascular fibrin characteristic of Purpuric Dermatosis.

CONCLUSION

In both conditions the production of TNF-a is important for the progression of the disease, probably is this cytokine that allowed the durability of vitiligo in the patient, producing the depigmentation of the melanocytes, as well as the development of purpura annularis telangiectodes of Majocchi after many years. The cytokine affected the capillary. There is the possibility that this cytotoxic protein is the link between both conditions.

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