Case report

Idiopathic atrophoderma of Pasini and Pierini, an unusual approach

DOI: 10.5377/alerta.v6i2.15578

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Abstract
Idiopathic atrophoderma of Pasini and Pierini is a rare entity of unclear etiology, occurring as much as six times more frequently in women than in men, with a possible association with localized scleroderma (morphea). Case presentation. It is about a 30 years old woman who consulted with an asymptomatic lesion of two years of evolution on the left gluteal region. Physical examination revealed an oval plaque, depressed and acromic in its center, measuring five by ten centimeters. A 30 years old female patient who consulted about an asymptomatic lesion of two years of evolution on the left gluteal region. Physical examination revealed an oval plaque, depressed and acromic in its center, measuring five by ten centimeters. The patient was previously treated with multiple topical therapies without clinical improvement. **Treatment.** Skin biopsy showed minimal changes in the epidermis, homogenization, and thinning of the collagen without adnexal involvement. After a correlation was made with the clinical findings, starting treatment with high-potency intralesional steroids (triamcinolone acetonide) was recommended. **Outcome.** After administering two applications of the drug, four weeks apart, the complete resolution of the dermatosis was evidenced. One month after the last dose, the patient showed no recurrence.
Keywords
Atrophy, localized scleroderma, steroids, atrophoderma.

Resumen
La atrofodermia idiopática de Pasini y Pierini es una entidad poco frecuente y de etiología aún no esclarecida, se presenta con una frecuencia hasta seis veces mayor en mujeres que en hombres y una posible asociación con la esclerodermia localizada (morfea). **Presentación del caso.** Paciente femenina de 30 años, quien consultó por una lesión asintomática de dos años de evolución en el glúteo izquierdo. En el examen físico se evidenció una placa ovalada, deprimida y acrómica en su centro, que mide cinco por diez centímetros. La paciente había sido tratada previamente con múltiples terapias tópicas sin obtener mejoría clínica. **Intervención terapéutica.** Se realizó la biopsia de piel que demostraba cambios mínimos en epidermis, homogenización y adelgazamiento de colágeno sin afección de anexos. Se hizo correlación con los hallazgos clínicos y se decidió iniciar tratamiento con esteroides intralesionales de alta potencia (acetónido de triamcinolona). **Evolución clínica.** Posterior a la administración de dos aplicaciones del medicamento, con cuatro semanas de diferencia entre ellas, se evidenció la resolución completa de la dermatosis. Un mes después de la última dosis la paciente no mostró recidivas.

Palabras clave
Atrofia, esclerodermia localizada, esteroides, atrofodermia.

Introduction

Idiopathic atrophoderma of Pasini and Pierini (IAPP) is an infrequent dermatosis that affects the organization of dermal collagen fibers. It manifests clinically as areas of atrophy. Nontheless, these features are not evident in all cases[1]. Although some authors consider it a localized variant of morphea, others consider it a tatally distinct nosological entity[2]. IAPP is a single or multiple well-delimited, hyperpigmented, non-indurated patches with a slight depression of the skin, which may converge and form a confluent area with subsequent atrophy[3]. It was first described in 1923 by Pasini, who assigned it the name "progressive idiopathic atrophoderma." Thirteen years later, Luis Pierini studied this dermatosis and established a possible relationship with localized scleroderma. Subsequently, Canizares assigned it the term idiopathic atrophoderma of Pasini and Pierini[1]. Its etiology still remains uncertain.

Atrofodermia idiopática de Pasini y Pierini, un abordaje inusual

Suggested citation:
Flores Hernández C, Moreno Ruiz J, Guevara Cerritos A, Maza A. Idiopathic atrophoderma of Pasini and Pierini, an unusual approach. Alerta. 2023;6(2):99-104. DOI: 10.5377/alerta.v6i2.15578

Received:
January 16, 2023.

Accepted:
May 29, 2023.

Published:
July 20, 2023.

Authors contribution:
CFH[1]: study conception, manuscript design, literature search, data collection, management, and analysis; writing, revising and editing. JMR[2]: literature search, data collection, management, and analysis; writing, revising and editing. AGC[3], AM[4]: writing, revising and editing.

Conflict of interests:
The authors declare there are no conflict of interests.

AIPP usually occurs more frequently in women than men, with a 6:1[4] ratio, mainly in young people between the second and third decade of life, albeit there are reports of congenital cases. There appears to be no predominance in any specific geographic region or ethnicity[4-5]. The lesions remain without significant changes and evolve asymptomatically during the course of the disease, increasing in size and acquiring their definitive characteristics. Once established, they do not regress spontaneously.

At the present time, there are few cases of successful therapeutic approaches reported in the literature, and their treatment is a challenge for dermatological practice, which is why those therapeutic approaches that appear to be promising are of great interest[1-4].

Case presentation

This case is about a 30 year old female patient who consulted at the Specialized Health Unit of San Jacinto, San Salvador, El Salvador, with a history of presenting an atrophic, acromic, and asymptomatic lesion for over two years. Physical examination revealed a dermatosis localized on the upper and lower outer quadrants in the left gluteus region. The dermatosis consisted of a smooth surface, depressed oval-shaped, atrophic plaque with defined cliff borders measuring 5 × 10 centimeters. Inside it, there was an oval acromic macule, with laking clear limits, measuring 3.8 x 7.5 centimeters, with a soft consistency, chronic evolution, and asymptomatic (Figure 1).

The patient had no relevant personal or family medical history. Multiple previous treatments administered included topical steroids, emollient creams, and systemic antibiotics, with no evidence of improvement. Skin biopsy reported mild perivascular and interstitial lymphocytic inflammatory infiltrate in the dermis. In the middle dermis, there was minimal homogenization of the collagen with no involvement of the adnexa skin. There were no significant changes in the epidermis (Figure 2).

Therapeutic intervention

Due to the previous topical therapeutic failure, the decision favored the use of the intralesional route with triamcinolone acetonide (50 mg vial in 5 mL) in two sessions with a difference of four weeks between applications. In each session, two milliliters of triamcinolone and one milliliter of lidocaine 2 % were applied to improve pain tolerance during the application.

Outcome

In the second control (four weeks after applying the first dose), the evolution of the lesion was assessed, noticing a marked clinical improvement. At this moment, the second dose of steroids was administered (Figure 3).

The patient was called for further follow-up four weeks after the second dose, and the complete resolution of the dermatosis was evidenced two months after starting the treatment (Figure 4).

One month after the end of the treatment, the satisfactory therapeutic results remained stable (absence of atrophy), with a slight residual hypercromia in the treated area.

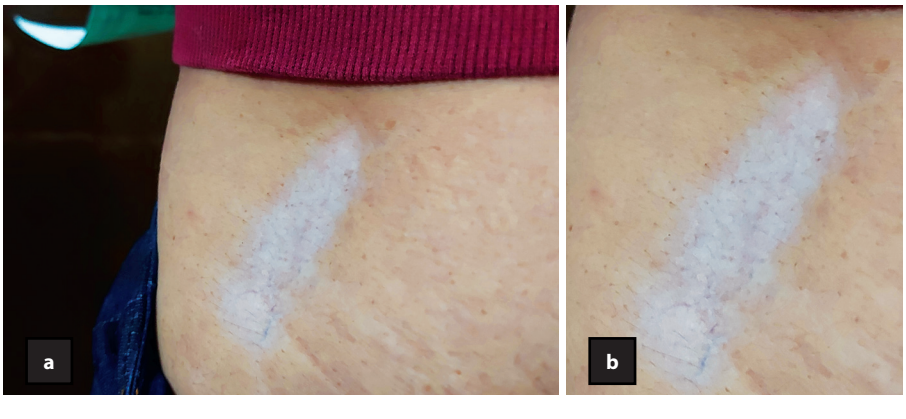

Figura 1. Lesión atrófica, ovalada, de bordes netos en acantilado. a. Presencia de área de piel acrómica en su interior, blanda a la palpación y asintomática. b. Mayor aumento

Clinical diagnosis

The histological findings on skin biopsy were non-specific and did not constitute a definitive diagnostic criterion by themselves. Therefore, they were correlated with the characteristics and time of evolution of the dermatosis in the clinical context of the patient, which led to the diagnosis of an idiopathic atrophoderma of Pasini and Pierini.

Discussion

Nowadays, IAPP remains a challenge in terms of its etiology and appropriate clinical and histological diagnostic criteria[4-6]. Since its original description by Pasini in 1923, different etiologies have emerged, even though none has enough weight to be accepted as a definitive explanation for this dermatosis[4-7].

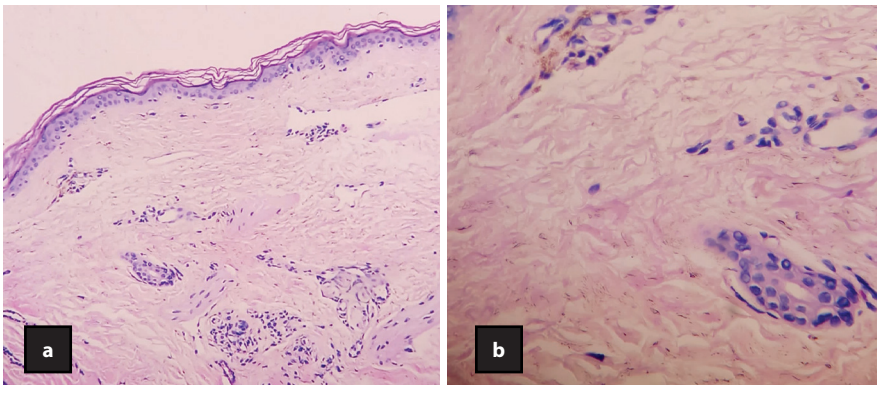

Figura 2. Hematoxilina y eosina. Epidermis sin cambios significativos. A nivel de la dermis superficial se observa leve infiltrado inflamatorio linfocitario perivascular. a. En dermis media se observa homogenización mínima de la colágena. No hay compromiso de los anexos (10x). b. Mayor aumento (40x). Fibras de colágeno homogenizado y adelgazado

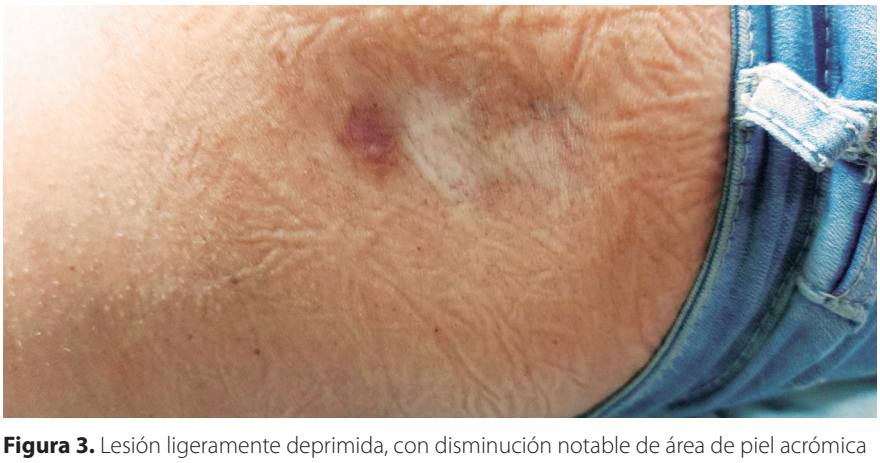

Figura 3. Lesión ligeramente deprimida, con disminución notable de área de piel acrómica

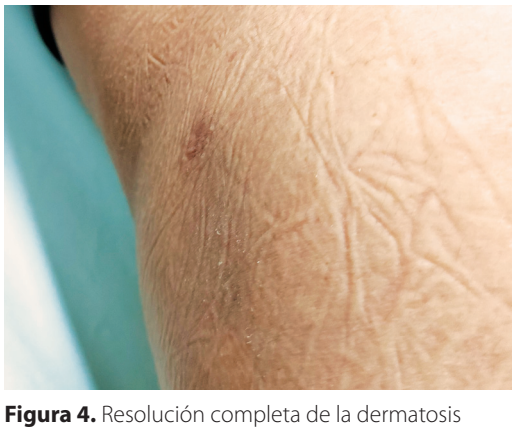

Figura 4. Resolución completa de la dermatosis

The neurogenic or zosteriform theory, which suggests some factors linked to a predisposition to errors in the migration of nerve cells from the neural crest during the embryonic period, might cause the appearance of atrophic lesions with unilateral distribution and zosteriform involvement based on a series of cases reported over the years[8-9]. On the contrary, the genetic theory supposes the existence of elements related to genetics that could allow hereditary transmission of the dermatosis. Weiner and Gant report the occurrence of IAPP in two siblings[10]. Kim Sung Kwon describes the case of a two years old girl whose dermatosis was present at birth; however, a similar clinical manifestation is not reported in another family member[11].

On the other hand, the infectious theory suggests that the association with a simultaneous infection by *Borrelia burgdorferi*. Different authors have described the presence of abnormally high antibody titles IgM and IgG against *B. burgdorferi*. Regardless of this, these patients did not meet the criteria for Lyme disease. A (discrete) clinical improvement is described in these patients when they were treated with antibiotics derived from penicillin, both orally and parenterally[9].

Although for many authors, IAPP is an "abortive" form (incomplete presentation, because it does not meet all the clinical and histologic criteria) of localized morphea, it is more appropriate to establish a classification that somehow facilitates the approach to this dilemma. The Argentine school (Pierini and Borda) proposes a classification into two variants of dermatosis: "true" IAPP, which remains unchanged in clinical and histological findings over the years; the other, a variant that seems to be clinically and histologically related to morphea. Of the latter, different forms of presentation are known: (1) IAPP with clinical and histological features that are similar to morphea ("abortive" variant); (2) IAPP presenting simultaneously with morphea lesions; (3) IAPP with lesions that over the years acquire typical morphea features; (4) IAPP progressing to systemic sclerosis[12-13].

The authors of this study consider that the patient falls into the first of the variants ("true"). The latter is based on the clinical evolution and histopathologic findings, which despite the length of the development remained unchanged in skin texture, a crucial element in the diagnosis of localized scleroderma. Also, knowing that the most relevant changes in the patient's dermatosis were the progressive size of the lesion and the accentuation of the accompanying dyschromia, the diagnosis can be oriented towards a "true" IAPP.

The lesions evolve asymptomatically during the progression of the disease, increasing in size and acquiring their characteristics. Once established, they do not reverse spontaneously. The most frequently involved anatomical sites are the trunk, back, thorax, abdomen, and arms, in order from highest to lowest. The face, hands, and feet are usually not compromised. Usually, the lesions are bilateral and symmetrical, although cases of zosteriform distribution have been described.

The dermatosis consists of depressed, hyperchromic, or skin-colored plaques. However, in this case, acromic areas may develop within the lesions. The borders are usually well delimited, giving it a "cliff" or "inverted plate" appearance. They can range in size, from two to five centimeters, but in rare cases, they can confluence and grow to a larger size, acquiring a crateriform appearance. In IAPP, the skin at the periphery of the lesions is normal, and there is no hyperchromic halo, which contributes to its distinction from morphea. On palpation, a slightly indurated consistency and a sclerotic appearance have been reported, but there may be no change in this appearance[1,4,9,14].

The primary differential diagnosis with which IAPP shares certain clinical and histologic features is localized morphea. However, there is no consensus on whether IAPP is a variant of morphea or a distinct entity[1,4,7,15]. Despite this, and based on various studies, the authors believe IAPP should be considered an independent entity from localized scleroderma[1,13,16]. In the case of the patient described above, and taking into consideration the clinical and histological characteristics, the diagnosis of IAPP seems appropriate since it meets many of the criteria, among which are the following: early age of onset, female sex, chronic evolution, clinical and histological aspects of the lesion compatible with IAPP, and no tendency to spontaneous resolution6. Other differential diagnoses may include primary anetoderma and linear atrophoderma of Moulin, each with differential histologic patterns[1,4,17].

Skin biopsy findings are usually minimal and non-specific and consist of a normally or slightly atrophic epidermis, with possible hyperpigmentation of the basal layer. A minimal perivascular and interstitial inflammatory infiltrate composed of lymphocytes, histiocytes, and plasma cells may be present in the papillary and mid-dermis. The collagen fibers present minimal changes consisting of thickening and homogenization at the level of the reticular dermis and

edema in the interior of these fibers in their upper segments[1,6,12,18], stressing that these findings are non-specific and are not a definitive diagnostic criterion by themselves. However, they can help when establishing a differential diagnosis with morphea. In the patient's case, many of the elements mentioned above were present, favoring the diagnosis of IAPP[4-6].

So far, this entity remains a challenge to treatment[4-6]. There are reports of variable data regarding the use of systemic antibiotics under the assumption that B. Burgdorferi is involved in the pathogenesis in patients with positive IgG antibodies. Oral penicillin and doxycycline 200 mg/day orally for two to three weeks have been used at variable doses with favorable results[5,9,14]. The use of antimalarial drugs is effective in a few cases[4,14,19]. Topical steroids do not seem to offer any improvement in these patientss[19].

The administration of intralesional corticosteroids is an approach that until now has not been used in this dermatosis, although topical and systemic steroids tend to be a relatively frequent approach. In several studies, this is now considered a first-line approach[4,6,19], due to its immunosuppressive and anti-inflammatory mechanism. Notwithstanding, the results were excellent in the patient when used intralesionally, caution should be applied, and further research must be conducted[4-6].

In hyperpigmented lesions, the use of a Q-switched laser has been suggested to potentially improve the condition[13].

Ethical aspects

For the presentation of this case, the patient's consent was requested and confidentiallity was respected; her approval was obtained through informed consent and it was developed in accordance with the principles of the Declaration of Helsinki.

Funding

No external funds were received for this work.

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