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## Idiopathic atrophoderma of pasini and pierini: A new case

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### Abstract

**Introduction:** The idiopathic atrophoderma of Pasini and Pierini is a rare dermatosis described for the first time by Pasini in 1923 and Pierini in 1936. Its etiopathogenesis remains controversial. We report a new case.

**Case report:** A 32-year-old woman with no history of any particular medical condition who consults for non-pruritic brownish and atrophic skin lesions without associated inflammation and sclerosis of the trunk and limbs. Histological examination of a cutaneous fragment confirmed the diagnosis of atrophoderma by Pasini and Pierini. The patient was on topical corticosteroids combined with hydroxychloroquine 400 mg daily. The evolution was marked by a stabilization of lesions with a decline of one year.

**Conclusion:** this is a rare entity, easy diagnosis and variable therapeutic options that remain disappointing, however, because of the absence of involution of the lesions.

**Keywords:** atrophoderma of Pasini and Pierini; morphea

### Introduction

The idiopathic atrophoderma of Pasini and Pierini is a rare dermatosis described for the first time by Pasini in 1923 and Pierini in 1936. Its etiopathogenesis remains controversial. We report a new case.

### Observation

A 32-year-old woman with no particular pathological history who consults for plaque-like skin lesions beginning with infra-centimetric achromatic macules (Figure 1) that initially appeared in the breasts, back and arms that have been evolving for a year. Clinical examination found achromal macules in the arms (Figure 1) and multiple, brown, non-pruritic, atrophic plaques with no associated inflammation or sclerosis of the breast and abdomen (Figures 2 and 3). and members; with a hyperpigmented closet slightly indurated at the level of the back (Figure 4). The histological examination of a cutaneous fragment had shown a normal epidermis and a fibrous dermis, seat of a discrete lymphocytic inflammatory infiltrate. The collagenous bundles are thickened with horizontalization. The diagnosis of atrophoderma Pasini and Pierini was retained. The patient was placed on topical corticosteroids for one daily application with progressive outgrowth, associated with hydroxychloroquine at a rate of 400 mg per day. The evolution was marked by stabilization of the lesions, there is no sign of progression of the cutaneous lesions nor systematization. The decline is one year.



**Fig 1:** Infra-centimetric achromatic macules of the right arm

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**Fig 2:** Brownish and atrophic plaques of the breasts.



**Fig 3:** Brownish and atrophic patches of the right flank



**Fig 4:** Hyperpigmented cupboard slightly indurated at the level of the back

### Comments

Idiopathic atrophoderma of Pasini and Pierini is a rare entity with less than 100 cases described in the literature. It affects young women. <sup>[1]</sup> Its cause remains unknown. Some authors have associated it with a *Borrelia burgdorferi* infection <sup>[1]</sup> Some cases of familial atrophoderma Pasini and Pierini have been described. However, no genetic link has been confirmed <sup>[1]</sup>. A neurogenic cause has been suggested because of the apparent zosteriform distribution of lesions in some cases <sup>[2]</sup>. The atrophoderma of Pasini and Pierini can be considered as a manifestation of localized scleroderma <sup>[3,4]</sup>.

Clinically, the disease is asymptomatic and evolves insidiously. It is characterized by single or multiple macules <sup>[3]</sup> roughly rounded and with irregular margins whose size varies from a few millimeters to several centimeters, they are atrophic and the erythematous border seen in the morphea lacks. The lesions are usually hyperpigmented but Saleh *et al.* showed in a retrospective study of 16 Lebanese patients that the lesions could be hypopigmented (56%) as in our patient <sup>[5]</sup>. The lesions do not show sclerosis or induration on palpation.

Histologically, mostly no alteration can be observed in the epidermis, but it may present a slight atrophy; the dermis may show a decrease in its thickness with changes in collagen and elastic fibers <sup>[5]</sup>.

The therapeutic approach includes tetracyclines, effective in patients with antibodies positive for *B. burgdorferi*, topical corticosteroids and antimalarials, including hydroxychloroquine, which appears to provide a good response in cases associated with lupus. In addition, topical treatments using calcineurin inhibitors have also been reported, although the responses have been variable <sup>[7]</sup>. An Alexandrite Q-switch laser treatment has also been reported to result in clinical improvement of the lesions hyperpigmented <sup>[8]</sup>.

Evolution is usually stationary. No definitive progression to systemic scleroderma was observed until Bisaccia *et al.* in 1982, reported one case <sup>[9]</sup>.

**Conflict of interest:** All authors have no conflict of interest

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