Infantile pityriasis alba and comorbid disorders

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Pityriasis alba is a common cutaneous disorder characterized by asymptomatic hypopigmented patches on the face, neck, trunk and proximal extremities of children and young adults. Its pathogenesis has not been definitely clarified. Pityriasis alba is considered to be a low-grade eczematous dermatitis and is regarded as a minor feature of atopic dermatitis. Possible comorbid disorders described in association with pityriasis alba are nutritional deficiency, anemia and parasitic infestations; however, these are exclusively observed in children of poor socioeconomic conditions. In the past, a pathogenic implication of several microorganisms, in particular Gram-positive bacteria, has repeatedly been proposed but never confirmed. Pityriasis alba has no reliable treatment, but it usually resolves with time. Emollients, mild topical steroids and pimecrolimus may aid to accelerate the repigmentation.

Pediatric HEALTH

Pityriasis alba (PA) is a common cutaneous disorder characterized by asymptomatic hypopigmented patches on the face, neck, trunk and proximal extremities of children and young adults. The single lesion has sharply demarcated margins and is covered by a fine branny scale (Figure 1). The cause of PA is still unknown, but the condition is widely considered as a mild form of atopic dermatitis. It often appears after sun exposures and result as a lack of pigmentation probably owing to a difficulty in pigmentation in the scaly hyperkeratotic patches. Lack of sunscreen use and frequent baths are also frequently reported.

Epidemiology

Pityriasis alba is found all over the world. It is quite common, affecting between 1.9 and 5.25% of preadolescent children. The peak of incidence is between the ages of 6 and 12 years [1]. It is more often observed in atopic subjects (77.9%) than in the general population [2]. Both sexes are equally susceptible. PA has a high prevalence in infants and children of low socioeconomics conditions in developing countries, being present in 12–90% of children [3–5].

Etiology & pathogenesis

Streptococcus β *haemoliticus* and *Staphylococcus aureus* infections, nutritional deficiencies and parasitic infections have been called out to explain the etiology of PA, but no definitive associations have been demonstrated, even if these disorders appear to be more frequent in patients with PA. Iron and copper deficiency have been described in patients with PA. In particular, copper is a cofactor for tyrosinase, an enzyme required for the production of melanin; hence, copper deficiency may in effect play a role in the pathogenesis of PA [1,6,7].

Since 1956, when O'Farrell suggested that PA could be considered a form of eczematous dermatitis [8], it has been widely considered as a mild form of atopic dermatitis [9]. With regard to this, it is not surprising that other possible favoring factors of PA may be sun exposure without sunscreens, frequent and hot baths, wind, soaps and cutaneous xerosis [7,9].

Clinical features

The individual lesion of PA is characterized by a rounded, oval or irregular plaque of a pale pink color. Furfuraceous or branny scaling covers the patch. One or several patches could be found with a diameter varying from 0.5 to 5 cm. Most lesions appear on the face, but upper extremities and the trunk may also be involved. Forehead and malar ridges are mainly involved in 63 and 57% of the cases, respectively. Angles of mouth and lateral superorbital regions are less involved, observed in 37 and 35% of patients, respectively. Only in 20% of affected children are symptoms present on the neck, shoulders or trunk as well as the face [1,10].

Once a pink patch with elevated borders has formed, PA usually fades over several weeks into a white patch covered with a powdery scale that can persist for months or years. Often erythema is so pale and of short course that it is unnoticeable. The lesions become more notable in summer with tanning of the surrounding skin. Recurrent crops of new lesions may develop at intervals.

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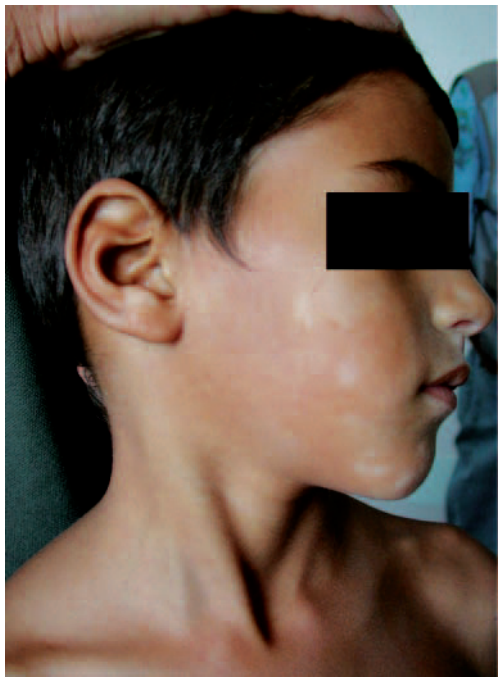

Figure 1. Clinical presentation of pityriasis alba. The clinical presentation of pityriasis alba is characterized by light-colored patches that are classically localized on the face.

Histology

Pityriasis alba is generally diagnosed on the basis of clinical features. Histological analysis is usually unnecessary and nonspecific. However, because differential diagnosis also includes disorders, such as vitiligo, hypochromic initial forms of leprosy and mycosis fungoides, histological analysis may sometimes be useful.

Microscopic examination can reveal in the epidermis hyperkeratosis, parakeratosis, acanthosis and mild spongiosis. In the hypomelanotic maculae, melanocytes and melanosome are decreased in the basal layer of the epidermis while no defect has been found in the melanosomal to keratinocytes transfer [1,6].

Additional histopathological features of early PA are follicular plugging, follicular spongiosis and atrophic sebaceous glands. Perivascular lymphocitic infiltrates and edema can be seen in the dermis. Late PA is usually characterized by the histology of a nonspecific chronic eczematous dermatitis.

Ultrastructural findings of hypopigmented maculae demonstrate reduced numbers of irregularly patterned melanocytes and melanosomes [1,6].

Atypical variants of pityriasis alba

In the past, two variants of PA have been described: pigmenting PA and extensive PA [10].

Pigmenting PA presents as a central dark hyperpigmented patch surrounded by a classical hypopigmented scaly area of PA. It appears almost always on the face and, in 30% of cases, a concurrent classic PA is associated. In 65% of cases affected, patients also present a superficial dermatophyte infection, but it is coinceivable that these cases should not be considered as pigmenting PA [1,6].

Extensive PA is mainly seen in young adults of female sex and mixed ethnic origin. It is a primary acquired hypopigmentation that affects principally the inferior portion of the trunk with a symmetrical involvement and relapsing course (Figure 2). It is usually asymptomatic, without scaling and does not appear to be related to atopic dermatitis. Recently, it has been suggested that extensive PA should not to be considered a clinical variant of PA, but rather as a distinct disorder [10]. Thus, Di Lernia and Ricci argued that extensive PA is a misnomer that could produce confusion and proposed 'progressive and extensive hypomelanosis' as the denomination for this disorder [11]. The controversy is not a mere question of semantics and nomenclature because classical PA could also present with widespread lesions, better classified as 'generalized PA' [12,13].

Differential diagnosis

Pityriasis alba must be differentiated from other disorders that often heal leaving a postinflammatory depigmentation, such as psoriasis or nummular eczema; thus, the anamnesis is mandatory.

Vitiligo must be distinguished by the presence of chronic, achromic (not hypochronic) patches not preceded by desquamation and characterized by sharp margins [9]. Wood's lamp examination can also help to differentiate achromic lesions of vitiligo from the hypochromic lesions of PA.

The small size and the particular shape (with geographic outlines) of coalescing macule lesions on the trunk differentiates pityriasis versicolor (tinea versicolor), although in childhood this condition can occur on the face and forehead as well (Figure 3). In these cases, microscopic examination for fungi can be very useful.

Nevus anemicus and nevus depigmentosus are usually characterized by a single lesion, occurring in the first infancy or at birth, without clinical evolution during time [1].

Mycosis fungoides may rarely present exclusively with hypopigmented scaly patches. It usually affects adults, but cases in children have been reported as well. Thus, the differential diagnosis of this cutaneous T-cell lymphoma must be kept

in mind and histological examination must be performed in long-lasting cases of hypomelanotic patches [14].

Pityriasis rotunda type II, probably a genodermatosis with temporary phenotypic expression, presents with circular, sharply delineated, sometimes confluent patches, giving rise to polycyclic figures. The surface shows a slight desquamation with adherent scales. The lesions are lighter in color than the surrounding skin and are distributed on the abdomen, upper and lower extremities, back and chest. Familial history, distribution and histopathologic findings are useful for the differential diagnosis with PA [15].

The identification of one or several hypochromic patches on the skin is a key stage in the diagnosis of leprosy on dark skin in patients living in (or immigants from) endemic areas [1]. Cutaneous lesions in leprosy-diagnosed patients are characterized by alterations in thermosensation, expressed as warm hypoesthesia, cold hypoesthesia or both. Skin biopsy is recommended in doubtful cases.

Comorbid disorders

Atopic dermatitis is probably more a comorbidity of PA than a etiopathogenetic factor. Indeed, PA is considered a low-grade eczematous dermatitis and is regarded as a minor feature of atopic dermatitis and a personal or family history of atopy has been found in 85.5% of patients affected by PA [3]. Brenninkmeijer *et al.* introduced the term of 'atopiform dermatitis' (also termed intrinsic atopic dermatitis) to distinguish the subgroup of patients with atopic dermatitis and absent allergen-specific IgE. The authors demonstrated a relevant negative association of PA with atopiform dermatitis (10.0% of cases), with respect to the strong association with atopic dermatitis (77.9% of cases) and speculated that atopiform dermatitis is an entity distinct from atopic dermatitis considering that, in addition to the absence of allergen-specific IgE, a history of atopy, recurrent conjunctivitis, palmar hyperlinearity, keratosis pilaris, PA and hand and/or food eczema is significantly less frequently present in atopiform dermatitis [2].

Skin dryness observed in many patients with PA is generally due to the inappropriate use of soaps, abrasives and frequent long and hot baths.

Additional comorbid disorders described in association with PA are nutritional deficiency, anemia and parasitic infestation. All these conditions are probably inscribed into a poor socioeconomic background, which has been widely reported in relationship to PA. Indeed, Ruiz-Maldonado proposes to differentiate PA into two types:

- Endemic PA, affecting over 90% of the child of poor socioeconomic condition;
- Atopic dermatitis-related PA [1,5,6].

Infections from *Streptococcus pyogenes*, *S. aureus* and other microorganisms have been proposed as comorbid disorders, but no clinical study has confirmed these infection as causal factors of PA [1,7].

Treatment

Before treating a patient with PA, it is important to explain that this disorder is benign and usually resolves spontaneously with growth. Emollients and mild topical steroids may be useful in reducing the subclinical inflammation and in accelerating repigmentation that could otherwise last for months or years. Recently, the use of the cream pimecrolimus 1% has also been suggested to avoid the long-term use of steroids and has proven to be efficacious [16,17]. The patient must also be informed to reduce sun exposure and aggressive detergents.

Conclusion

Pityriasis alba is a common, mainly localized, hypopigmentation that is observed in children and young adults, and has no reliable treatment but it usually resolves with time. It is more evident in subjects of high phototype

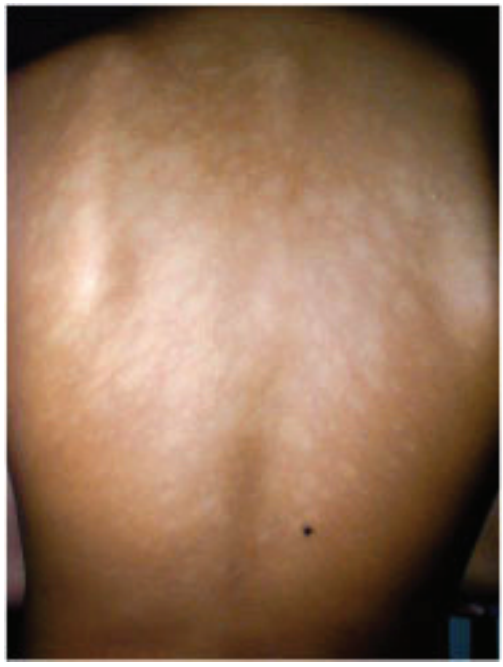

Figure 2. Pityriasis alba extensa. Pityriasis alba extensa, better named 'progressive macular hypomelanosis', occurs mainly on the trunk of young females. It is often mistaken for pityriasis alba.

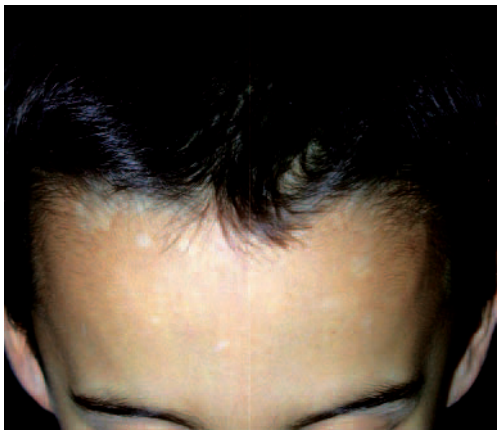

Figure 3. Pityriasis versicolor of childhood. Facial lesions, preferentially on the forehead, are frequently observed in pityriasis versicolor of childhood.

but it occurs in all ethnic groups. Etiology is still unknown even if association with atopy and low-grade socioeconomic conditions are probably the most relevant favoring factors. Emollients, mild topical steroids and pimecrolimus may aid to accelerate the repigmentation. Pigmenting and generalized PA are two true variants of PA, while the so-called extensive PA appears to be a different disease that should be better termed as progressive and extensive hypomelanosis.

Future perspective

Although PA is a well-known skin disorder for many years, its pathogenesis has not been definitely clarified. The role of atopy is still not well understood and, recently, a major relationship with extrinsic atopic dermatitis has been proven. The responsibility of poor socioeconomic conditions has to be elucidated. In this context, a relationship with a possible pathogenic role of iron and copper deficiency should be evaluated. Finally, a pathogenic implication of several microorganisms, in particular Gram-positive bacteria, have repeatedly been proposed and never confirmed.

Surely a better knowledge of the understanding pathogenic factors of PA could lead to correcting the possible nutritional defects in malnourished children or in populations with difficult access to medical care and, in general, to a more specific therapy.

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Executive summary

- Pityriasis alba (PA) is a common cutaneous disorder characterized by asymptomatic hypopigmented patches on the face, neck, trunk and proximal extremities of children and young adults.
- It is more often observed to be present in atopic subjects (32%) than in the general population (1%).
- Etiology is still unknown, but it is widely considered a form of eczematous dermatitis – it is included among the minor diagnosis criteria of atopic dermatitis.
- The individual lesion of PA is characterized by a rounded, oval or irregular plaque of a pale pink color covered by furfuraceous or branny scales, which progress in a long-standing hypochromic macule. The initial inflammatory phase can be unremarkable.
- PA is generally diagnosed on the basis of clinical features; histological analysis is usually unnecessary and nonspecific.
- Pigmenting and generalized PA are two clinical variants of classical PA.
- The so-called extensive PA is a different disease that should be better named as 'progressive and extensive hypomelanosis'.
- Topical emollients, mild topical steroids and pimecrolimus 1% can be used to control the disorder, even though complete resolution does not always take place. A definitive treatment is not available, but the condition usually disappears slowly. Relapses in the following years are common.

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