Ashy dermatosis in pediatric age. Report of a new case: clinical, histological and dermoscopic features

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Summary

Ashy Dermatosis (AD) is a rare acquired dermatosis characterized by bluish-grey coloured macules, of variable shape and size, more frequently involving the trunk and the arms. AD generally affects dark phototypes, more frequent in Central and South America. It usually appears in young adults, more often in women, but few cases have also been reported in childhood. Over the years there has been much confusion about terminology: in fact, many terms have been used to describe similar clinical features such as erythema dyschromicum perstans, idiopathic eruptive macular pigmentation and lichen planus pigmentosus. Usually the diagnosis of AD is performed on the basis of the clinical rash, even if sometimes the characteristic features could be absent in consideration of the stage of the disease. Then in the hypothesis of AD usually also a skin biopsy is necessary to assess the final diagnosis.

We report the use of a new tool, recently described, that in our opinion could be helpful in the diagnosis of AD. Dermoscopy is a non invasive exam that can be performed easily also in children. We suppose that the recognition of detailed dermoscopic features could represent a valid diagnostic aid and could allow to avoid a skin biopsy.

KEY WORDS: ashy dermatosis; erythema dyschromicum perstans; pediatric dermatology.

Introduction

Ashy Dermatosis (AD) was first described in El Salvador 1957 by Ramirez as a macular hyperpigmentated rash of a characteristic bluish-grey colour which he called “los cenicientos” (1).

AD is considered a rare dermatosis. It usually appears in young adults, more often in women, but few cases have also been reported in childhood, in dark phototypes. AD in adults is more frequent in Central and South America, while children are usually Caucasian.

Recently Vazquez-Lopez et al. have described the dermoscopic subpatterns of AD (2).

Case report

We report the case of an Italian 6-year-old girl, with Fitzpatrick type III skin.

Clinical observation revealed the presence of asymptomatic lesions distributed symmetrically on her neck, back, chest and proximal arms (Figure 1 a, b). The macules were of a particular blue-greyish colour, of variable diameter from few millimeters to 1 centimeter. The rash appeared about 3 months before the observation and at the beginning there were only few little macules on the neck, then gradually increasing involving also back, chest and arms in about one month. Her parents had not given so much importance to these macules as they thought they were ink spots, even if her parents tried to remove the spots with water and soap. They had not noticed any previous erythema or dermatitis in the involved area. The girl was healthy and was not taking any medication or dietary supplement, and had not been in contact with new substances. No similar cases or other dermatological disorders were reported in the patient’s family.

Laboratory tests showed normal blood cell count, kidney and liver function, erythrocyte sedimentation rate, C-reactive protein, and anti-streptolysin title. The assay for autoimmunity (anti-nuclear, anti-ENA, anti-DNA ds), Immunoglobulins E, endocrine exams and serology for viral infections (HIV, B and C hepatitis included) were all negative.

A skin biopsy was performed in a blue-greyish macule on the neck area. Histology showed lymphocitic perivascular inflammatory infiltrate and pigment incontinence (Figure 2).

On the basis of clinical appearance and histological features, a diagnosis of Ashy dermatitis was carried out.

To better define the clinical features, we also per-
formed dermoscopy on these hyperpigmented macules. It showed grey-brownish dots and globules symmetrically distributed over the entire lesion, sometimes with linear distribution. The limits of the single macules were given exclusively by the presence of such dots-globules and a greyish background, not always recognizable. In some cases, moreover, the dots-globules, despite of small size, appeared of variable, bizarre, sometimes elongated shape. In this dermoscopic appearance, constant and symmetrical, we did not observe other dermoscopic structures and no sign of “dynamism” of the lesions (for ex. striae, signs of melanocytic activation, aspects of regression, colour heterogeneity) (Figure 3 a, b).
Due to our patient’s young age and in consideration of the benign course of the skin disease, sometimes even autoresolutive, no treatment was prescribed. We reported this case for the rare incidence of AD in pediatric age and to confirm the dermoscopic pattern of AD as recently described.

Discussion
AD is a rare acquired chronic hyperpigmentation disorder of unknown origin. In literature, the pediatric cases reported have been described more frequently among Caucasian population, while adult ones in Hispanic people. AD is characterized by eruptive asymptomatic rash of ash-coloured macules, sometimes surrounded by an erythematous border, of different size from millimeter to centimeter, that involves the trunk and the proximal extremities (3-5).

As a matter of fact, since the first description of AD by Ramirez in 1957, there has been a debate about the existence of AD as an individual entity. Even more, in the past a lot of confusion was due to several terms, such as erythema dyschromicum perstans (EDP), lichen planus pigmentosus (LPP) and idiopathic eruptive macular pigmentation (IEMP), used to describe similar clinical features. Actually, the difference between these diseases was based on clinical and histological features.

The term of erythema dyschromicum perstans (EDP) was introduced by Sulzberger to describe the clinical features of the lesions: the red border of the macules and the persistence of the dyschromia (6). Although these characteristics can be found, the red border is not a constant feature, it is often hard to find and it is related to the disease activity. Despite this there isn’t an univocal consensus, today most of the authors agree on considering EDP and AD synonymous and these terms are employed indifferently (7, 8).

Some authors consider AD as a variant of lichen planus, while other as a distinct entity. In both these cutaneous disorders histologically we can found melanophages and vacuolization of the basal layer, while Mall-Joseph spaces usually are not observed in AD. LP is very unusual in pediatric age (9).

AD and LP have also been described occurring in the same patient. This coincidental eruption has also been called lichen planus pigmentosus (LPP), a macular variant of LP (10, 11).

Idiopathic eruptive macular pigmentation (IEMP) is a disorder of pigmentation first described in 1978 as a new clinical entity (12). It has been proposed diagnostic criteria to differentiate IEMP from AD. At time of writing, these two entities are now considered as the same disorder (13).

Many clinical entities have to be considered for the differential diagnosis of AD and among these we have to include: post-inflammatory pigmentation, fixed drug eruption, Addison’s disease, hemochromatosis, pityriasis versicolor, acanthosis nigricans, pseudoacanthosis nigricans, terra-firma forme dermatosis, idiopathic eruptive macular pigmentation, phytophotodermatoses, cutaneous amyloidosis (14-16).

The cause of acquired skin hyperpigmentation may vary. It can be a sign of a systemic disease or can be due to environmental factors (exogenous substances, sunlight, drugs).

To discriminate among these many differential diagnosis, the clinical history and dermatological observation are of great importance. In doubtful cases, the main help can be represented by the histology, which contributes to the final diagnosis.

The pathogenesis of AD is still unknown. It has been reported in association with autoinflammatory diseases, viral and parasitic infections, allergy to cobalt and radiological contrast material. However, no casual or trigger factors have been identified in most of the reported cases, especially those in pediatric age (17, 18).

An abnormal response to antigenic stimulation and an alteration in the expression of adhesion molecules, in particular LFA-1α and ICAM-1, have been hypothesized to play a role in the development of the damage of the basal layer.

Besides, it has been suggested that an important genetic susceptibility could be conferred by genes located within the major histocompatibility complex region. In particular, the presence of HLA-DR4*0407 is considered a risk factor in Mexican mestizos (19).

Vasquez-Ochoa et al. carried on a study to characterize the histopathology of AD. A dermal lymphocytic infiltrate was observed in all cases, most frequently with a perivascular distribution. Other histological characteristic features included melanin incontinence with presence of melanophages in the dermis, vacuolization of the basal layer, colloid bodies and exocytosis of lymphocytes (20).

This is probably due to the variety of histopathological features that can be observed are related to the site of skin biopsy and also to the clinical evolution of the skin lesions.

The diagnosis of AD can be performed on clinical and histological features. Unfortunately, the characteristic clinical features is not always present in each patient, for example the erythematous border of the lesion often cannot be appreciated. Sometimes the histology of AD couldn’t be pathognomonic, since probably the histological features change in the different stages of the clinical course and according to the site where the skin biopsy has been performed.

Recently the use of dermoscopy has been described as a new tool to better define the clinical features of AD and to help making the diagnosis. Dermoscopy shows regressing grey macules with clustered granular grey-brown dots and globules. The authors hypothesized that the amount of granules could suggest the clinical course: a large amount of granules could be related to a slower course (2).

It has been also described the features of AD in electron microscopy, that showed vacuolization of basal keratinocytes, widening of intercellular spaces with the retraction of desmosomes, the presence of melanophages and dermal perivascular infiltrate (21).
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The clinical course is not the same in every patient. The improvement or the resolution of the lesions is more often observed in children than in adults, in which spontaneous remission is rare. The clinical course of AD in childhood has been reported to be resolved within 2-3 years (5). Different treatment such as sun blocking agents, topical steroids, dapsone, griseofulvin, chemical peelings, laser therapy, have been proposed from different authors but appear to be not so effective. Clofazimine has been employed in the treatment of AD in consideration of its anti-inflammatory and immunomodulatory effects, and of the involvement of cell adhesion and activation molecules in the pathogenesis (22). Actually, in consideration of the benign course of this condition and of the possible adverse events of drugs, often therapeutic efforts are avoided.

Conclusions

Recently dermoscopy and electron microscopy has been proposed to better define the features of AD and it could represent useful tool for a certain diagnosis. We have reported a case of pediatric AD and we confirmed the dermoscopic features previously reported in literature. Since AD in children is a rare dermatosis, it is difficult to obtain many data on these two techniques. However, more reports are necessary to define dermoscopic and electron microscopy features to provide with the diagnostic criteria. In particular, dermoscopy is a non invasive diagnostic procedure that may be useful in pediatric patients in whom a skin biopsy could be difficult to perform.

References