Persistent Grayish Pigmentation in a Child

E Roche, I Febrer, and V Alegre
Servicio de Dermatología, Hospital General Universitario de Valencia, Spain

Clinical History

The patient was a 7-year-old child of gypsy origin, with no relevant personal history, who presented with progressively developing, asymptomatic, hyperpigmented lesions, first observed 11 months earlier. The lesions had started to appear on the upper part of the trunk and had shown a centrifugal progression, becoming generalized over the course of a few months. No triggering event was identified. The mother had observed that, having reached their maximum development, the lesions were slowly fading.

Physical Examination

The patient presented multiple, oval macules with sizes ranging from 0.5 to 3 cm, which became confluent and covered large areas. The lesions were of a slate-blue color, and had no inflammatory signs at their margins. They were distributed predominantly over the trunk, neck, and proximal parts of the limbs, including the axillas, and their long axis was orientated along the skin tension lines (Figures 1 and 2). The scalp, mucosas, palms, soles and nails were unaffected.

Histopathology

The biopsy showed preservation of the epidermis with hyperpigmentation of the basal layer. A mild, predominantly lymphocytic, perivasculary inflammatory infiltrate was observed in the papillary dermis, with pigmentary incontinence and melanophages (Figure 3).

Complementary Tests

Blood tests were performed, including a complete blood count, basic biochemistry, coagulation studies, antistreptolysin antibodies, antinuclear antibodies (antinuclear antibody and extractable nuclear antibody), and viral serology: hepatotropic viruses (hepatitis A, B, and C [HCV] viruses), cytomegalovirus and Ebstein-Barr virus. The results were within the normal ranges.
What Was the Diagnosis?

Diagnosis

Erythema dyschromicum perstans or ashy dermatosis.

Comment

Erythema dyschromicum perstans was first described in 1957 by Ramírez as an acquired dermatosis that occurs in individuals of Hispanic descent, though it has subsequently also been reported in non-Hispanic whites, African Americans, Indians and Asians. It usually presents in young adults under 30 years of age, and is uncommon in prepubertal children.\(^1\) It has been suggested that its appearance in children is more common in non-Hispanic whites. It shows no sex predominance, though it appears to be more frequent in women.

Its etiology is unknown, although it has been associated with endocrine diseases, vitiligo,\(^1\) infections (human immunodeficiency virus [HIV], HCV),\(^2\) drug administration (antibiotics, benzodiazepines, iodine derivatives), and exposure to environmental allergens (pesticides, fungicides, ammonium nitrate, coal).

The lesions consist of oval macules and plaques, between 0.5 and 3 cm diameter, of bluish or grayish color. This latter characteristic was the reason that the patients were initially referred to as ashen. The lesions first appear on the trunk and extend centrifugally and symmetrically towards the limbs, with the long axis orientated along the skinfolds. In 2 prepubertal cases, the lesions were orientated along the Blaschko lines.\(^4\) The macules are most common on the neck, trunk and proximal parts of the arms, while the scalp, palmar-plantar surfaces, nails and mucosas are usually unaffected. Initially the lesions may be surrounded by an erythematous border that eventually disappears; however, this inflammation is often subclinical, and the lesions arise and progress with no erythema. They develop over weeks or months and are asymptomatic or present mild pruritus.\(^1,2\)

Improvement or resolution of the lesions is more common in children than in adults; improvement occurs in 50% of cases in children,\(^1,2\) whereas the disorder can persist for life in adults.

The histological findings vary according to the phase of the lesion. In the initial stages, the inflammatory border shows vacuolization of the basal layer, with occasional colloid bodies, edema of the papillary dermis, and a variable degree of lichenoid infiltrate interspersed with melanophages. The inflammatory infiltrate is less pronounced in the mid-dermis and shows a perivascular distribution. Lymphocytic exocytosis can develop in the epidermis. In the stable, hyperpigmented phase, the epidermis appears thinner, with mild follicular hyperkeratosis, mild inflammatory infiltrate, and marked pigmented incontinence with abundant melanophages.\(^2\)

Colloid bodies with immunoglobulin (Ig) M, IgA, IgG, fibrinogen or C4 have at times been detected using immunofluorescence. In other cases, deposits of granular IgM and fibrinogen have been found at the dermoepidermal junction.\(^2\)

Many treatments have been reported but with poor efficacy. Topical measures have included corticosteroids, hydroquinones, keratolytics, chemical peels, laser therapy, and phototherapy. Systemic treatments tested include corticosteroids, antihistamines, isoniazid, dapson,\(^7\) clofazimine, griseofulvin, estrogens, ascorbic acid, and chloroquine.

In children, it is recommended to inform the parents and patients about the good prognosis of the disease, to insist on the use of sunscreens to prevent the hyperpigmentation from increasing, and to await a possible spontaneous resolution.\(^2\)

The differential diagnosis should include other disorders that cause clinical hyperpigmentation, such as fixed drug eruption, lichen planus, pigmented incontinence, mastocytosis, hyperpigmentation in Addison disease, typhus, and discoid lupus erythematosus, among others.\(^8\)

Conflicts of Interest

The authors declare no conflicts of interest.

References