Idiopathic eruptive macular pigmentation

K. Trčko, P. B. Marko and J. Miljković

SUMMARY

We present the case of a 10-year-old girl with a six months history of disseminated asymptomatic, brown pigmented macules on the trunk and proximal parts of the extremities. The clinical picture, histological findings, and the course of disease were similar to those of idiopathic eruptive macular pigmentation. The cutaneous lesions gradually disappeared over the next two years without any treatment, and no relapse occurred.
The knowledge of this disease is important in order to avoid unnecessary treatment as spontaneous resolution of the lesions may be expected within months or a few years. The spontaneous regression without any treatment is an additional diagnostic criterion.

Introduction

Idiopathic eruptive macular pigmentation (IEMP) is a rare skin disorder characterized by asymptomatic, brown pigmented macules involving the neck, trunk and proximal portions of the extremities. It was first described by Degos et al in 1978 (1). The disease occurs primarily during childhood and adolescence usually without a history of erythema, drug medication or any other skin disorder. The lesions usually appear abruptly and gradually disappear spontaneously over the period of a few months to years without any treatment (2). The etiology is still unknown. Histopathologic examination shows normal epidermis and many melanophages in the upper dermis. Electronmicroscopy shows an increased number of melanosomes in basal and suprabasal keratinocytes, and clustered melanosomes in dermal melanophages.

KEY WORDS

idiopathic eruptive macular pigmentation, 10-year-old girl, spontaneous regression

Case report

A 10-year-old girl was examined because of the occurrence of brown pigmented macules on the trunk

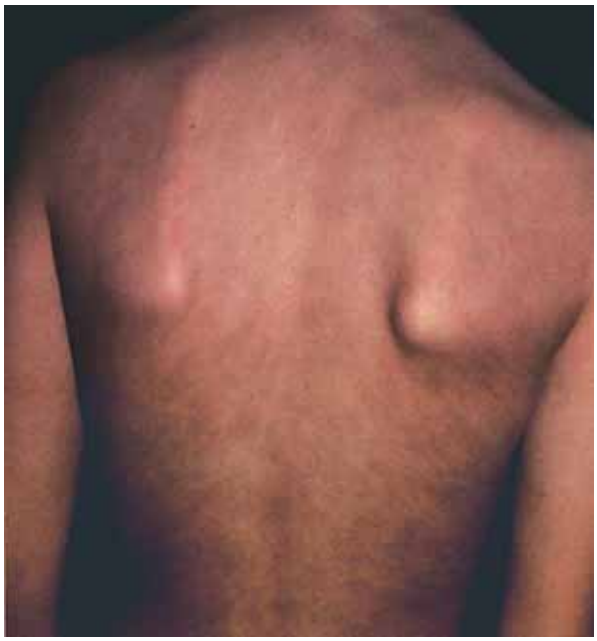

Figure 1.Idiopathic eruptive macular pigmentation: brown pigmented macules on the back.

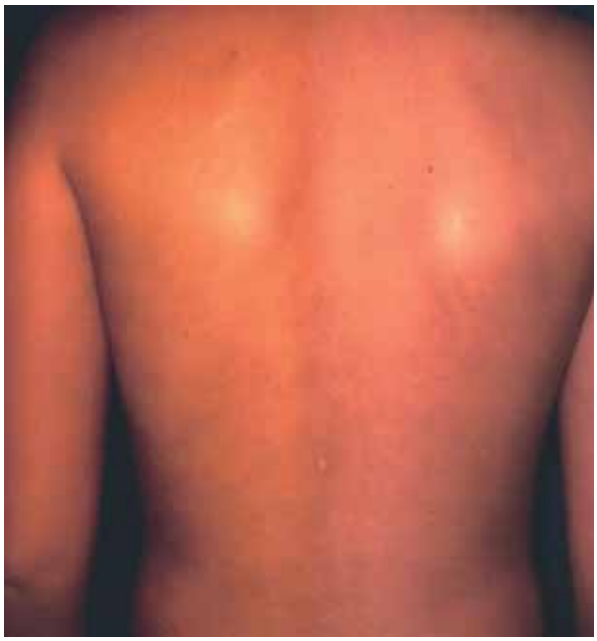

Figure 4. Idiopathic eruptive macular pigmentation: The cutaneous lesions on the back gradually disappeared after 24 months.

and proximal parts of the extremities which had appeared 6 months earlier. There was no history of preceding inflammatory erythema, insolation, pruritus, atopy, drug medication or any other skin disorder. The patient was otherwise healthy. The other members of patient's family did not have similar skin lesions.

Over the trunk and proximal parts of the extremities symmetrically disseminated, brown pigmented macules were seen (Figures 1,2). Mucous membranes

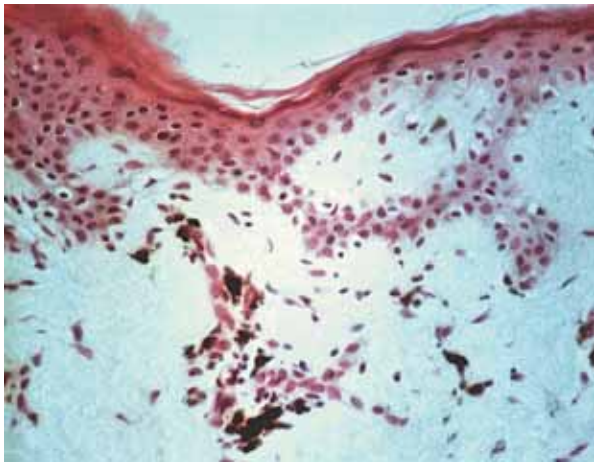

Figure 2. Idiopathic eruptive macular pigmentation: normal epidermis and great number of melanophages in the papillary dermis (hematoxylin-eosin, x200).

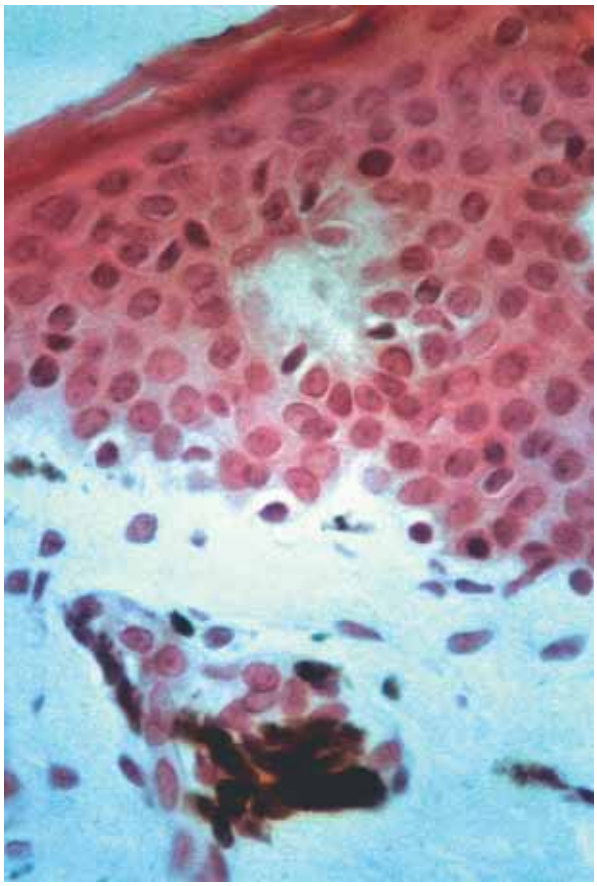

Figure 3. Same, magnification x 320.

were spared, palms and soles were clear, and the Darieržs sign was absent.

Laboratory studies including complete blood cell counts, and a panel of serum chemistry showed that serum glucose, protein electrophoresis, iron, ferritin and thyroid hormones were all within a normal range. The serum IgE level was normal. Abdomen ultrasound examination revealed no abnormalities.

Histological examination of a skin biopsy specimen revealed normal epidermis and a great number of melanophages in the papillary dermis (Figure 3). The special staining did not detect iron in the vicinity of melanophages, which is frequently observed in drug eruptions.

The cutaneous lesions gradually disappeared over a 24 month period without any treatment (Figures 4,5). No relapse occurred.

On the basis of the clinical and histological features and the course of the disease the diagnosis of idiopathic eruptive macular pigmentation was made.

Discussion

The etiology and pathogenesis of the disease are still unknown. Degos et al. in 1978 observed seven cases of a pigmented dermatosis different from ashy dermatosis and from erythema dyschromicum perstans. Some authors believe that hormonal factors may be involved in the pathogenesis of IEMP because the disease primarily occurs during childhood and adolescence (3). Because of ultramicroscopic findings other authors suspect that in some cases it represents the end stage of lichen planus (4). Dupré et al. have described two cases of acquired hyperpigmented macular eruption following pityriasis rosea (5).

De Galdeano et al. have studied five cases of IEMP (6). They concluded that the condition appears to be a distinct clinical and histopathologic entity, and summarized some basic diagnostic criteria necessary to recognize this rare disease:

1) eruption of brownish, nonconfluent, asymptomatic macules involving the trunk, neck, and proximal extremities in children or adolescents;

2) absence of a preceding inflammatory process;

3) no previous drug exposure;

4) basal cell layer hyperpigmentation of the epidermis and prominent dermal melanophages without visible basal layer damage or lichenoid inflammatory infiltrate; and

5) normal mast cell count.

The occurrence of hyperpigmentation is usually linked to the activity of melanocytes. A diffuse hyperpigmentation may also occur as a manifestation of drug eruption or in the course of systemic diseases, like hemochromatosis, hyperthyroidism or Addison disease. The differential diagnosis of IEMP includes ashy dermatosis (erythema dyscromicum perstans), fixed drug eruption, café-au-lait macules, mastocytosis and postinflammatory hyperpigmentation.

Ashy dermatosis occurs almost exclusively among Latin Americans, there is a preceding inflammatory erythema and the lesions are relatively stable. Post-inflammatory hyperpigmentation following viral exanthemas or drug eruptions are the most likely differential diagnosis. In the case we present the possibility of fixed drug eruptions was excluded .

Café-au-lait macules can be single or multiple and are seen in normal individuals or can be associated with syndromes such as neurofibromatosis, McCune-Albright syndrome, ring- chromosome syndromes, and Watson syndrome (7). These can be easily differentiated from IEMP because there is no spontaneous regression and the other signs are missing. Neurofibromatosis can be easily excluded because there is no spontaneous regression and further signs are expressed. Related to neurofibromatosis is Watson syndrome, characterized by café-au-lait macules, pulmonary stenosis and a low intelligence (7). McCune-Albright syndrome is a rare disorder characterized by precocious puberty, polyostotic fibrous dysplasia and café-au-lait spots. Additional endocrine abnormalities which may be present include hyperthyroidism, growth hormone excess and hyperprolactinemia (8). Ring chromosome syndrome 15 (r15) is characterized by specific facial features, café-au-lait (dark) spots, hyperpigmentation, failure to thrive, mental retardation and typically by a terminal deletion of the long arm of chromosome 15. The hyperpigmentation and the dark spots are present in 30% of patients and are probably due to a gene deletion on the distal 15 q arm (9).

In mastocytosis a positive Darier's sign, and a characteristic histopathologic pattern with an increased number of mast cells are easily recognizable signs of disease.

Post-inflammatory hyperpigmentation is the consequence of an inflammatory process at the epidermal-dermal junction occurring in lichen planus, lupus erythematosus, atopic dermatitis, lichen simplex chronicus, psoriasis, erythema multiforme, fixed drug eruption, photosensitivity or in other allergic processes.

Although histopathologic findings cannot offer an accurate diagnosis, the post-inflammatory hyperpigmentation in the early stage of the disease may show liquefaction degeneration of basal layer keratinocytes (10,11). In addition to histopathologic differences, clinical manifestations without any history of preceding dermatoses support the diagnosis of IEMP. In our opinion spontaneous regression of the lesions over a period of a few months or years without any treatment is an additional diagnostic criterion.

Conclusion

In our case all the diagnostic criteria were fulfilled. We believe that knowledge of this entity is important to avoid unnecessary medical treatment because of the expected spontaneous resolution within a period of a few months or years. Probably an epidemiologic approach including a meta-analysis of a larger number of cases would be helpful in establishing the pathogenesis. We believe the creation of a European registry for IEMP would be helpful.

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