Idiopathic eruptive macular pigmentation – a rare pigmentary disorder

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Abstract

Skin pigmentation disorders are frequent problems in dermatological practice. They have a heterogeneous aetiology and clinical picture, and demand a flexible therapeutic approach. We present a case of a 10-year-old boy with idiopathic eruptive macular pigmentation. It is a rare chronic entity which causes only an aesthetic defect and demands no treatment.

Key words: idiopathic eruptive macular pigmentation, ashy dermatosis, macules.

Introduction

Idiopathic eruptive macular pigmentation (IEMP) belongs to the group of skin pigmentation disorders. It is a very rare entity, with the presence of characteristic reddish-brown or brown, sharply separated macules of diameter 0.5–2.5 cm. Lesions usually occur on the neck, trunk and extremities and are not accompanied by any subjective symptoms. The disease usually appears during childhood or puberty, with similar frequency in males and females. In the histopathological picture, increased pigmentation of basal layer cells and the presence of numerous melanophages are observed. The disease has a chronic course with spontaneous outcome and does not require any treatment.

Case report

A boy, aged 13, was admitted to the Outpatient Ward of the Dermatology Department of the Medical University of Gdańsk with skin lesions characterized by numerous, greyish-brown, oval macules of diameter 0.5–1.5 cm, located symmetrically on the trunk and neck (Fig. 1). Lesions were sharply separated from uninvolved skin, without a tendency to merge. Skin lesions did not cause any complaints. Darier's sign was negative. Macular lesions appeared for the first time around 1.5 years before admission to the ward and were located only in the pelvic area. During the progression of the disease, new lesions affected the skin of the patient's back, abdomen, chest and side parts of the neck. The occurrence of new macules was not related to any trigger factor. The patient did not present any signs of systemic disease and did not take any medication before the onset of the disease. The patient's family history toward skin diseases was negative. Laboratory test: red blood count, urine analysis, liver and renal parameters, antistreptolysin O titre, and C-reactive protein were negative or did not vary from the normal levels.

In the histopathological examination of the skin biopsy from the trunk, hyperpigmentation of basal membrane cells and accumulation of melanophages in the dermis were observed (Fig. 2). Based on the microscopic picture, patient's history and clinical picture, a diagnosis of IEMP was established.

During the nine months of follow-up, skin lesions presented a stable course, without any subjective complaints. New lesions did not occur. The patient did not receive any treatment.

Discussion

Idiopathic eruptive macular pigmentation is an entity which is rarely found in everyday dermatological practice. Up till now, only a few dozen cases have been described

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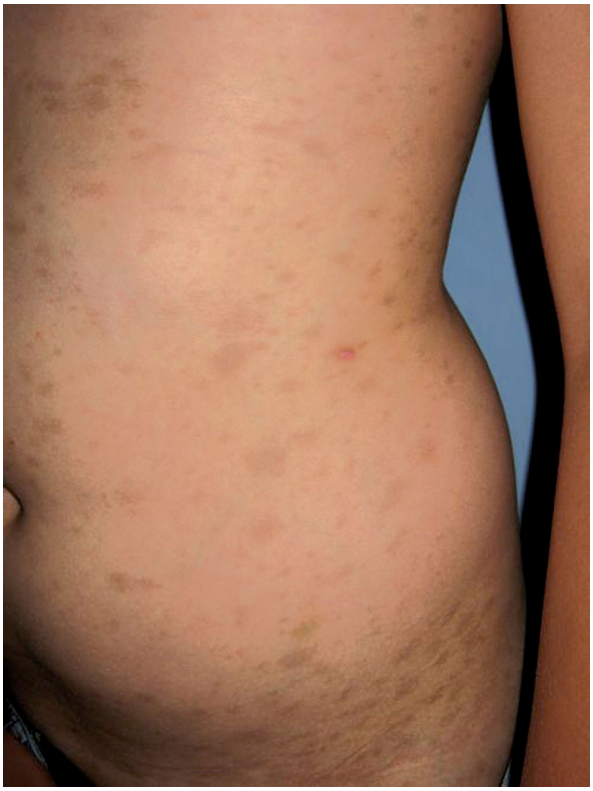

Fig. 1. Disseminated greyish-brown macules of the trunk

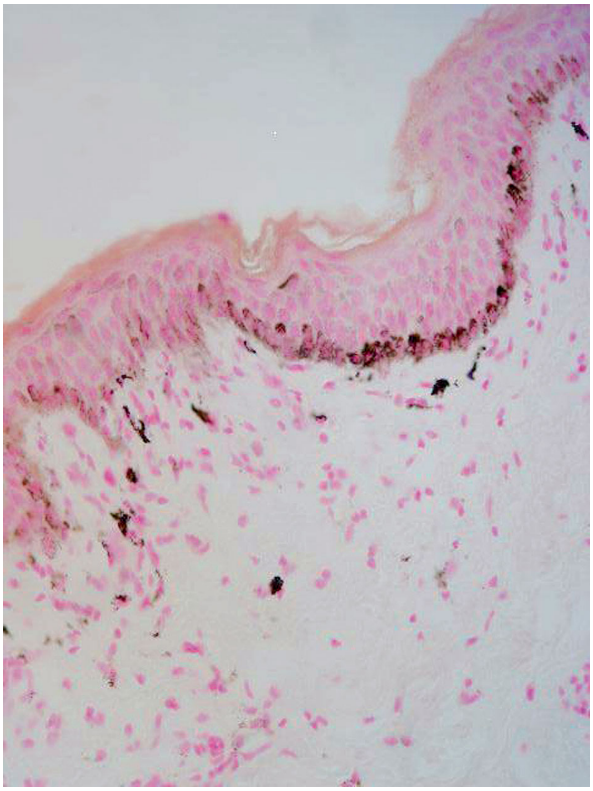

Fig. 2. Hyperpigmentation of the basal layer of the epithelium with accumulation of melanophages in the dermis (Fontana-Masson staining, original magnification 40×)

[1–3]. The aetiopathogenesis of IEMP remains unclear. It appears that the disease does not have a genetic background; only sporadic cases of the disease are observed. The dermatosis is not related to excessive exposure to ultraviolet light, according to the fact that lesions appear on sun-protected skin. There is a hypothesis that IEMP may be related to hormonal disorders, as the lesions often appear during puberty [4]. Other authors classify IEMP as an end step of lichen planus, due to some common patterns in the microscopic picture in both diseases [5].

The entity was described for the first time in 1978 by Degos *et al.* The scientist performed a comparative analysis of various skin diseases with disturbed pigmentation, including the most common one, erythema dyschromicum perstans (ashy dermatosis), and distinguished distinctive clinical and histopathological characteristics of the disease, later named by the authors as idiopathic eruptive macular pigmentation [6]. Eighteen years later, Sanz de Galdeano *et al.* recommended the diagnostic criteria of IEMP (Tab. 1) [7]. According to the authors, the presence of asymptomatic, well-separated, brownish macular lesions, with a tendency to affect the skin of the trunk, neck and proximal parts of extremities, and with higher frequency in children and teenagers, is necessary for the diagnosis of IEMP. Absence of inflammation in the skin and a negative history of drug intake indicate the diagnosis of IEMP. The next diagnostic criterion is the presence of increased pigmentation of basal layer cells and the presence of melanophages in the histopathological picture. The basal layer of the epidermis usually remains intact; nevertheless, perivascular inflammatory infiltrations can be noticed. The last criterion is the proper count of mast cells in the microscopic picture.

The differential diagnosis of idiopathic eruptive macular pigmentation includes several entities. Diseases with similar macular lesions should be taken into consideration (Tab. 2). Erythema dyschromicum perstans usually affects dark-skinned patients or, in our latitude, those with skin phototype III or IV. In the clinical picture, disseminated, macular hyperpigmentation on the sides of the trunk and extremities can be observed. Skin lesions, in contrast with IEMP, have a greyish-brown colour, can be surrounded by an inflammatory rim and frequently show a tendency to blend. A significant symptom which differentiates ashy dermatosis from IEMP is erythematous skin inflammation, preceding lesions, usually provoked by infection or exposure to toxic factors. The classification and the relation between these two entities is still a matter of discussion.

Tab. 1. Diagnostic criteria of idiopathic eruptive macular pigmentation according to de Galdeano *et al*. [7]

1. Brownish, not confluent, asymptomatic macular lesions on trunk, neck and proximal extremities in children and youth
2. Absence of skin inflammation preceding the occurrence of lesions
3. Absence of drug exposure
4. Hyperpigmentation of epidermal basal layer cells and noticeable melanophages without destruction of basal layer or inflammatory infiltration
5. Normal number of mast cells in histopathological picture

Tab. 2. The most frequent entities needed to be differentiated with idiopathic eruptive macular pigmentation

Ashy dermatosis
Incontinentia pigmenti
Drug-induced exanthema
Macular amyloidosis
Café au lait macules
Systemic diseases: haemochromatosis, hyperthyroidism, Addison disease
Post-inflammatory hyperpigmentation (i.e. acne, atopic dermatitis, chronic neurodermatitis)
Other dermatoses: urticaria pigmentosa, lichen planus pigmentosus, atrophoderma of Pasini and Pierini, linear atrophoderma

There are suggestions that concerning the similarity of the histopathological picture, IEMP is only a variant of erythema dyschromicum perstans. Other authors suggest that due to the different features in the personal history and clinical picture, IEMP should be distinguished as a distinctive skin pigmentation disorder [8].

Hyperpigmentation is a common feature of urticaria pigmentosa. Due to characteristic features in the clinical (macular, reddish-brown lesions with positive Darier's sign) and histopathological picture (increased number of mast cells), this dermatosis is relatively easily distinguished from IEMP.

Another group of disorders that should be differentiated from IEMP is genetic, including incontinentia pigmenti (Bloch-Sulzberger syndrome) and neurofibromatosis (von Recklinghausen disease). These genodermatoses affect not only skin, but also internal organs such as eyes, teeth, and the skeletal system. The background of incontinentia pigmenti is inability of cells of the epidermal basal layer to accumulate melanin. Subsequently, the pigment accumulates in the dermis, inducing inflammation. Skin lesions may occur even in the neonatal period and have polymorphic character – apart from macular lesions, erythema, blisters and papules might be present.

The presence of *café au lait* macules is a characteristic feature of von Recklinghausen disease. Unlike IEMP, lesions do not resolve spontaneously and are one of multiple symptoms of von Recklinghausen disease. *Café au lait* macules, it should be emphasized, are present in approximately 10% of the healthy population and are not accompanied by anomalies in organs other than the skin [9].

The IEMP requires differentiation with macular and lichenoid variant of amyloidosis – a disease from the group of storage diseases. The presence of confluent macular lesions with a wrinkled surface is one of the clinical symptoms of amyloidosis. Hyperpigmentation may appear in similar body areas as in IEMP, i.e. on extremities. Skin lesions are accompanied by pruritus, a symptom that is usually absent in IEMP. The fundamental differentiating feature is seen in the microscopic picture of the skin biopsy. Skin variants of amyloidosis present amyloid aggregations in the papillary layer of the dermis.

The macular hyperpigmentation may be a side effect of systemic and topical application of some medications. A classical example of drug-induced hyperpigmentation is a bluish-grey or black-purple pigmentation of skin after the antiarrhythmic drug amiodarone. Hyperpigmentation can be a result of various other drugs, most commonly tetracycline derivatives – minocycline, antidepressants (imipramine), anti-malarial drugs, antivirals (zidovudine), chemotherapeutics (bleomycin, busulfan, 5- fluorouracil), psoralens, oral contraceptive drugs or heavy metal salts.

Macular lesions may indicate disorders of the endocrine system. Excessive secretion of melanotropic hormone, which is present in acromegaly or hypothyroidism, causes various skin pigmentation disorders. Excessive melanogenesis and the presence of disseminated hyperpigmentations may be caused by adrenal gland disorders. In all those conditions, skin lesions are secondary to systemic diseases.

Observations of the natural course of idiopathic eruptive macular pigmentation indicate that the disease has a chronic but spontaneously resolving character. Macules usually disappear within 1-2 years of the disease duration [2, 7]. Occasionally, a remarkable chronic course, over 20 years, has been observed [10].

Concerning the significant aesthetic defect, attempts of various methods of IEMP treatment have been made. However, observations indicate that none of the described therapies – topical ketoconazole, steroids, retinoids or laser therapy – accelerated the resolution of macular lesions [10].

In the context of the above observations, idiopathic eruptive macular pigmentation should be considered as a dermatosis of favourable prognosis without the necessity of therapeutic interventions.

References

1. Plantin P, Le Berre A, Le Roux P, et al. Idiopathic macular pigmentation. Arch Fr Pediatr 1993; 50: 607-8.
2. Stinco G, Favot F, Scott CA, et al. Pigmentatio maculosa eruptive idiopathica: a case report and review of the literature. Int J Dermatol 2007; 46: 1267-70.
3. Marins de Arruda Câmara V, Lupi O, Piñeiro-Maceira J. Idiopathic eruptive macular pigmentation. Int J Dermatol 2008; 47: 272-5.
4. Jang KA, Choi JH, Sung KS, et al. Idiopathic eruptive macular pigmentation: Report of 10 cases. J Am Acad Dermatol 2001; 44: 351-3.
5. Trcko K, Marko PB, Miljkovic J. Idiopathic eruptive macular pigmentation. Acta Dermatovenerol Alp Panonica Adriat 2005; 14: 30-4.
6. Degos R, Civatte J, Belaich S. La pigmentation maculeuse eruptive idiopathique. Ann Dermatol Venerol 1978; 105: 177-82.
7. De Galdeano CS, Leaute-Labreze C, Bioulac-Sage P, et al. Idiopathic eruptive macular pigmentation: report of five patient. Pediatr Dermatol 1996; 13: 274-7.
8. Volz A, Metze D, Bohm M, et al. Idiopathic eruptive macular pigmentation in a 7-year-old girl: case report and discussion of differences from erythema dyschromicum perstans. Br J Dermatol 2007; 157: 799-846.
9. Landan M, Krafchik B. The diagnostic value od café-au-lait macules. J Am Acad Dermatol 1999; 40: 877-90.
10. Mehta S, Aasi S, Cole R, et al. Idiopathic eruptive macular pigmentation: a case of 21 years' duration. J Am Acad Dermatol 2003; 49: 280-2.