Idiopathic Eruptive Macular Pigmentation: A Rare and an Under-reported Entity

Sonam Sharma
Department of Pathology, Kalpana Chawla Government Medical College, Karnal, Haryana, India

Corresponding author: Sonam Sharma, Department of Pathology, Kalpana Chawla Government Medical College, Karnal, Haryana, India, Tel: 09999841393; E-mail: drsonamsharma@gmail.com

Received date: April 21, 2018; Accepted date: June 25, 2018; Published date: August 2, 2018

Abstract

Idiopathic eruptive macular pigmentation is an unfamiliar dermatological disorder which is characterized by the presence of multiple discrete asymptomatic brownish-black macules and flat plaques involving the face, neck, trunk, and proximal extremities especially in pediatric population. It is important to differentiate this uncommon entity from other pigmented dermatosis as it undergoes spontaneous resolution without any residual pigmentation or scarring and thus contraindicating any unnecessary aggressive therapeutic interventions. An experience of one such case in a 6-year-old Indian boy is being reported herewith so as to create awareness amongst the dealing physicians.

Keywords: Acanthosis nigricans; Children; Idiopathic eruptive macular pigmentation; Pigmented papillomatosis; Urticaria pigmentosa

Introduction

Idiopathic eruptive macular pigmentation (IEMP) is a rare, benign, self-limiting and an under-diagnosed pigmented skin disorder of an unknown etiopathogenesis [1]. This term was first coined in French by Degos et al. in the year 1978 [2]. This entity was first described in English literature by Sanz de Galdeano et al. in 1996 who published 5 cases of IEMP and based on the history, clinical presentation and histological findings of these cases, they also established its diagnostic criteria which comprised of following identifying characteristics: [a] eruption of brownish, nonconfluent, asymptomatic macules involving the trunk, neck, and proximal extremities in children and adolescents, [b] absence of preceding inflammatory lesions, [c] no previous drug exposure, [d] basal cell layer hyperpigmentation of the epidermis and prominent dermal melanophages without visible basal layer damage or lichenoid inflammatory infiltrate, and [e] normal mast cell count [3].

This criteria for IEMP diagnosis has been widely accepted and subsequently quoted in the published writings since then. However, recently an attempt has been made to revise the diagnostic criteria as authors have speculated that IEMP can be considered as an eruptive variant of acanthosis nigricans because of its close clinical and histological resemblance to acanthosis nigricans i.e. velvety lesions on atypical sites and histological findings of pigmented papillomatosis [4]. Paucity of the knowledge about this unfamiliar melanosis and debates over its controversial existence as a separate disease entity prompted me to report a rare case of IEMP in a 6-year-old Indian boy who was clinically misdiagnosed as a case of urticaria pigmentosa.

Case Report

A 6-year-old healthy boy presented with multiple brownish-black lesions over the face, neck, trunk and bilateral proximal extremities since last 9 months. The lesions gradually increased in number and size over a period of 2 months and became stable since last month. There was no history of any preceding skin lesion, associated pruritis and photoaggravation of the lesions. History of any oral medications/supplements or any topical medications prior to the onset of the eruption was non-contributory. On detailed history taking, parents of child revealed that he was being treated for urticaria pigmentosa earlier but there was no improvement. The child was born of a non-consanguineous marriage. His birth, developmental, personal and family history was insignificant. The general physical and systemic examination revealed no abnormality. His dermatological examination exhibited multiple (approximately 40-50 in number), discrete, smooth, non-scaly, non-atrophic, normesthetic, round to oval hyperpigmented (brownish-black to gray) macules ranging from 0.5 cm to 3 cm in diameter over the face, neck, trunk, and both the upper extremities including flexural folds (Figure 1).

Few of the lesions had a peculiar velvety surface. Mucosae, hair, nails, palms, and the lower extremities were spared. Darier’s sign was negative. The routine haematological, biochemical and microbiological investigations revealed no abnormality. Skin biopsy from one of the pigmented back lesion showed orthokeratosis, mild irregular acanthosis, slight papillomatosis and basal cell layer hyperpigmentation. The upper dermis showed sparse superficial perivascular lymphohistiocytic infiltrate with numerous melanophages.
in the papillary dermis. The mast cell number was normal (Figure 2). Based on these clinical manifestations and histopathological findings, a final diagnosis of IEMP was made. The patient was started on topical tretinoin 0.05% application once daily. On 6 months follow-up the preexisting lesions remained unchanged however, no worsening or development of new lesions was seen.

![Figure 2: Histopathology of the lesional skin (A) Epidermis exhibiting orthokeratosis, mild irregular acanthosis, slight papillomatosis, prominent melanin in the basal cell layer with the upper dermis showing melanophages and sparse superficial perivascular lymphohistiocytic infiltrate (Hematoxylin and eosin, 40X) (B) Magnified view (Hematoxylin and eosin, 200X).](image)

Discussion

IEMP is a rare under-reported skin disorder which is now regarded as the clinical reminiscent of acanthosis nigricans lesions [5-7]. After careful review of the pertinent literature till date, less than 50 cases of IEMP have been documented worldwide with handful of them being reported from India [1,5,6,8-12]. The largest series of 10 cases has been published by Jang KA et al. from Korea [13]. The age of onset of the disease ranges from 1 to 31 years with no sex predilection [14]. The youngest and oldest case described in the literature is that of a 1-year-old and 50-year-old [13,15]. The exact pathogenesis of this disease still remains unclear. Researchers have postulated that hormonal factors and autoimmunity are involved in increased pigment production in IEMP cases [13,14], whereas sunlight which plays a pivotal role in many cutaneous disorders is not at all an important factor for it to occur, as most of these lesions occur in photoprotected areas [16]. Pang YZ et al. have observed that since majority of the IEMP cases reported in the literature involved the young patients of Indian descent (like in the present case), a genetic basis of this disease cannot be ruled out and requires further exploration [17].

Clinically, the characteristic eruption of IEMP is composed of round to oval, circumscribed, non-pruritic, homogeneous pigmented macules and plaques that appear without previous erythematous, papular, or hypochromic lesions, occurring mostly on the trunk, neck, and proximal portion of the extremities. However, lesions occurring in a Christmas tree pattern and limited to flexural areas of the body have also been documented in the literature [18,19]. IEMP needs to be differentiated clinically and histologically from friction melanosis, post-inflamatory hyperpigmentation, fixed drug eruption (FDE), urticaria pigmentosa, lichen planus pigmentosus (LPP), erythema dyschromicum perstans (EDP), pigmentary disorders that involve the flexural areas like Dowling-Degos disease and acanthosis nigricans as these conditions can cause diagnostic conundrum and unnecessary therapeutic interventions [13,19].

Friction melanosis occurs after prolonged rubbing of the skin with nylon towels/brushes and is characterized by skin hyperpigmentation over the bony regions of the back and limbs clinically and pigmentary incontinence with sporadic deposition of amyloid in the papillary dermis on histopathology [20]. Post-inflammatory pigmentation is excluded by the lack of clinical manifestations of a previous dermatosis that commonly affects the dermal-epidermal interface, such as lichen planus, benign lichenoid keratosis, and erythema multiforme [13]. In FDE, there is history of drug intake prior to the onset of eruption. Darier's sign positivity and increase in the number of mast cells histologically is seen in urticaria pigmentosa. LPP and EDP are considered to be the clinical variants of the same disease, as their histological findings are similar [21-23].

LPP is an uncommon variant of lichen planus which is characterized by hyperpigmented, dark brown macules in sun-exposed areas and flexural folds [24] whereas in EDP, slate gray macules with rim of erythema without any predilection for photoexposed sites are seen. Histologically, an atrophic epidermis, vacuolar alteration of the basal cell layer with a scarce lymphohistiocytic lichenoid infiltrate and pigment incontinence are seen in LPP while in EDP, vacuolar changes of the basal layer and prominent pigmentary incontinence are evident on histopathology. Ultrastructural studies in IEMP shows numerous mature melanosomes in basal and suprabasal keratinocytes and macrophages containing clustered melanosomes, but no vacuolar pattern of the basal cell layer, discontinuity of the basal lamina, colloid bodies, or a lichenoid infiltrate that might indicate a diagnosis of LPP or EDP [3,23]. Dowling-Degos disease has an autosomal dominant mode of inheritance and shows a reticulate or confluent distribution. Pigmented comedone-like lesions, follicular hyperkeratotic papules with rim of erythema without any predilection for photoexposed sites are seen. Histopathologically, Dowling-Degos disease and acanthosis nigricans as well as the clinical syndrome of acanthosis nigricans are all considered to be variants of the same disease, as their characteristic histopathological findings are similar [21-23].

IEMP has a benign course and is a self-resolving condition. It has been reported to disappear spontaneously within months to years [13]. However, two unusual cases have been documented in the literature i.e. of a 24-year-old woman lasting 21 years who was characterized by...
several periods of spontaneous resolution followed by recurrences [27] while the other case is of a 24-year-old man who presented with IEMP lesions of 20 years duration. The patient never had spontaneous resolution but his disease stopped progressing followed by a sudden aggravation 16 years later [12].

Conclusion

IEMP is a rare dermatological condition which should always be considered among differentials of the skin pigmentary disorders especially in pediatric age group patients as it a separate entity with characteristic clinical and histological features. Medical unfamiliarity and reluctance of children to undergo skin biopsy has added to its rarity and under-reporting. Nevertheless, a high index of suspicion and knowledge about this entity is important so as to avoid unnecessary treatment because this condition spontaneously resolves with time. However, more insight is required to understand its genesis and to revise its diagnostic criteria in near future.

References