Progressive cribriform and zosteriform hyperpigmentation

Rameshwar M. Gutte

ABSTRACT

Here we report a case of a healthy 30-year-old male who presented to us with progressively increasing reticulate pigmented lesions following lines of Blaschko on the right side of abdomen and back. Skin biopsy revealed increased basal layer pigmentation without nevus cells. A diagnosis of progressive cribriform and zosteriform hyperpigmentation (PCZH) was made on clinicopathological correlation. We also discuss clinicopathological differentials of this peculiar pigmentary disorder.

Key words: Blaschkoid, cribriform, hyperpigmentation, progressive, zosteriform

INTRODUCTION

Progressive cribriform and zosteriform hyperpigmentation, first described by Rower et al. in 1978, is a distinctive clinicopathological pigmentary disorder of the skin.[1] The following criteria was suggested for diagnosis of PCZH (1) uniformly tan cribriform macular pigmentation in a zosteriform distribution; (2) a histologic pattern that consisted of a mild increase in melanin pigment in the basal cell layer and complete absence of nevus cells; (3) no history of rash, injury, or inflammation to suggest postinflammatory hyperpigmentation; (4) onset well after birth with gradual extension - age at onset was in the second decade of life in every case; and (5) lack of other associated cutaneous or internal abnormalities.[2,3] Here we report a case of PCZH for its rarity in Indian literature.

CASE REPORT

A healthy 28-year-old male presented with progressively increasing brown to black macules and patches arranged along the lines of Blaschko on the right side of abdomen and back. The onset of lesions was first from back and the spread towards abdomen. The lesion appeared at the age of 20 and has been progressive since then. He had no history of any preceding eruption, drug intake or trauma to the area. There was no evidence of internal diseases and no family history of skin abnormalities.

Cutaneous examination revealed linear, cribriform, brown to black pigmentation along lines of Blaschko extending from back to abdomen till pubic area on right side [Figure 1]. Multiple macules coalescing at places to form patches were seen and at some places lesions were slightly palpable with velvety appearance. Laboratory studies, including complete blood cell count, liver and renal function tests and serum electrolyte, were within normal limits.

A diagnosis of zosteriform lichen planus, non-hypertrichotic variant of Becker’s nevus and nevoid acanthosis nigricans was thought. A skin biopsy specimen was obtained from most recent lesion which revealed uniformly increased pigmentation within the basal keratinocytes [Figures 2 and 3]. There were few dermal melanophages with mild pigment incontinence without much inflammatory infiltrate in the dermis [Figure 4]. Nevus cells were absent. Absences of lichenoid infiltrate and interface change ruled out lichen planus. Lack of an increase in smooth muscle fiber and hair follicles ruled out Becker’s nevus. Lack of church-spire hyperkeratosis, presence of heavy basal layer pigmentation with mild pigment incontinence ruled out epidermal nevus and acanthosis nigricans. Also clinically there were many skip
Gutte: Progressive cribriform and zosteriform hyperpigmentation

Indian Dermatology Online Journal - January-March 2014 - Volume 5 - Issue 1

Figure 1: Multiple hyperpigmented patches in a cribriform pattern along the lines of Blaschko

Figure 2: Skin biopsy showing moderate epidermal hyperplasia with increased basal layer pigmentation. (H and E, ×40)

Figure 3: Uniformly increased basal layer pigmentation with pigmented basal layer keratinocytes (H and E, ×100)

Figure 4: High power view of one of the rete-ridge showing increased melanin pigmentation and mild pigment incontinence (H and E, ×400).

areas giving a cribriform appearance. Thus, the diagnosis of PCZH was made on clinicopathological correlation.

DISCUSSION

PCZH is a pigmentary disorder of the skin which is a distinctive clinical entity.[4]

Somatic mosaicism that develops during embryogenesis appears to be the underlying etiology. According to a new genetic concept postulated by Happle, for mosaicism in human skin,[5] somatic mosaicism that develops during embryogenesis leads to the proliferation and migration of two mixed populations of melanocytes with different potential for pigment production. The lesions following Blaschko’s lines in PCZH may reflect the clonal migration and proliferation of embryonic melanoblasts, resulting in peculiar cutaneous patterns.[2,3]
Clinically it should be differentiated from other lesions following the lines of Blaschko, such as incontinentia pigmenti, early epidermal nevi, linear and whorled nevoid hypermelanosis, non-hypertrichotic variant of Becker’s nevus, nevoid acanthosis nigricans and linear lichen planus etc., Clinicopathological correlation will be helpful for differentiation of these entities. For example, histopathologically, epidermal nevi may be distinguished by findings of church-spire hyperkeratosis, papillomatosis, acanthosis and mild increased basal layer pigmentation but no pigment incontinence.\(^{[3,4,6,7]}\)

In our case, lichen planus (LP) was the closest differential diagnosis both clinically and histologically. Though pigment incontinence was seen, other features of LP viz-Interface change, lichenoid band of lymphocytic infiltrate were absent while epidermal hyperplasia with increased basal layer pigmentation was against it. Moreover, Cho E et al.,\(^{[3]}\) in their study found pigment incontinence in 43% cases of PCZH. Also in a case reported by Cho SH et al., histology showed pigmentary incontinence.\(^{[8]}\)

Another close differential diagnosis was non-hypertrichotic variant of Becker’s nevus. But lack of increase in dermal smooth muscles and presence of pigment incontinence was against it.

Linear and whorled nevoid hypermelanosis (LWNH) was first described by Kalter and colleagues in 1988.\(^{[6]}\) It has a similar clinical appearance but exhibits diffuse streaks and whorls rather than localized hyperpigmented lesions as in PCZH and has an early onset. In a study involving 16 children with segmental, linear, or swirled hyperpigmentation distributed along the lines of Blaschko, six patients were found to have widespread hyperpigmentation of the LWNH type, and the other 10 demonstrated unilateral hyperpigmentation of the PCZH type.\(^{[7]}\) Choi et al. suggested that PCZH may be considered as late onset LWNH.\(^{[2]}\) Probably many other clinical entities bearing different names such as “zosteriform lentiginous nevus”, “zebra-like hyperpigmentation in whorls and streaks,” “reticulate hyperpigmentation distributed in a zosteriform fashion,” and “reticulate hyperpigmentation of Iijima” may represent PCZH-LWNH spectrum. Blaschko’s lines have been confused with dermatomes most commonly as both the distribution patterns are characterized by a striking demarcation of cutaneous lesions at the midline. However, Blaschko’s lines represent a form of human ‘mosaicism’ and they do not relate to any vascular, neural or lymphatic structures in the skin.\(^{[9]}\) Cho E et al. in their case series found that, all the lesions corresponded to the lines of Blaschko.\(^{[8]}\)

Also since mosaicism is most likely underlying etiology lesions should be Blaschkoid rather than zosteriform.

Accordingly the term zosteriform should not be used for this entity. So considering different names being used for probably the same entity we recommend that progressive cribiform and Blaschkoid hyperpigmentation may be used for all these conditions representing PCZH-LWNH spectrum. It can be further divided into infantile form associated with other anomalies and adult form less likely to have extra-cutaneous features.

REFERENCES

1. Rower JM, Carr RD, Lowney ED. Progressive cribiform and zosteriform hyperpigmentation. Arch Dermatol 1978;114:98-9.
2. Choi JC, Yang JH, Park HS, Chun DK. Progressive cribiform and zosteriform hyperpigmentation – the late onset linear and whorled nevoid hypermelanosis. J Eur Acad Dermatol Venereol 2005;19:638-9.
3. Cho E, Cho SH, Lee JD. Progressive cribiform and zosteriform hyperpigmentation: A clinicopathologic study. Int J Dermatol 2012;51:399-405.
4. Kim SJ, Kim MB, Oh CK, Jung HS, Kwon KS. Three cases of progressive cribiform and zosteriform hyperpigmentation. Korean J Dermatol 2002;40:181-6.
5. Happle R. Mosaicism in human skin. Understanding the patterns and mechanisms. Arch Dermatol 1993;129:1460-70.
6. Kalter DC, Griffiths WA, Atherton DJ. Linear and whorled nevoid hypermelanosis. J Am Acad Dermatol 1988;19:1037-44.
7. Di Lernia V. Linear and whorled hypermelanosis. Pediatr Dermatol 2007;24:205-10.
8. Cho SH, Ha JH, Choi HC, Bae EY, Lee JD. A case of atypical progressive cribiform and zosteriform hyperpigmentation. Pediatr Dermatol 2003;20:792-5.
9. Tagra S, Talwar AK, Walia RS. Lines of Blaschko. Indian J Dermatol Venereol Leprol 2005;71:57-9.
