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A Mosaic of Pigmentation: Adult-Onset Progressive Cribriform and Zosteriform Hyperpigmentation With a Congenital Pigmentary Lesion

Afsina A. Palakkathodi¹, Arun Vignesh¹, Leena Dennis Joseph², Sudha Rangarajan¹, Adikrishnan S¹

1. Department of Dermatology, Venereology and Leprosy, Sri Ramachandra Institute of Higher Education and Research, Chennai, IND 2. Department of General Pathology, Sri Ramachandra Institute of Higher Education and Research, Chennai, IND

Corresponding author: Afsina A. Palakkathodi, afsinaarafap@gmail.com

Abstract

Progressive cribriform and zosteriform hyperpigmentation (PCZH) is a rare acquired pigmentary disorder characterized by unilateral, gradually progressive hyperpigmented macules occurring in the absence of preceding inflammation or systemic abnormalities. Increasing evidence suggests that PCZH may represent a localized manifestation within the spectrum of pigmentary mosaicism. The coexistence of a congenital pigmentary lesion and adult-onset PCZH is rarely described.

We report a 24-year-old man who developed gradually progressive, asymptomatic unilateral hyperpigmented macules over the right neck, shoulder, upper back, upper chest, and mandibular region over two years. The lesions respected the midline. Dermoscopy demonstrated a diffuse light-brown reticular pigment network, while histopathological examination showed basal epidermal hypermelanization with increased epidermal melanocyte density, without interface dermatitis, pigmentary incontinence, or nevocytic proliferation. The clinical and histopathological findings were consistent with PCZH. An additional speckled hyperpigmented patch over the contralateral hip, present since birth, was clinically consistent with pigmentary mosaicism.

This unusual coexistence of adult-onset PCZH and a congenital pigmentary lesion clinically consistent with pigmentary mosaicism highlights the possible relationship between PCZH and the broader spectrum of cutaneous pigmentary mosaicism.

Categories: Pathology, Dermatology

Keywords: acquired blaschkoid hyperpigmentation, blaschko lines, cutaneous mosaicism, dermoscopy, linear and whorled nevoid hypermelanosis, pigmentary mosaicism, progressive cribriform and zosteriform hyperpigmentation

Introduction

Progressive cribriform and zosteriform hyperpigmentation (PCZH) is a rare acquired pigmentary disorder first described by Rower et al. in 1978. It is characterized by gradually progressive, unilateral tan-to-brown macular hyperpigmentation developing after birth without preceding inflammation or associated systemic abnormalities [1]. The term "cribriform" describes a sieve-like or reticulate pigmentary pattern, while "zosteriform" refers to a segmental distribution resembling herpes zoster, distinct from a Blaschkoid distribution that follows the developmental lines of Blaschko. Histopathology typically demonstrates increased basal epidermal pigmentation without nevocytic proliferation [2].

The nosologic position of PCZH remains uncertain. It has been proposed to represent a localized, late-onset variant of linear and whorled nevoid hypermelanosis (LWNH), with both conditions considered within the broader spectrum of pigmentary mosaicism, which refers to patterned pigmentation arising from genetically distinct cutaneous cell populations [3,4].

We report an unusual case of adult-onset PCZH coexisting with a contralateral congenital pigmentary lesion. This presentation highlights the clinical overlap among patterned pigmentary disorders and raises the possibility of a broader relationship between acquired and congenital forms of pigmentary mosaicism.

Case Presentation

A 24-year-old man presented with gradually progressive, asymptomatic hyperpigmentation over the right upper back of two years' duration. The pigmentation initially appeared as multiple light-brown macules over the right upper back and gradually extended to involve the right posterior shoulder, posterolateral neck, supraclavicular region, upper anterior chest, and mandibular region, following the lines of Blaschko. The lesions remained asymptomatic, with no history of pruritus, pain, burning sensation, erythema, scaling,

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preceding inflammatory dermatosis, trauma, or topical or systemic drug exposure. The pigmentation remained strictly unilateral without crossing the midline.

The patient also had a separate asymptomatic hyperpigmented lesion over the left hip that had been present since birth without appreciable progression. There was no family history of similar pigmentary disorders or other genodermatoses. His past medical history was unremarkable, with no history of developmental, neurological, skeletal, or ophthalmological abnormalities.

Cutaneous examination revealed multiple irregular tan-to-light-brown hyperpigmented macules coalescing into reticulate patches in a unilateral segmental distribution involving the right side of the neck and upper chest (Figure 1), right mandible, neck, and shoulder (Figure 2), and posterolateral neck and upper back, without crossing the midline (Figure 3). The distribution was clinically Blaschkoid, extending across multiple anatomical regions rather than being confined to a single dermatome. The lesions were flat, non-scaly, non-indurated, and non-atrophic.



FIGURE 1: Anterior view showing tan brown cribriform macules and patches over right side of neck and upper chest.



FIGURE 2: Lateral view showing tan brown macules and patches over right side of mandible, neck, and shoulder.



FIGURE 3: Posterior view showing cribriform tan brown macules and patches over right posterolateral neck and upper back.

A separate, well-demarcated hyperpigmented patch composed of multiple clustered and coalescing dark-brown macules in a speckled configuration, with irregular but relatively well-defined margins, was present over the left hip (Figure 4). The lesion had been present since birth. Its clinical morphology raised the possibility of a speckled lentiginous nevus (nevus spilus) or another form of patterned pigmentary mosaicism. However, in the absence of dermoscopic and histopathological evaluation, definitive classification was not possible.



FIGURE 4: Congenital speckled hyperpigmented patch over the left hip.

Clinical photograph demonstrating multiple clustered and coalescing dark-brown macules forming an irregular speckled hyperpigmented patch over the left hip.

Based on its congenital onset and clinical morphology, this lesion was considered clinically consistent with pigmentary mosaicism. No associated vascular malformations, connective tissue nevi, limb asymmetry, or skeletal abnormalities were identified.

General physical and systemic examinations were unremarkable.

Dermoscopy of the right-sided lesions revealed a subtle diffuse light-brown reticular pigment network with focal accentuation on a background of homogeneous light brown pigmentation (Figure 5).



FIGURE 5: Dermoscopy of a right-sided lesion showing a subtle light-brown reticular pigment network with focal accentuation on a background of light-brown pigmentation.

In some areas, closely spaced curvilinear brown lines produced a subtle fingerprint-like appearance (Figure 6).

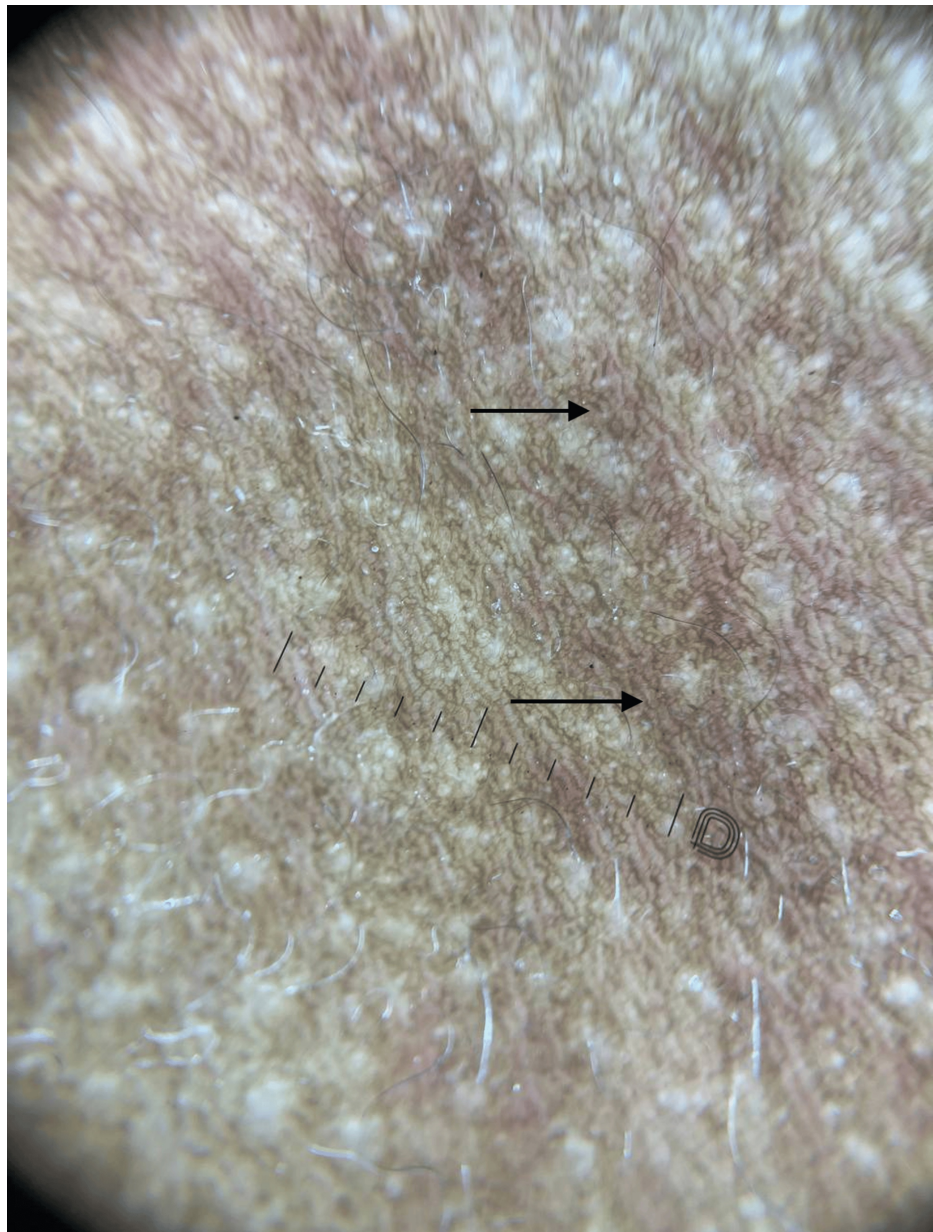


FIGURE 6: Dermoscopy of right-sided lesion showing focal, closely spaced curvilinear brown lines producing a fingerprint-like appearance (black arrows).

Blue-gray granules, annular structures, and features suggestive of lichenoid dermatitis were absent.

Routine hematological and biochemical investigations were within normal limits.

Histopathological examination of a biopsy obtained from a lesion over the right upper back revealed basket-weave orthokeratosis with mild acanthosis and preserved epidermal architecture. The basal layer showed diffuse hypermelanization without interface dermatitis, basal vacuolar degeneration, pigmentary incontinence, dermal melanophages, or significant inflammatory infiltrate. Increased epidermal melanocyte density was reported on histopathological examination, without nevocytic proliferation or cytological atypia (Figures 7, 8). Melanocytic immunohistochemical staining was not performed. Dermoscopy and histopathological examination of the congenital left-hip lesion were not performed.

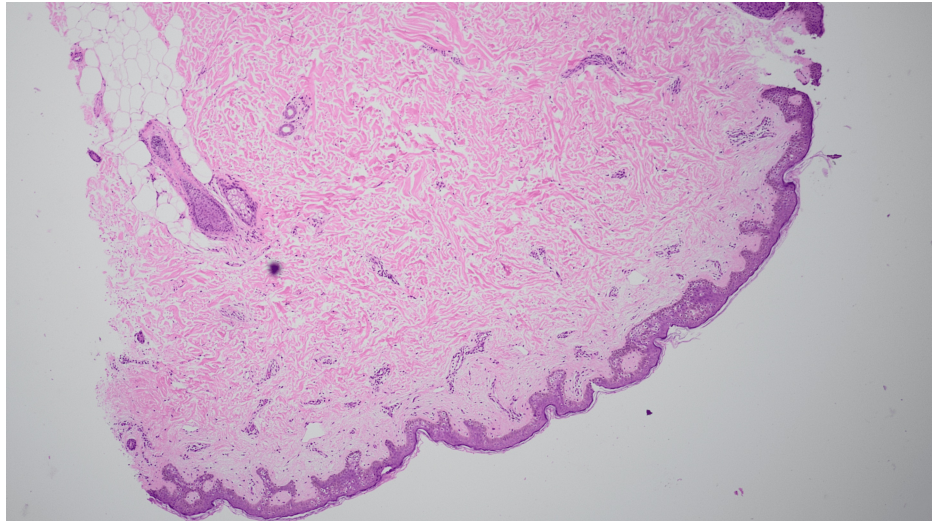


FIGURE 7: Low-power histopathological features of the right-sided lesion.

Low-power photomicrograph of a biopsy from the right upper back showing preserved epidermal architecture with mild acanthosis and elongation of rete ridges, without a conspicuous dermal inflammatory infiltrate (hematoxylin and eosin, 40x).

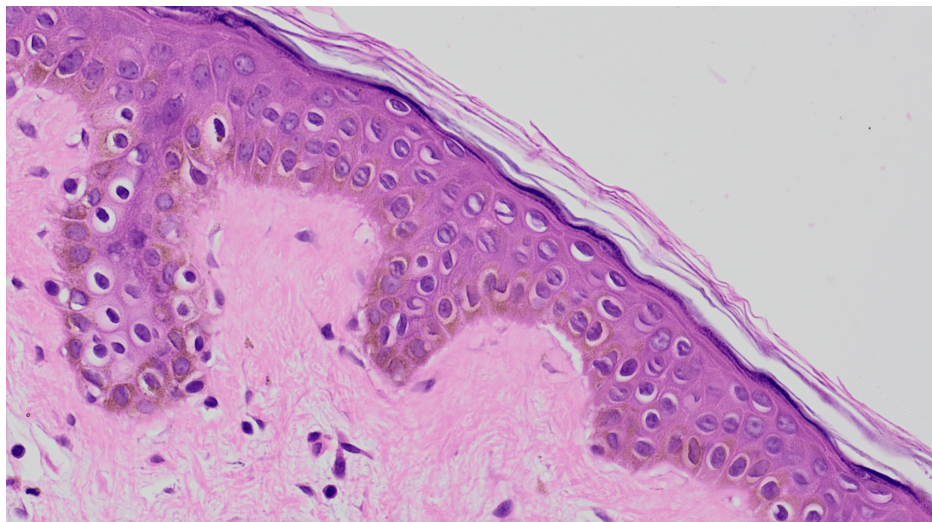


FIGURE 8: Higher-power histopathological features of the right-sided lesion.

High-power photomicrograph of a biopsy from the right upper back showing increased basal epidermal melanocytes, mild acanthosis, and preserved epidermal architecture, without significant interface changes or dermal inflammatory infiltrate (hematoxylin and eosin-stained section, x400).

Based on the clinical presentation and histopathological findings, a diagnosis of PCZH was made. The patient was counselled regarding the benign nature of the condition and available treatment options, and the patient opted for observation.

Discussion

PCZH is a rare acquired pigmentedary disorder first described by Rower et al. in 1978. It is characterized by unilateral cribriform macular hyperpigmentation in a zosteriform distribution, onset well after birth with gradual progression, absence of preceding inflammation, increased basal pigmentation without nevus cells on histopathology, and absence of associated systemic abnormalities [1]. Our patient fulfilled the diagnostic criteria proposed by Rower et al., with adult onset, gradual progression, unilateral Blaschkoid hyperpigmentation respecting the midline, absence of preceding inflammation or trauma, compatible

histopathological findings, and no extracutaneous abnormalities [1].

The clinical presentation was consistent with previous reports of PCZH, including the largest clinicopathological series by Cho et al. and individual reports from different geographic regions [2,5-8]. Although PCZH was initially described predominantly in younger individuals, Cho et al. reported an age of onset ranging from birth to the fifth decade, supporting adult-onset PCZH as part of its clinical spectrum [2].

Dermoscopy in our patient demonstrated a subtle reticular pigment network with focal fingerprint-like structures over a homogeneous light-brown background (Figures 5, 6). These findings are compatible with the limited dermoscopic descriptions of PCZH reported in the literature [9]. Given the paucity of published dermoscopic data, further reports may help characterize its dermoscopic spectrum.

Histopathologically, PCZH typically demonstrates increased basal melanization without nevocytic proliferation [1,2]. Cho et al. reported significantly increased basal melanin without a statistically significant increase in melanocyte number [2]. In our patient, diffuse basal hypermelanization was accompanied by a mildly increased number of epidermal melanocytes, without cytological atypia or nevocytic proliferation. This finding may represent histopathological variation within PCZH. The absence of interface dermatitis, basal vacuolar degeneration, pigmentary incontinence, dermal melanophages, or significant inflammation also argued against an inflammatory or lichenoid pigmentary disorder.

The principal differential diagnoses included LWNH, Becker nevus without hypertrichosis, and segmental lentiginosis. LWNH was considered because of its Blaschkoid distribution and histopathological similarities; however, its usual congenital or early-life onset and more extensive linear and whorled distribution contrasted with the localized adult-onset presentation in our patient [3]. Becker nevus without hypertrichosis was considered less likely because of the absence of characteristic dermal smooth-muscle and pilosebaceous hyperplasia [10]. Segmental lentiginosis was considered less likely because of the reticulate Blaschkoid morphology rather than discrete lentigines and the absence of a lentiginous histopathological pattern [11]. Lichen planus pigmentosus was unlikely because of the absence of clinical inflammation and interface dermatitis, pigmentary incontinence, and dermal melanophages on histopathology [10].

The nosological position of PCZH remains debated. Di Lernia proposed that PCZH may represent a localized, late-onset variant of LWNH, as both disorders share similar clinical distribution, histopathological features, and presumed pathogenesis, differing mainly in age at onset and extent of involvement [3]. More broadly, patterned pigmentary abnormalities following the lines of Blaschko are recognized within the clinical spectrum of pigmentary mosaicism [12]. The Blaschkoid distribution in our patient is compatible with this concept, although molecular analysis was not performed.

Pigmentary mosaicism comprises a heterogeneous group of pigmentary disorders resulting from genetic mosaicism, often arising from postzygotic genetic alterations that give rise to genetically distinct cell populations. Rather than representing a single disease entity, it encompasses a spectrum of hyperpigmented, hypopigmented, or mixed pigmentary abnormalities distributed in characteristic patterns, including the lines of Blaschko, checkerboard, phylloid, patchy patterns without midline separation, and lateralization [13]. Current terminology favors the umbrella term pigmentary mosaicism over several older descriptive diagnoses, reflecting a common framework of cutaneous mosaicism despite variable clinical phenotypes [12].

Pigmentary mosaicism frequently becomes apparent at birth or during early childhood and may occur either as an isolated cutaneous finding or in association with extracutaneous abnormalities [12]. In our patient, the congenital left hip lesion had remained stable since birth, with no associated systemic abnormalities. Based on its congenital onset and clinical morphology, the lesion was considered clinically consistent with pigmentary mosaicism. However, as histopathological and molecular evaluation of this lesion was not performed, its precise classification could not be established.

The coexistence of these two clinically distinct pigmentary phenotypes is noteworthy. El-Komy et al. reported PCZH occurring with congenital nevus depigmentosus, while Han and Chang described PCZH associated with vitiligo [14,15]. These observations demonstrate that PCZH may coexist with other pigmentary disorders, although they do not establish a common molecular mechanism. In the present case, whether the congenital lesion and adult-onset PCZH represent independent mosaic events or phenotypic manifestations within a broader mosaic process remains uncertain. Molecular analysis of affected skin would be required to investigate this possibility.

This case highlights adult-onset PCZH occurring in association with a contralateral congenital pigmentary lesion clinically consistent with pigmentary mosaicism. The coexistence is of interest in the context of the evolving classification of PCZH and other patterned pigmentary disorders within the spectrum of cutaneous mosaicism. Further reports incorporating molecular genetic analysis may help clarify the relationship between PCZH, LWNH, and pigmentary mosaicism.

Conclusions

This case highlights the rare coexistence of adult-onset PCZH with a contralateral congenital pigmentary lesion. The distinctive clinical presentation, supported by dermoscopic and histopathological findings, emphasizes the importance of clinicopathological correlation in diagnosing PCZH. The presence of two distinct pigmentary patterns in the same individual adds to the clinical spectrum of pigmentary disorders and warrants further investigation into their possible associations.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

Concept and design: Afsina A. Palakkathodi, Arun Vignesh

Acquisition, analysis, or interpretation of data: Afsina A. Palakkathodi, Arun Vignesh, Leena Dennis Joseph, Sudha Rangarajan, Adikrishnan S

Drafting of the manuscript: Afsina A. Palakkathodi, Leena Dennis Joseph

Critical review of the manuscript for important intellectual content: Afsina A. Palakkathodi, Arun Vignesh, Sudha Rangarajan, Adikrishnan S

Disclosures

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