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CASE AND RESEARCH LETTERS

Progressive Cribriform and Zosteriform Hyperpigmentation[☆]

Hiperpigmentación zosteriforme y cribriforme progresiva

To the Editor

Progressive cribriform and zosteriform hyperpigmentation (PCZH) is a skin disease first described by Rower et al.¹ in 1978. Since then, several cases have been published in Korea but only 3 articles in the dermatology literature in English.²⁻⁴

This letter reports the case of a woman with hyperpigmented lesions on the right arm and on the right side of the trunk since childhood. These lesions were consistent with a diagnosis of PCZH.

The woman, aged 54 years, was white and had a personal history of fibromyalgia and asthma. She attended the clinic because of an asymptomatic hyperpigmented lesion on the back that had been present since she was 9 years old. Examination revealed several macular lesions with a homogeneous brown color and well-defined but irregular borders, grouped in a segmental pattern on normal skin on the right flank and arm, and the right side of the back and abdomen. These lesions did not cross the midline of the trunk (Figs. 1 and 2). No café-au-lait spots or neurofibromas were seen on unaffected skin.

The patient did not recall any rash or prior trauma of the affected area although the lesions did increase in size in the first years after onset. She did not have any extracutaneous abnormalities or a family history of neurofibromatosis or similar skin lesions. A biopsy was taken from the affected area of the back. Histopathology showed an increase in cytoplasmic pigmentation of the cells of the basal epidermal layer (Fig. 3). In view of the clinical and histologic characteristics, a diagnosis of PCZH was made.

Rower et al.¹ reported 5 patients with pigmentary abnormalities with certain characteristics in common that constituted the diagnostic criteria for PCZH. These characteristics were uniform and cribriform brown macular hyperpigmentation with a zosteriform distribution; a his-



Figure 1 Macular lesions of even brown color with a reticulate pattern on normal skin, on the right side of the back, without crossing the midline.

topologic pattern consistent with a mild increase in melanin pigment in cells of the basal layer and complete absence of nevus cells; no history of rash, injury, or inflammation that could suggest postinflammatory hyperpigmentation; onset after birth with gradual increase in extension; and absence of other cutaneous or internal abnormalities.

In 2012, Cho et al.⁴ published demographic, clinical, and histopathologic data from 30 Korean patients with PCZH. The authors observed a slight predominance of men, with an age of onset of PCZH between birth and 54 years (77% between 1 and 15 years). Only one individual reported a family history of similar lesions and none had associated extracutaneous congenital abnormalities. In general, the lesions were asymptomatic, cribriform, with a Blaschkoid distribution, and located preferentially on the trunk and limbs with a slight tendency towards right-sided predominance. The affected skin, in comparison with adjacent skin, showed increased pigmentation (more melanin granules) of the basal layer. Nevus cells were not present and there were no significant differences in the number of melanocytes. Likewise, there were no differences in other skin disorders except for cases with pigmentary incontinuity. No effective treatment has been reported for PCZH. Terms such as *reticulate hyperpigmentation of Iijima*⁵ and *reticulate zosteriform hyperpigmentation*⁶ have been used to describe conditions with similar clinical and histopathologic findings.

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Figure 2 Macular hyperpigmented lesions, with well-defined but irregular borders, grouped in a unilateral segmental pattern on the right side of the body. A, Lateral aspect of the right arm. B, Right chest wall. C, Right side of the abdomen.

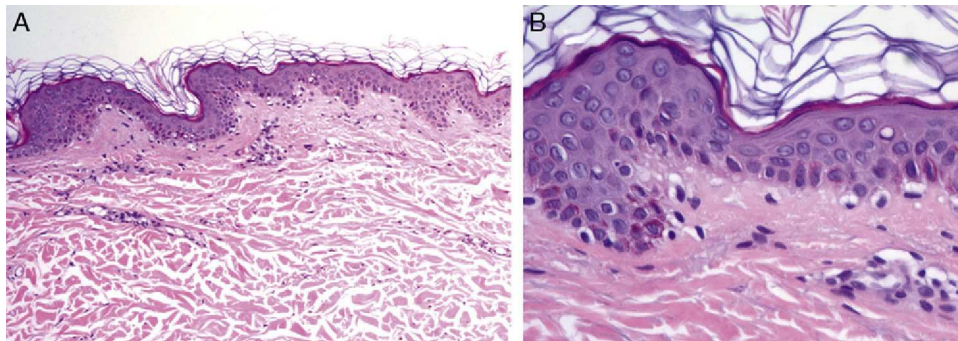


Figure 3 A, In the epidermis, increased pigmentation in cells of the basal layer (hematoxylin-eosin, original magnification $\times 100$). B, Higher magnification, cytoplasmic pigmentation in cells of the basal epidermal layer (hematoxylin-eosin, original magnification $\times 400$).

Differential diagnosis of PCZH includes skin conditions that present with segmental hyperpigmented lesions such as linear and whorled nevoid hypermelanosis,^{4,5,8} café-au-lait spots,^{8,9} segmental pigmentation disorders with onset in the first months of life,^{8,9} and others such as Becker nevus (variant without hypertrichosis).^{4,8,10} Linear and whorled nevoid hypermelanosis is a diffuse asymmetric hyperpigmentation in lines or swirls along the Blaschko lines. The lesions appear at birth or within the first few weeks of life and then spread or darken during the first 2 years. These lesions are sometimes associated with extracutaneous disorders, particularly of neurologic, cardiac, and musculoskeletal nature.^{4,7,8} Despite the diffuse pattern, associated congenital abnormalities, and the different age of onset compared to PCZH, the clinical manifestations may sometimes overlap,⁷ and so some authors consider these conditions as part of spectrum of the same disease.^{2,3} Café-au-lait spots are usually round or oval, although they can follow a segmental distribution in segmental neurofibromatosis type 1 and have irregular borders in the McCune-Albright and Jaffe-Campanacci syndromes.^{8,9} Segmental pigmentation abnormalities appear in the first months of life as

segmental hypopigmentation or hyperpigmentation, mainly on the trunk. They follow a block-like pattern with sharp confinement at the midline, particularly anteriorly, and can be associated with extracutaneous abnormalities.^{8,9}

In conclusion, the findings in our case are similar to those reported by Cho et al.⁴ Bearing in mind the absence of associated conditions and the lack of therapeutic options, it is likely that PCZH is in fact more common than the limited number of reports in the literature would have us believe.

References

1. Rower JM, Carr RD, Lowney ED. Progressive cribriform and zosteriform hyperpigmentation. *Arch Dermatol.* 1978;114:98–9.
2. Schepis C, Alberti A, Siragusa M, Romano C. Progressive cribriform and zosteriform hyperpigmentation: the late-onset feature of linear and whorled nevoid hypermelanosis associated with congenital neurological, skeletal and cutaneous anomalies. *Dermatology.* 1999;199:72–3.
3. Choi JC, Yang JH, Lee UH, Park HS, Chun DK. Progressive cribriform and zosteriform hyperpigmentation - the late onset

- linear and whorled nevoid hypermelanosis. *J Eur Acad Dermatol Venereol.* 2005;19:638–9.
4. Cho E, Cho SH, Lee JD. Progressive cribriform and zosteriform hyperpigmentation: a clinicopathologic study. *Int J Dermatol.* 2012;51:399–405.
 5. Iijima S, Naito Y, Naito S, Uyeno K. Reticulate hyperpigmentation distributed in a zosteriform fashion: a new clinical type of hyperpigmentation. *Br J Dermatol.* 1987;117:503–10.
 6. Patrizi A, di Lernia V, Varotti C. Reticulate hyperpigmentation distributed in a zosteriform fashion. *Br J Dermatol.* 1989;121:280.
 7. Di Lernia V. Linear and whorled hypermelanosis. *Pediatr Dermatol.* 2007;24:205–10.
 8. Gibbs NF, Makkar HS. Disorders of hyperpigmentation and melanocytes. In: Eichenfield LF, Frieden IJ, Esterly NB, editors. *Neonatal dermatology.* 2nd ed. Philadelphia: Saunders Elsevier; 2008. p. 397–422.
 9. Hogeling M, Frieden IJ. Segmental pigmentation disorder. *Br J Dermatol.* 2010;162:1337–41.
 10. Molho-Pessach V, Schaffer JV. Blaschko lines and other patterns of cutaneous mosaicism. *Clin Dermatol.* 2011;29:205–25.
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Dermatoscopy of Apocrine Tumors: Report of 4 Cases[☆]

Dermatoscopia de tumores apocrinos: presentación de 4 casos

To the Editor

Hidrocystomas and hidradenomas are adnexal tumors that originate from sweat glands and are typically of apocrine origin. They are usually diagnosed histologically because they lack characteristic clinical findings.^{1–5}

Dermatoscopy is a noninvasive technique that can help in the diagnosis of these tumors. We present the dermatoscopic features of 3 cases of apocrine hidrocystomas and 1 case of a solid-cystic apocrine hidradenoma.

Case 1: A 68-year-old man attended the clinic because of an asymptomatic lesion that had appeared 6 months earlier in the right parietal region (Fig. 1A). The mildly erythematous nodule measured 1 cm in diameter. The dermatoscopic image showed a homogeneous pattern of pale grey/bluish color, with whitish cotton wool-like structures and linear irregular vessels (Fig. 2A). Histopathology showed a cystic structure lined by an epithelium formed of a double layer of cuboidal cells with no cytologic atypia and areas of decapitation secretion, consistent with diagnosis of apocrine hidrocystoma (Fig. 3A).

Case 2: A 62-year-old woman attended the clinic because of an asymptomatic lesion on the right helix that had been present for several years. The translucent cystic lesion measured 1 cm in diameter (Fig. 1B). The dermatoscopic image showed a homogeneous pattern of pale grey/bluish color, with brown-orange regions to the left of the lesion, whitish cotton wool-like structures, and linear irregular vessels

(Fig. 2C). Histology was consistent with apocrine hidrocystoma (Fig. 3B).

Case 3: A 30-year-old woman had a 1 cm nodule on a sebaceous nevus in the interparietal region (Fig. 1C). The dermatoscopic image showed a homogeneous pattern of pale grey/bluish color, with whitish cotton wool-like structures, and linear irregular vessels (Fig. 2C). Histology confirmed the diagnosis of apocrine hidrocystoma.

Case 4: An 81-year-old woman attended the clinic because of an asymptomatic lesion on the right arm. Clinical examination revealed a well-defined, erythematous nodule measuring 1.5 cm in diameter (Fig. 1D). The dermatoscopic image showed a homogeneous pattern of pale grey/bluish color, with brown-orange areas, whitish cotton wool-like structures, and linear irregular vessels peripherally (Fig. 2D). Histologically, the lesion was a nonencapsulated dermal tumor comprised of cells with no atypia and abundant eosinophilic cytoplasm and cells whose nuclei had a more basaloid and hyperchromatic appearance. Foci of clear cells could be seen. The lesion showed cystic formations with mucin in the lumen, as well as a sclerotic and vascularized stroma (Fig. 3C). The above was compatible with a diagnosis of solid-cystic apocrine hidradenoma.

Apocrine hidrocystomas are benign adnexal cystic tumors of unknown etiology. Possible causes include sweat duct occlusion or adenomatous cystic proliferation of apocrine glands (apocrine cystadenoma). Histologically, they usually have multiple cystic spaces, with papillary protrusions and a wall composed of an inner layer of cuboidal cells and an outer layer of myoepithelial cells. These lesions are usually surrounded by a fibrous stroma with small-diameter vessels inside.^{2,3}

Apocrine hidradenoma is a benign adnexal tumor of uncertain histogenesis. It is a multilobed, nonencapsulated, well-defined neoplasm comprised mainly of neoplastic epithelial cells and small ductal lumens. There are several histologic patterns: nodular hidradenoma, solid-cystic hidradenoma, and clear-cell hidradenoma.^{1,4,5}

Correct diagnosis of hidrocystoma and hidradenoma is difficult solely on the clinical presentation and subsequent

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