Dermatology Practical & Conceptual

Verrucous Epidermal Nevus: Dermoscopy, Reflectance Confocal Microscopy, and Histopathological Correlation

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Introduction

Verrucous epidermal nevus (VEN) is a benign, noninflammatory malformation usually present at birth or occurring within the first years of life. It is composed of keratinocytes that arise from pluripotential germ cells in the basal layer of the embryonic ectoderm and results from mosaic postzygotic mutations. VEN clinically appears as skin-colored to brown, sharply demarcated, papillomatous papules coalescing into plaques. The diagnosis is usually based on clinical presentation and, in selected cases, on histopathology examination that reveals epidermal hyperplasia, papillomatosis with elongation of rete ridges, and pigmented keratinocytes surrounding the dermal papillae. Dermoscopy and reflectance confocal microscopy (RCM) are valuable, noninvasive techniques that support the diagnosis in clinical practice in several fields of dermatology including cutaneous tumors as well as inflammatory and infectious dermatoses [1]. Dermoscopy has shown in 8 cases of VEN the characteristic presence of large brown circles, consisting of hyperchromic brown edge surrounding a hypochromic area.

The aim of this study was to evaluate and correlate dermoscopy (Dermlite; 3Gen, San Juan Capistrano, CA, USA), RCM (VivaScope3000; Caliber I.D., Rochester, NY, USA) and histopathological findings in a series of 9 patients with a clinical and/or histopathologically proven diagnosis of VEN. In all cases VEN had been present since early infancy. All participants provided informed consent.

Case Presentation

All examined lesions—3 located on the head, 4 on the neck, and 2 on the trunk—clinically appeared as velvety plaques with linear distribution and variable degree of pigmentation, ranging from light to dark brown (Figure 1A). At dermoscopy, all lesions showed the presence of large brown circles on a brownish background. No pigment network, globules, comedo-like openings, or milia-like cysts were observed (Figure 1B). RCM revealed, in all cases, a normal epidermal honeycomb pattern (bright polygonal cellular outlines and dark nuclei), papillomatosis, and the constant presence of a well-defined, strongly bright rim of

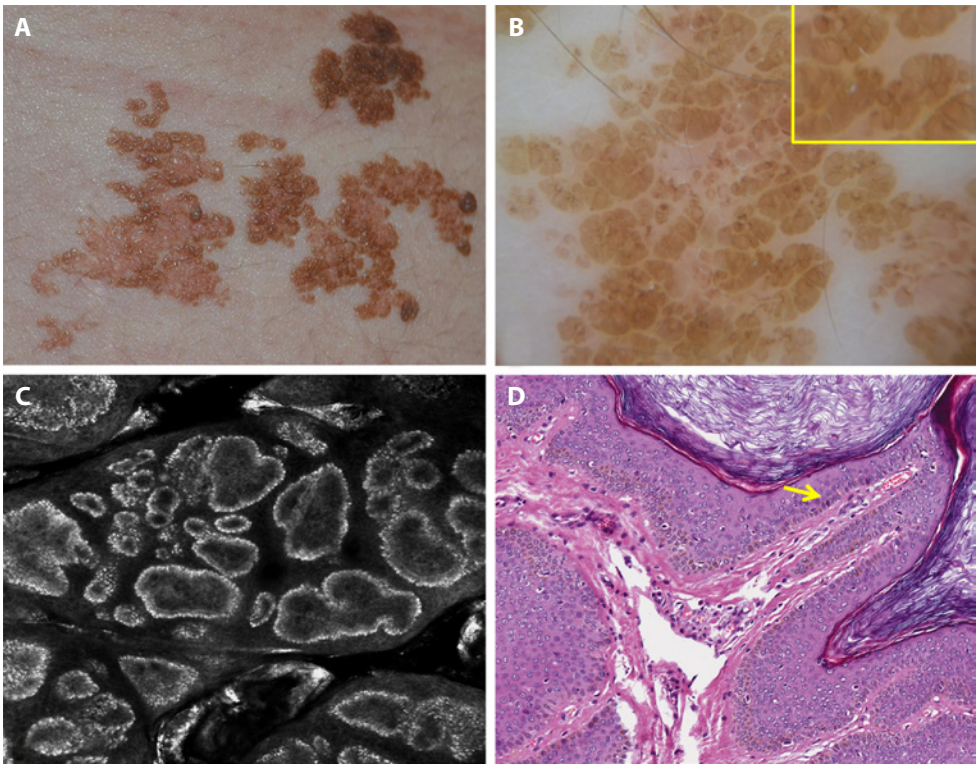

Figure 1. VEN of the neck. (A) Clinical aspect. (B) Dermoscopy showing large brown circles (insert: a detail) on a brownish background. (C) RCM showing a well-defined, strongly bright rim of monomorphous keratinocytes demarcating each dark dermal papilla. (D) Histopathology showing arrangement of pigmented keratinocytes surrounding the dermal papillae (arrow). [Copyright: ©2019 Verzì et al.]

monomorphous cells demarcating each dark dermal papilla throughout the lesion (Figure 1C).

Conclusions

To the best of our knowledge, this is the first study describing the RCM features of VEN. In our series, VEN showed peculiar dermoscopic and RCM findings. In particular, the typical presence at dermoscopy of large brown circles, as previously reported [2], correlated with dermal papillae demarcated by a rim of monomorphous and strongly bright cells seen at RCM and corresponds to the histological arrangement of pigmented keratinocytes surrounding the dermal papillae (Figure 1D).

The dermoscopic brown circles have been observed, along with other findings, in seborrheic keratosis and pigmented squamous cell carcinoma, 2 disorders that, however, show different age at onset and clinical features. A similar ringed pattern at RCM may also be observed in solar lentigo, lentigo simplex, and ink spot lentigo; in the first case, the bright ring is due to hyperpigmentation of the basal keratinocytes, and in the remaining 2 to hyperproliferation of melanocytes. However, these disorders are clinically different from VEN presenting with flat, nonpalpable, pigmented lesions.

In conclusion, our study suggests that the combined use of dermoscopy and RCM may assist in the enhanced diagnosis of VEN, especially in those cases with unusual clinical presentation.

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