Dermoscopic Findings in Laugier-Hunziker Syndrome

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Background: Laugier-Hunziker syndrome (LHS) is a rare, acquired mucocutaneous hyperpigmentation often associated with longitudinal melanonychia. The clinical behavior of mucocutaneous pigmented lesions ranges from benign to highly malignant. Therefore, in most cases, the clinical diagnosis should be confirmed by further diagnostic methods. Dermoscopy is a noninvasive technique that has been used to make more accurate diagnoses of pigmented skin lesions. Nevertheless, to our knowledge, the dermoscopic features of the pigmented lesions in LHS have not been described previously. Herein, we report a case of LHS together with its dermoscopic features.

Observations: The clinical examination revealed macular hyperpigmentation on the oral and genital mucosa, conjunctiva, and palmoplantar region together with longitudinal melanonychia. Dermoscopic examination of mucosal lesions on the patient’s lips and vulva revealed a parallel pattern. Longitudinal homogeneous pigmentation was observed on the toenails. The pigmented macules on the palms and the sole showed a parallel furrow pattern. A skin biopsy sample taken from the labial lesion was compatible with a diagnosis of mucosal melanosis.

Conclusions: By means of this case report, the dermoscopic features of the pigmented lesions in LHS are described for the first time, which facilitates diagnosis with a noninvasive technique. Future reports highlighting the dermoscopic features of this syndrome may simplify the diagnosis of LHS, which is thought to be underdiagnosed.

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Augier-Hunziker Syndrome (LHS) is a rare, acquired disorder characterized by benign macular hyperpigmentation of the oral and genital mucosa, which is associated with longitudinal melanonychia in 50% to 60% of cases.1 Dermoscopic examination should be indicated for patients with LHS who have several pigmented lesions on the mucosa, nails, or skin. However, to our knowledge, this is the first description of the dermoscopic features of the lesions in LHS.

Report of a Case

A 24-year-old female patient was referred to our dermoscopy unit because she had bluish-brown maculae on her lips (Figure 1A) and gingiva and pigmented papillae on her tongue (Figure 1B). In addition, a longitudinal hyperpigmented macule on the vulva extending from the labia minora to the labia majora (Figure 1C), an ill-defined hyperpigmented conjunctival macule (Figure 1D), 3 small pigmented macules on the palms and 1 on the sole, and linear melanonychia on the toenails were also observed. She had no medical history of receiving medication for, or family history of, pigmentation disorder or intestinal polyposis. Findings from the laboratory tests were within reference range, and the assessment of her gastrointestinal system included radiological examinations and a colonoscopy, the findings from which were also within reference range. Findings from the histological examination of the lesion on her lip revealed hyperpigmentation of basal keratinocytes. There were also perivascular melanin-laden macrophages in the papillary dermis (Figure 2A). Masson-Fontana staining was positive for melanin (Figure 2B), but staining with Prussian blue was negative for hemosiderin. Melanocytic proliferation was not detected immunohistochemically with S100 antibody. According to these findings, a diagnosis of LHS was made.
Dermoscopic examination of mucosal lesions on the patient’s lips and vulva revealed parallel patterns. On the lips, linear, streaklike brown pigmentation caused by the skin furrows and reliefs was associated with multiple brown dots of different sizes distributed regularly throughout the lesion (Figure 3A). The parallel pattern seen on the vulva was partially linear and partially curvilinear, with light- to dark-brown streaks following the cutaneous profile (Figure 3B). Homogeneous, brownish, regular bandlike pigmentation with indistinct borders were seen on 4 toenails (Figure 3C). The pigmented macules on the palms and on the sole showed a parallel furrow pattern (Figure 4).

In the case reported described herein, a parallel pattern was observed on the patient’s lips and vulva. On the lips, it was accompanied by regularly distributed multiple brown dots without any other dermoscopic variable suggestive of a malignancy. To our knowledge, the presence of multiple dots has not been reported in the limited number of articles published on dermoscopic features of mucosal melanosis.2,3 The parallel pattern with multiple brown dots correlated histopathologically with melanin pigmentation of basal keratinocytes and dermal melanophages, and these dots would be an expected finding when a patient has a superficial pigmentary incontinence. On the vulva, the pigmentation observed on dermoscopy was seen as partially linear and partially curvilinear brown streaks following the cutaneous profile, and this observation was concordant with the characteristic features of benign genital melanosis as defined by Soyer et al.4

The longitudinal melanonychia in our patient revealed homogeneous brownish regular bands with indistinct borders, and the pigmented macules on the palms and the sole revealed a parallel furrow pattern on dermoscopy.

By means of this case, the dermoscopic features of LHS are described for the first time. Future reports will improve our perception of this rarely seen entity.

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REFERENCES


Correction

Error in Author Order. In the Evidence-Based Dermatology Study by Helfrich et al, published in the March issue of the ARCHIVES (2007;143:397-402), there was an error in the author order in the byline. The names should have appeared as follows: Yolanda R. Helfrich, MD; Le Yu, MD; Abena Olori, MD; Ted A. Hamilton, MS; Jennifer Lambert, MS; Anya King, MPH; John J. Voorhees, MD; Sewon Kang, MD. The online version of this article was corrected on March 15, 2007.