

Large congenital nevus spilus— improved follow-up through the use of in vivo reflectance confocal microscopy

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ABSTRACT **Background:** Nevus spilus (NS) is a potential precursor of melanoma; the vast majority of cases reported in the literature were histologically classified as superficial spreading melanoma. **Objective:** To demonstrate the diagnostic value of reflectance confocal microscopy (RCM) in this subtype of congenital nevi. **Methods:** We report a case of a large congenital NS with equivocal clinical and dermoscopic findings in which RCM was applied for diagnosis and follow-up. **Results:** There was a good correlation of RCM with histopathology and a lack of dynamic changes during follow-up. **Conclusion:** Our observations indicate that RCM, as a non-invasive tool, can be useful for diagnosis and follow-up of clinically and dermoscopically equivocal NS.

Introduction

Nevus spilus (NS) is a relatively common cutaneous lesion that is seen in 2-3% of the population. It is characterized by numerous small, darkly pigmented macules or papules on a tan background pigmentation and is also referred to as “speckled lentiginous nevus,” “nevus on nevus” or “spotty

nevus” [1] The background tan patch is usually present at birth and NS is therefore considered a subtype of congenital melanocytic nevus [2]. The typical dimension of NS ranges from 2 to 10 cm, but some NS may cover extensive skin areas with regional or zosteriform distribution. Large NS involve especially the trunk as well as the upper and lower extremities. Histologically, the background hyperpigmenta-



Figure 1. Clinical image. Multiple light to dark brown macules and papules are seen on the tan background pigmentation. One newly developed dark brown macule that was excised for histopathologic examination is encircled; another one that was followed up with dermoscopy and RCM is marked by an arrow. [Copyright: ©2013 Proding et al.]

tion has been described as having the features of a lentigo simplex or a café-au-lait macule. The darker speckles of NS are classically reported as superimposed junctional or compound nevi, but intradermal, Spitz, and blue nevi may occasionally occur [3].

NS are potential precursors of melanoma, which underlines the need for a precise examination and a close lifelong follow-up to exclude malignancy [4]. The risk that melanoma develops within a NS is thought to be proportional to the size of the lesion and has been reported to be higher for macular than for papular NS [5]. More than 20 cases of melanoma arising within a NS have been reported in the literature; over 90% of these cases were histologically classified as superficial spreading melanoma.

With the following case report, we will show for the first time that in vivo reflectance confocal microscopy (RCM), as a convenient non-invasive method for skin investigation, is useful in diagnosis and follow-up of NS.

Case report

A 15-year-old girl was referred to our outpatient department because of a giant congenital NS covering her right scapular region and upper aspect of her right arm. Recently, the

patient had noticed the development of multiple darkly pigmented spots within the nevus.

Skin examination revealed multiple light to dark brown macules and papules within the NS, measuring 0.2 to 3 cm in diameter (Figure 1). Dermoscopy of the newly developed lesions showed a brown to black homogeneous or homogenous-reticular pigment pattern with focal gray-blue areas, suggestive of regression (Figures 2A and 3A, C). In addition, many of the lesions displayed irregularity of the pigment network and irregular dots, altogether raising the suspicion of malignancy.

In vivo RCM (Vivascope 1500, MAVIG GmbH, Munich, Germany) of a representative lesion showed a typical honeycomb pattern at epidermal layers and a regular ringed pattern at the dermoepidermal junction; uniformly distributed dermal papillae surrounded by a rim of bright monomorphous cells, correlating to pigmented keratinocytes and melanocytes, were seen (Figure 2B, C). Additionally, small junctional aggregates of melanocytes protruding into the dermal papillae were found. At upper dermis, there were focal aggregates of non-nucleated plump bright cells compatible with melanophages. These results correlated well with the histopathologic findings, which were consistent with the diagnosis of “junctional melanocytic nevus without atypia” (Figure 2D).

The remaining newly developed lesions within the NS showed similar confocal findings suggestive of benign nevi. These lesions were monitored with digital dermoscopy and RCM and did not show any changes at a three-month follow-up visit (Figure 3).

Discussion

The progressive appearance of macules and papules within a NS, as observed in our case, is not unusual. Although the majority of these newly arrived lesions are benign, a risk for melanoma exists; therefore, a close follow-up of large NS is essential since their dimensions rarely permit a complete surgical excision.

RCM, as a new method capable of producing real-time in vivo sections of the skin at a nearly histologic resolution, permits cytological analysis to complement dermoscopy and histopathology for melanoma detection. Multiple studies have already demonstrated excellent correlation between confocal microscopy and conventional histology for melanocytic lesions [6]. Despite its maximum in vivo imaging depth of approximately 250 μm , RCM reaches particularly good diagnostic accuracy in diagnosing melanoma at the dermoepidermal junction. Therefore, RCM should be adequate for the detection of melanoma within NS, since most cases reported were superficial spreading melanomas. In addition,

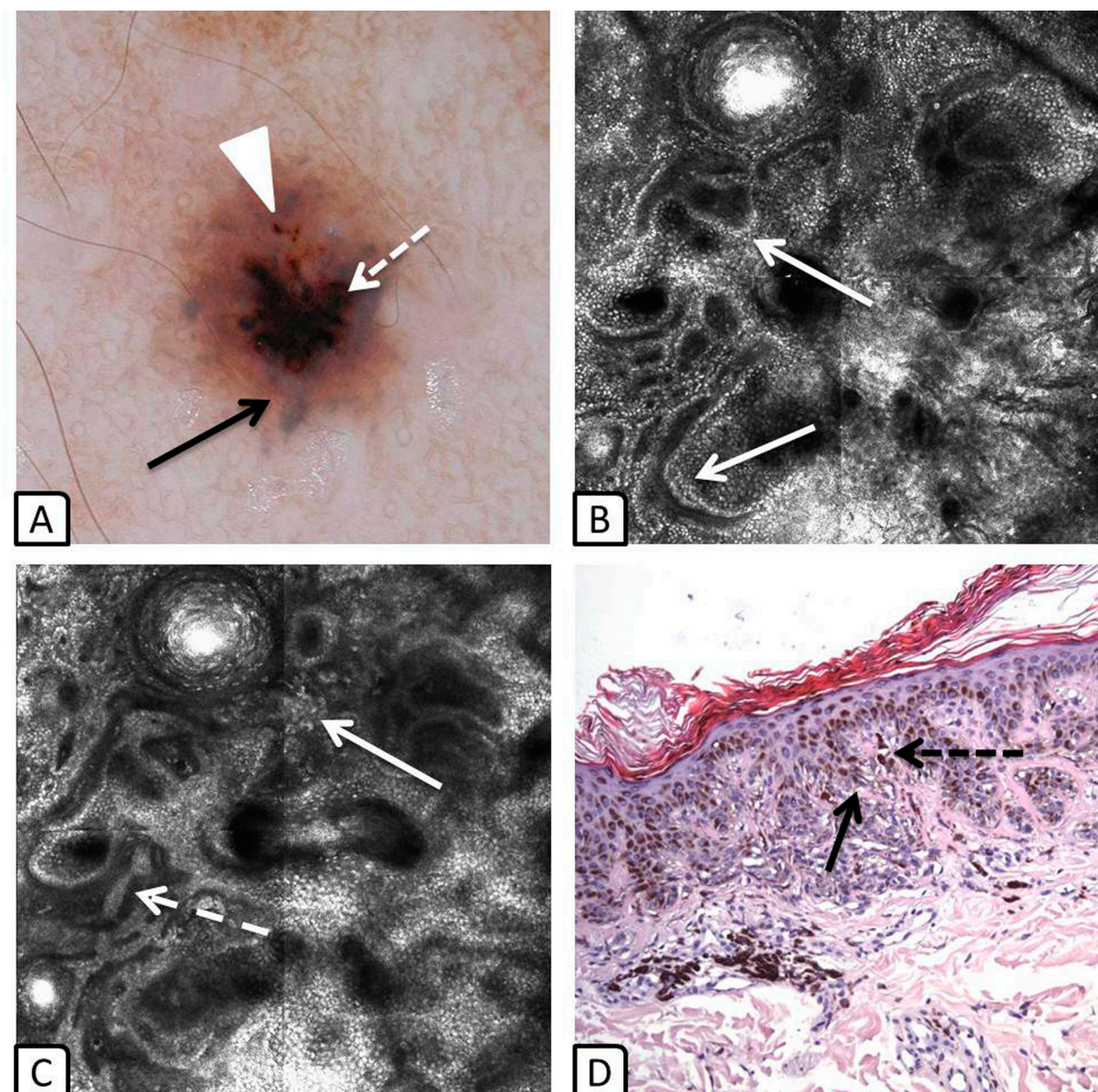


Figure 2. (A) Dermoscopic picture (non-polarized) of the newly developed 2 mm macular lesion encircled in Figure 1. A homogeneous brown pigment pattern with focal gray-blue areas (arrow), black dots (arrowhead) and a central, irregularly outlined black blotch (dashed arrow) are observed. (B) RCM image at the level of the dermoepidermal junction (high magnification; 1 x 1 mm field-of-view). Dermal papillae are demarcated by a rim of bright monomorphic cells (arrows) corresponding to pigmented keratinocytes and melanocytes. (C) Deeper RCM section at the level of the dermo-epidermal junction (high magnification; 1 x 1 mm field-of-view). Focal aggregates of non-nucleated plump bright cells, compatible with melanophages (arrow) and regular compact cell aggregates, correlating to melanocytic nests (dashed arrow) are visualized within the papillary dermis. (D) Histopathology reveals features of a junctional nevus without cellular atypia; melanocytes are seen in single units and in small clusters along the dermoepidermal junction (arrow). Focal aggregates of melanophages are detected within the papillary dermis (dashed arrow). (H&E, magnification 10x). [Copyright: ©2013 Prodingler et al.]

this technique allows completely safe and painless repetitive application and observation of dynamic changes in situ over time and consequently may reduce unnecessary excisions.

The good correlation of RCM with histopathology in our case indicates that RCM, as a non-invasive tool, can be used as an auxiliary for differentiation of nevi from melanoma in clinically and dermoscopically equivocal NS.

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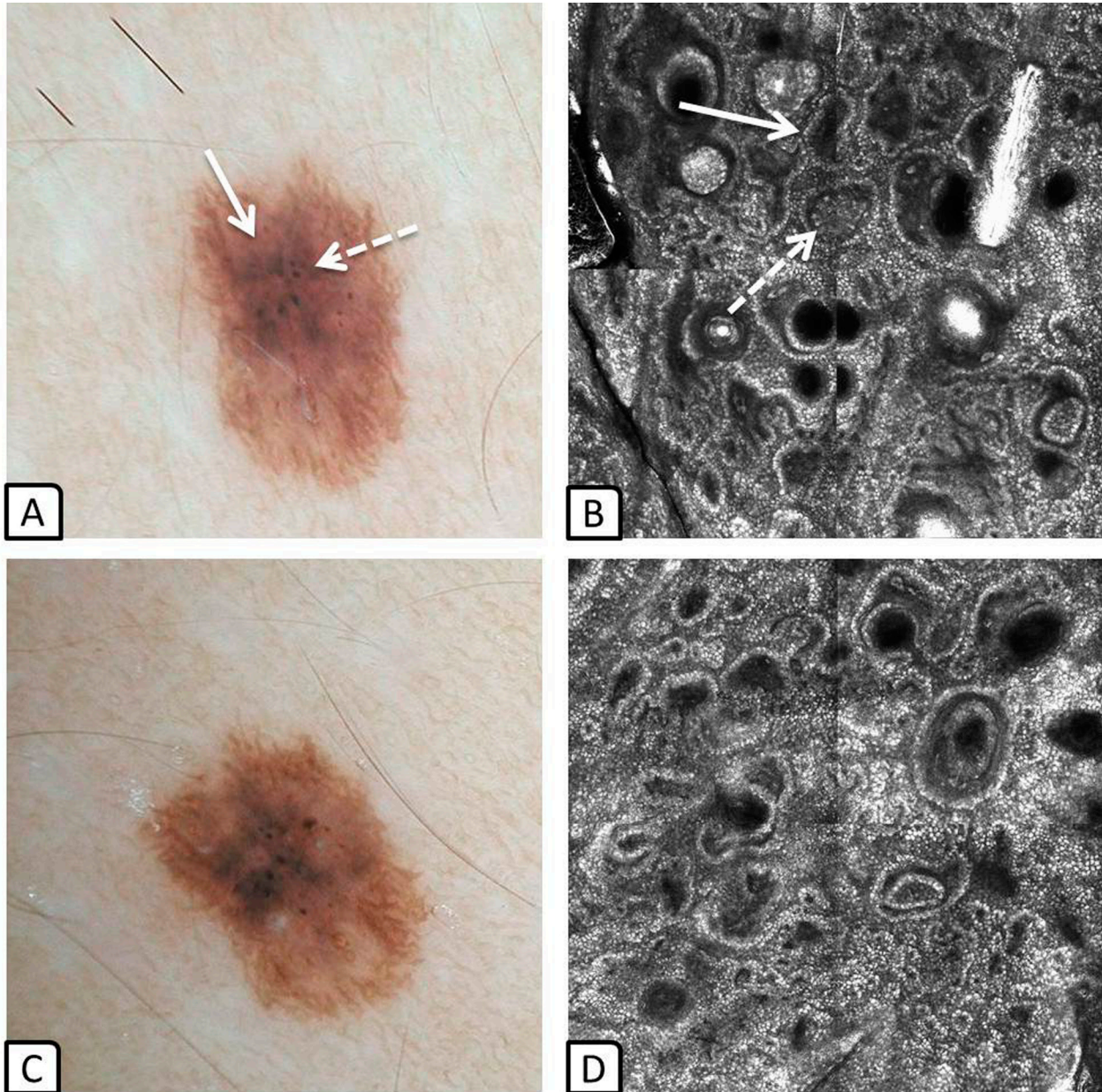


Figure 3. (A) Polarized dermoscopy of a newly developed nevus that was followed up with dermoscopy and RCM (see arrow in Figure1). A homogenous-reticular pattern with central gray hue (arrow) and focal black dots (dashed arrow) is seen. (B) RCM at the level of the dermoepidermal junction (high magnification; 1 x 1 mm field-of-view). Edged dermal papillae (arrow) surrounded by a rim of monomorphous bright cells (pigmented basal keratinocytes and melanocytes) as well as compact cell aggregates within dermal papillae (melanocytic nests; dashed arrow) are observed. (C) Non-polarized dermoscopy of the nevus shown in Figure 3A shows no major morphological changes at the three-month follow up visit. (D) Three-month RCM follow-up image at the dermoepidermal junction of the nevus shown in Figure 3B (1 x 1 mm field-of-view). A regular ringed pattern composed by edged dermal papillae is again observed. [Copyright: ©2013 Proding et al.]

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