Videodermatoscopic Approach to Porokeratosis of Mibelli: A Useful Tool for the Diagnosis

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Sir,
Dermatoscopy was introduced in dermatology to support the clinical diagnosis of pigmented skin lesions, and after a long period of development it is now commonly used in dermatological practice (1–5). Porokeratosis of Mibelli (PM), a dermatosis characterized by an alteration of epidermal keratinisation, has a wide variety of morphological clinical presentations: classic PM, linear PM and punctate PM (6). It is characterized by single or by a few papules and plaques, varying in size, with elevated borders and a slightly evident longitudinal furrow, usually located in the extremities; their central portion is generally hypopigmented, hairless and atrophic. The histopathological hallmark is the presence at the borders of the lesions of the cornoid lamella, formed by parakeratotic cells filled in invaginations of epidermis through the orthokeratotic stratum corneum (6, 7). We performed a dermatoscopic examination in 10 patients with classic PM in order to point out dermatoscopic features enabling a non-invasive diagnosis, always taking into account the clinico-pathological correlation.

MATERIALS AND METHODS

We enrolled 10 patients (7 women and 3 men, aged between 15 and 65, average age 47 years), presenting an overall number of 31 plaques (1 to 5 lesions each), with a clinically suspected diagnosis of classic PM. PM plaques were located mostly in the upper and lower extremities; 8 patients had multiple lesions located on extremities and trunk; single lesions were observed in 2 patients.

The examination was performed using a computerized optical probe videomicroscope (Videocap 200, DS Medi-Group); lesions were covered with a drop of immersion oil and observed using lenses of $\times 20$ and $\times 50$ magnification, provided with a glass slide, and a $\times 200$ lens (the actual sizes of the sites thus photographed were 196.4 mm^2, 25 mm^2 and 1.8 mm^2, respectively). Clinical and dermatoscopic photographs of the lesions were taken and four independent, but experienced, observers completed evaluation of the dermatoscopic clues in a blind manner. Histopathological examination confirmed the clinical suspicion of PM in the patients, all of whom were fully aware of the nature of the study and had given their informed consent.

RESULTS AND DISCUSSION

In the clinical images of all the lesions ($\times 20$ and $\times 50$ lenses), a raised border with an inland longitudinal furrow was clearly visible. The four observers agreed in pointing out two further clues for the diagnosis of PM:

1. In 31 lesions (100%), dark-brown close globules and dots circumscribing a central hypopigmented scar-like area were present. In 11 lesions (36%), these globules/dots joined to form a continuous line (Fig. 1).

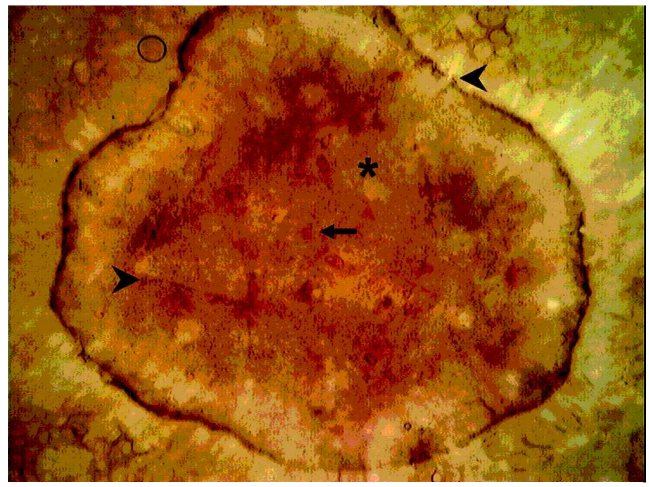

Fig. 1. Dark-brown close globules and dots (arrowhead) circumscribing a central hypopigmented scar-like area (asterisk) with multiple scattered brown globules and dots (arrowhead). Red dots (arrow) in the centre of the lesions ($\times 50$ lens).

2. In the central area of 28 lesions (90%), multiple scattered brown globules and dots were present. In 13 lesions (42%), red dots were present, too (Fig. 1).

The brown dots all over the lesion histologically, probably correspond to a post-inflammatory accumulation of melanophages at the dermal-epidermal junction and in the papillary dermis, whereas the scar-like area corresponds to fibrosis degeneration of the dermis in the central portion of the lesions.

The $\times 200$ lens is not commonly used in dermatoscopic examination, but it proved to be effective in the diagnosis of PM, in that white scales were observed arising from follicular ostia in 12 lesions (39%); histologically, these scales probably correspond to cornoid lamella emerging from involved hair follicles.

The epidermis in the central area of PM lesions is usually fairly atrophic. We believe that the thin epidermic layer allowed clear sight ($\times 200$ lens) of the dermal blood vessels below, which by contrast were not so clearly visible in the healthy surrounding skin. They may correspond to the red dots observed with the $\times 50$ lens, so they could be considered as "dotted vessels" (8).

The younger PM lesions showed slightly evident scar-like features and numerous red dots in the central area, whereas in the older lesions the scar-like features were prominent and the red dots were not present. This is probably related to a prevalence of the epidermal atrophy in the central area of younger lesions combined with a progressive dermal fibrosis in the older ones.

Other pigmented skin lesions presenting dermatoscopic features resembling that of PM are Clark naevus, Spitz naevus, melanoma, dermatofibroma and basal cell carcinomas. Clark

naevi and Spitz naevi can present a central hypopigmented area with multiple brown dots, but this area lacks the scar-like feature of PM and, instead, is surrounded by a pigment network.

Dots and globules are considered a major dermatoscopic criterion for the diagnosis of melanocytic lesions (9), and dotted vessels are commonly believed suggestive of melanoma (8). Their presence in PM must be considered an exception to the rule; however, other cases are present in the literature in which the dermatoscopic criterion for the diagnosis of melanocytic lesions was not respected (10). It is possible that by extending the use of dermatoscopy to non-melanocytic lesions, a revision of dermatoscopic patterns will be required.

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Ultraviolet A Sunbed Used for the Treatment of Scleroderma

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Sir,

Ultraviolet A has been used in recent years in the treatment of localized and systemic scleroderma with good results (1). In most studies, UVA-I (340–400 nm) has been utilized (2–6), but there are some investigations showing that ordinary UVA alone or with psoralen is also effective (7–10). It has been shown that UVA increases collagenase in fibroblast cultures and in human skin, suggesting that this may be the basic mechanism by which UVA is beneficial in scleroderma (11, 12). Also, modulation of the immunosystem by UVA could contribute to the useful effects (13).

Since UVA-1 devices are relatively expensive, and not available in all dermatologic departments, there is a need to use other treatment modalities. Accordingly, a girl with extensive localized scleroderma was recently successfully treated with UVA from a sunbed. This treatment has now been used for other scleroderma patients with good results, prompting us to report our experience from an open study.

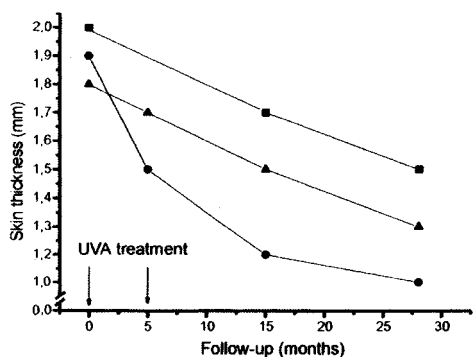

Fig. 1. Skin thickness of patient 1 was repeatedly measured by ultrasound before UVA treatment and for up to 26 months. The time between arrows is the period when altogether 60 UVA treatments were given by UVA sunbed. Note the interrupted scale on the y-axis (■ = shin; ● = forearm; ▲ = abdomen).

CASE REPORTS

Patient 1, a 12-year-old girl, had had gradually expanding generalized morphea for a year. At the first visit to a dermatologist, she had tightening and thickening of the skin on her arms and legs and over most of her body. She had difficulty extending her arms owing to the skin changes, and she had therefore refused to take part in gymnastics at school. A skin biopsy was taken and skin thickness was measured and recorded by Dermascan-A from abdominal skin, upper and lower arm, leg and back. Histology revealed markedly thickened dermis with numerous eccrine ducts surrounded by thick collagen bundles.

When the diagnosis was established, UVA treatment with ordinary solaria was started. The girl was treated using a sunbed (Solana computer sunbed, with Philips Performance lamps 100 W). Lamp output was 18–20 mW/cm^2, and mostly within 340–400 nm. The patient was treated three times a week for a maximum of 20 min at a time. She was treated 60 times, and the total UVA dose was estimated to be about 1100 J/cm^2.