

Case Report/Case Series

A Digital Dermoscopy Follow-up Illustration and a Histopathologic Correlation for Angulated Lines in Extrafacial Lentigo Maligna

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IMPORTANCE The diagnosis of extrafacial lentigo maligna can be a challenge because the dermoscopic features are not well described. These features can share some of the criteria of superficial spreading melanoma and some of facial lentigo maligna; however, these features are often absent. A new dermoscopic pattern has recently been described as forming angulated lines.

OBSERVATIONS We report 4 cases of extrafacial lentiginous lesions in 3 patients in whom straight angulated lines have been observed as the only key for diagnosis. This is the first time, to our knowledge, that these lines were seen in the follow-up by digital dermoscopy during a lapse time of a few months. This morphologic change led to the excision of the lesion. The pathologic diagnosis of lentigo maligna was established in each case. Moreover, a histologic correlation of these straight angulated lines is documented.

CONCLUSIONS AND RELEVANCE Angulated lines could be considered as a new warning sign during the follow-up of extrafacial lentigo. More studies are required to establish the sensitivity and specificity of this newly described dermoscopic feature.

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Lentigo maligna (LM) constitutes the noninvasive phase of lentigo maligna melanoma. Although LM is still in situ, it should be diagnosed and treated given the risk of progression to invasive melanoma. Lentigo maligna typically occurs on chronically sun-exposed skin, such as that of the head and neck, and less frequently on the extremities of elderly individuals. Extrafacial LM (EFLM) was first described by Sir John Hutchinson in 1890¹ and is therefore called Hutchinson melanoma. These EFLMs account for 17.5% of all cases of LM according to a 10-year analysis of the Scottish Melanoma Group database.²

The dermoscopic features of LMs of the head and neck are well known and are related to the facial skin anatomy characterized by the absence of rete ridges, multiple pilosebaceous units, and photoinduced elastosis. The dermoscopic features of LM were defined in 2000 by Schiffner et al.³ The progressive invasion by malignant cells can be observed in dermoscopy by the appearance of asymmetrical pigmented follicular openings, a perifollicular annular-granular pattern, pigmented rhomboidal structures, and obliterated hair follicles. Slutsky and Marghoob⁴ described a zigzag pattern of angulated, linear pigmented lines on facial LMs. Pralong et al⁵ described 4 additional original criteria: increased vascular network, targetlike images, red rhomboidal structures, and darkening at dermoscopic examination.

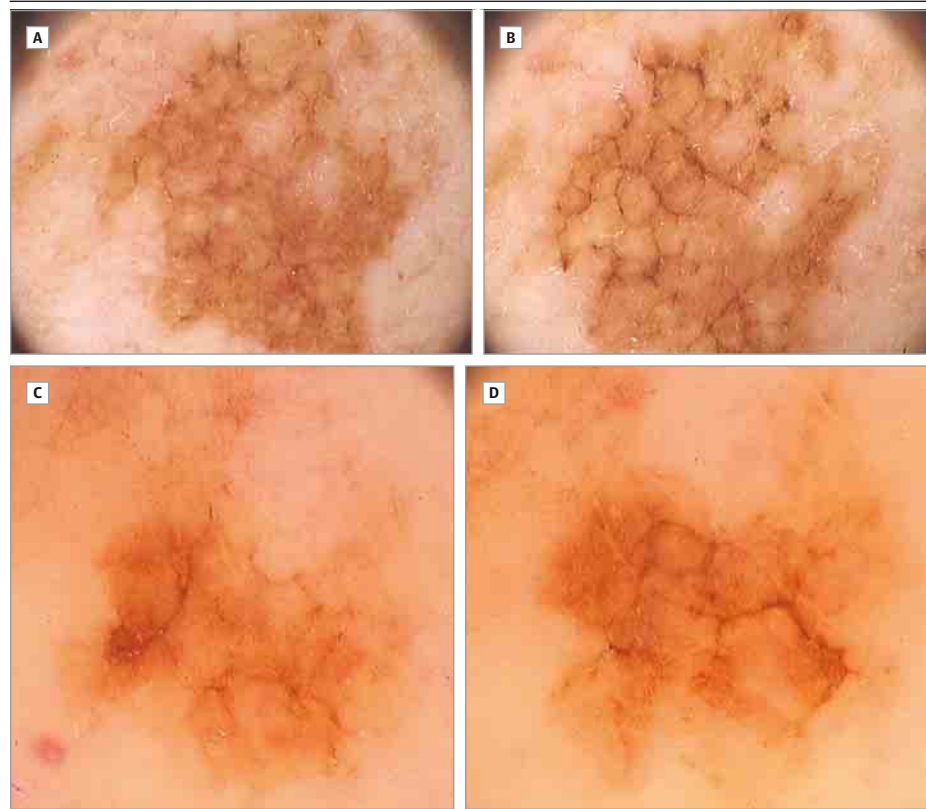
Less is known regarding the dermoscopic features of EFLM. In 2013, Lau et al⁶ performed a study of 3 cases featuring the dermoscopy of extrafacial LMs. They proposed that they could share a combination of dermoscopic features of LM (pigmented rhomboidal structures and asymmetrical perifollicular pigmentation) and superficial spreading melanoma in situ (irregular dots and streaks and an irregular pigmented network).

Two new EFLM criteria have been described by Keir^{7,8}: a lentigo-like pigment pattern lacking a lentigo-like border and complete or incomplete large polygonal shapes defined by darker gray or brown straight lines or formed by the straight edge of a junction between lighter and darker areas of the lesion.

Jaimes et al⁹ recently proposed the new term *angulated lines* to integrate all of the different linear dermoscopic patterns previously described, such as the rhomboidal structures by Schiffner et al³ and the zigzag pattern by Slutsky and Marghoob,⁴ both features described on facial LMs, and the large polygons described by Keir⁷ on EFLM. However, even though they may look geometrically similar superficially, it is not proven whether they share the same biological or pathologic process.⁹

We report 3 cases by digital follow-up in which straight angulated lines were seen on extrafacial pigmented lesions and

Figure 1. Lentigo Maligna (LM) of the Back and Left Lower Leg in Patient 1



A, LM of the back. B, LM of the back: evolution after 10 months. C, LM of the left lower leg. D, LM of the left lower leg: evolution after 5 months. In both cases, straight angulated lines are discrete on the initial pictures. Additional lines then appear and grow darker and more easily seen after several months of follow-up. Pictures were taken with a FotoFinder dermoscope (Teachscreen Software GmbH).

how the occurrence of only these lines over time allowed the diagnosis of LM. In addition, we attempt to explain how this phenomenon can be histologically correlated.

Report of Cases

Patient 1 was a woman in her 50s who was diagnosed in her teens as having xeroderma pigmentosum. The patient had been followed up in our department for 11 years via digital dermoscopy. She presented with severe actinic skin damage with a history of multiple carcinomas and 82 melanomas of different subtypes. We observed over time the appearance of straight angulated lines on several pigmented lesions, which were excised because of this morphologic change. All of the lesions were diagnosed as LM (Figure 1 and Figure 2). On the lesion shown in Figure 2, the section of the surgical sample was performed perpendicularly to the lines. At microscopic examination, an atrophic epidermis and a proliferation of isolated atypical melanocytes along the dermoepidermal junction were observed. The accumulation of melanophages in the superficial dermis was seen underneath the clusters of melanocytes. A second nest of melanocytes was found approximately 1 mm from the first where the section intersects the second line; underlying melanophages were also observed. An inflammatory infiltration was also present. The diagnosis of LM was made.

Patient 2 was a man in his 70s with a personal history of 3 superficial spreading melanomas and several squamous cell

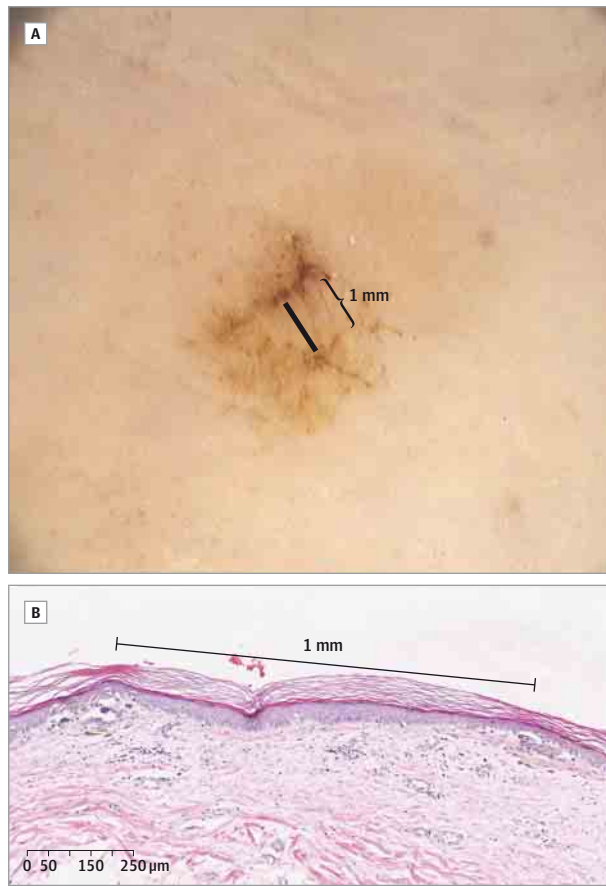
and basal cell carcinomas. A pigmented lesion located on the patient's back drew our attention because of the occurrence of straight angulated lines. The lesion was excised, and the diagnosis of LM was confirmed (Figure 3).

Patient 3 was a woman in her 80s who had been followed up via digital dermoscopy for the past 2 years because of a large pigmented lesion on her lower leg. The lesion was stable until slight modifications displaying straight angulated lines appeared (Figure 4). Histologic examination confirmed the diagnosis of LM.

Discussion

Little is known about specific dermoscopic features of EFLM. Keir^{7,8} described a lentigo-like pigment pattern that lacked a lentigo-like border and complete or incomplete large polygonal shapes defined by darker gray or brown straight lines or formed by the straight edge of a junction between lighter and darker areas of the lesion. The polygonal shapes appear to be much larger than the rhomboidal structures seen in facial LM and are described to be rhomboidal, pentagonal, or hexagonal and in some cases centered on follicular openings. However, because the angulated lines do not always form completed polygons, we believe the term *angulated lines* recently proposed by Jaimes et al⁹ seems to be the most appropriated. Angulated lines must then be differentiated from the zigzag pattern described by Slutsky and

Figure 2. Lentigo Maligna of the Right Lower Leg in Patient 1



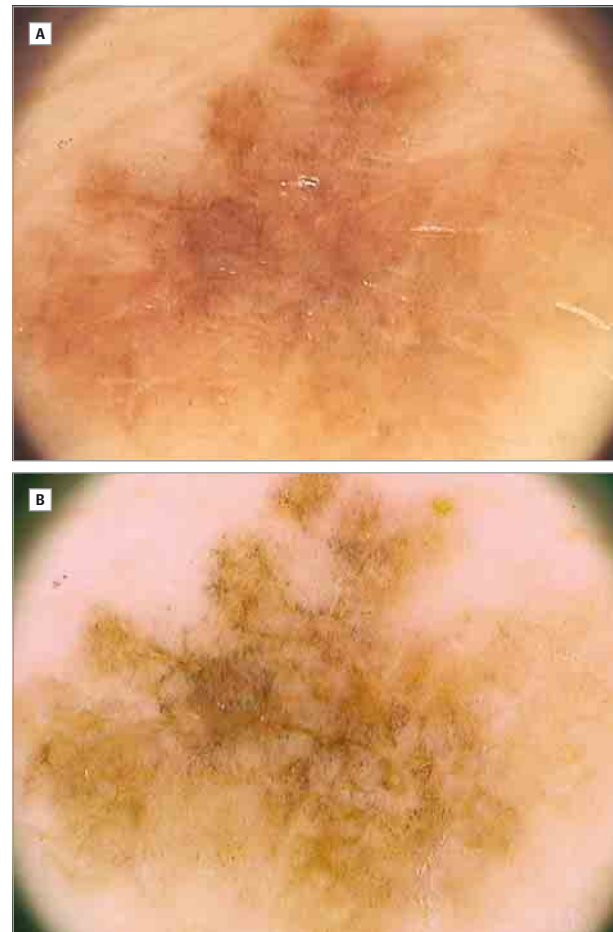
A, A distance of 1 mm between the 2 lines was found dermoscopically and histologically. B, A proliferation of isolated atypical melanocytes focally clustered in 2 nests is observed at the dermoepidermal junction and is associated with an underlying accumulation of melanophages in the superficial dermis directly underneath those nests (hematoxylin-eosin, original magnification $\times 10$).

Marghoob,⁴ which, according to the authors, probably represents incompletely formed rhomboidal structures related to the facial skin anatomy.

Also known as Hutchinsonian melanotic freckles, LM occurs histologically in skin with severe actinic damage, manifested by epidermal atrophy and solar elastosis. They are characterized by a proliferation of atypical melanocytes along the dermoepidermal junction and often down the walls of hair follicles and sweat ducts.^{10,11} This proliferation is confined to the basal layer and is formed by a linear accumulation of single cells and/or irregular nests and a slight pagetoid invasion of the epidermis.^{10,12} In addition to solar elastosis, the papillary dermis contains a lymphocytic infiltration, fibroplasias, and telangiectasia.¹⁰ Regression phenomena (fibrosis, hypervascularity, melanophages, and lymphocytic infiltration) may be seen and should prompt a careful search for invasion by atypical melanocytes.^{10,11} Most of these features are observed in all of our cases.

Figure 2 clearly shows an atypical proliferation of melanocytes, focally organized in nests underneath which accu-

Figure 3. Lentigo Maligna (LM) of the Back in Patient 2



A, LM of the back. B, LM of the back: straight angulated lines are seen appearing after 3 months of evolution. Pictures were taken with a FotoFinder dermoscope (Teachscreen Software GmbH).

mulations of melanophages were found. The localization of these 2 associations exactly matched the perpendicular section of the 2 lines, which had been observed dermoscopically. Indeed, the distance between the 2 lines (1 mm) in dermoscopy was also found during pathologic analysis.

In accordance with the report by Keir,⁸ we could clearly see a focal accumulation of melanophages in the superficial dermis. In addition, nests of atypical melanocytes were found right above the melanophages at the dermoepidermal junction. No relation with the hair follicle was observed because it is described for the rhomboidal structures of the facial LMs, which are related to a proliferation of melanoma cells around the hair follicles.³

Conclusions

Angulated lines are a pattern observed in LMs, but the physiopathologic mechanisms appear to be different in EFLM than in facial LM. We describe 3 patients in whom angulated lines

Figure 4. Lentigo Maligna (LM) of the Leg in Patient 3



A, LM of the leg. B, LM of the leg: evolution after 12 months; straight angulated lines are becoming darker and more visible. Pictures were taken with a FotoFinder dermoscope (Teachscreen Software GmbH).

were observed, which were the only criteria in support of their malignant lesion. These angulated lines could correspond to a host reaction against the lesion and therefore be a warning sign that the lesion is becoming malignant. The emergence of these lines in dermoscopy could become a new diagnostic ele-

ment in the follow-up of extrafacial lentiginous lesions. Their appearance or presence should alert the dermatologist. If the angulated lines change over time, they should be considered as a criterion for excision of the lesion. This new postulate should be confirmed in future studies.

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