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Dermatoscopic features of extrafacial lentigo maligna

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Summary

The dermatoscopic features of classic lentigo maligna (LM) are well described; however, there is little literature available on extrafacial LM, which is a less frequently reported condition. The skin architecture is especially rich in adnexae on sun-exposed areas such as the face, relative to other parts of the body, thus it is possible that the dermatoscopic findings of extrafacial LM will differ from the usual findings of LM. We carried out a dermatoscopic study on three patients with extrafacial LM. The dermatoscopic patterns reflected the underlying histological features of the disease, with increased melanocytes seen along the rete ridges and around follicular ostia, which result in a unique pigment network architecture.

Lentigo maligna (LM) is a distinct form of melanoma *in situ*, which is characterised by an increased number of histologically atypical melanocytes situated along the dermoepidermal junction. If untreated, the condition can progress to invasive LM melanoma (LMM).[1] LM is more common in fair-skinned and older people. Long-term sun exposure is the main risk factor, which explains why most LM are seen on the head and neck region. It has been reported that LM represents a prolonged preinvasive phase, and when associated with LMM, the prognosis is better than with other forms of invasive melanoma. Extrafacial LM, occurring at sites distant from the face, is rarer, and is often confused with superficial spreading melanoma (SSM) *in situ*. The existing literature on extrafacial LM is scarce. One study analysing 71 cases of LM from the Scottish Melanoma Group database over 10 years found that extrafacial LMM accounted for 17.5% of all cases of LMM.[1] In that study, the mean age of diagnosis in patients with extrafacial LMM was significantly younger than that of patients with head and neck LMM (63 years vs. 72 years),[1] and extrafacial LMM was seen predominantly on the trunk in men but on the limbs in women.[1] The tumours of extrafacial LMM were also thinner than LMM of the head and neck.[1]

In this study, we investigated the dermatoscopic features of three patients with extrafacial LM, and compared these with the recognized dermatoscopic features of LM of the head and neck.

Report

Patient 1, a 61-year-old woman, presented with a long-standing pigmented skin lesion on the dorsum of her right wrist, which had recently increased in size and changed in appearance (Fig. 1a). Clinically, the differential diagnosis included dysplastic naevus, SSM *in situ* or invasive SSM. The lesion was excised. On histological examination, there were signs of sun-damaged tissue with lentiginous and nested proliferation of melanocytes with severe nuclear atypia. There was little pagetoid spread, but atypical melanocytes were seen extending down the follicular epithelium, with no involvement of the dermis, findings that were consistent with LM. The dermatoscopic features of the lesion were of irregular reticulate pigmentation with dots and globules (Fig. 1b), and rhomboidal structures and asymmetrical perifollicular pigmentation (Fig. 1c) were also visible.

Patient 2 was a 75-year-old woman, who presented with a 4-month history of a new pigmented skin lesion that had appeared on her right upper arm

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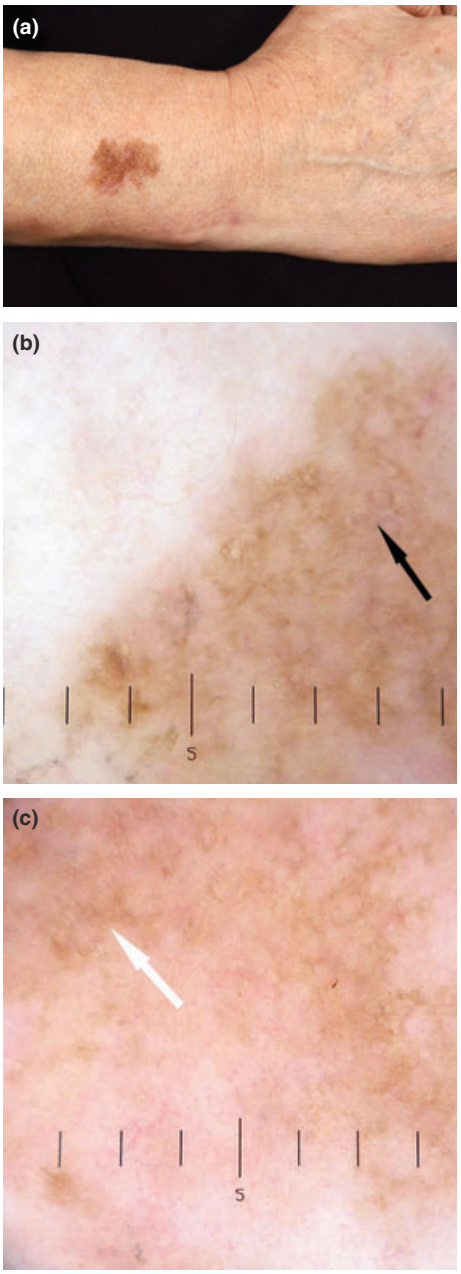


Figure 1 Patient 1: (a) lesion on right wrist; (b) rhomboidal structures (arrow) and (c) asymmetrical perifollicular pigmentation (arrow).

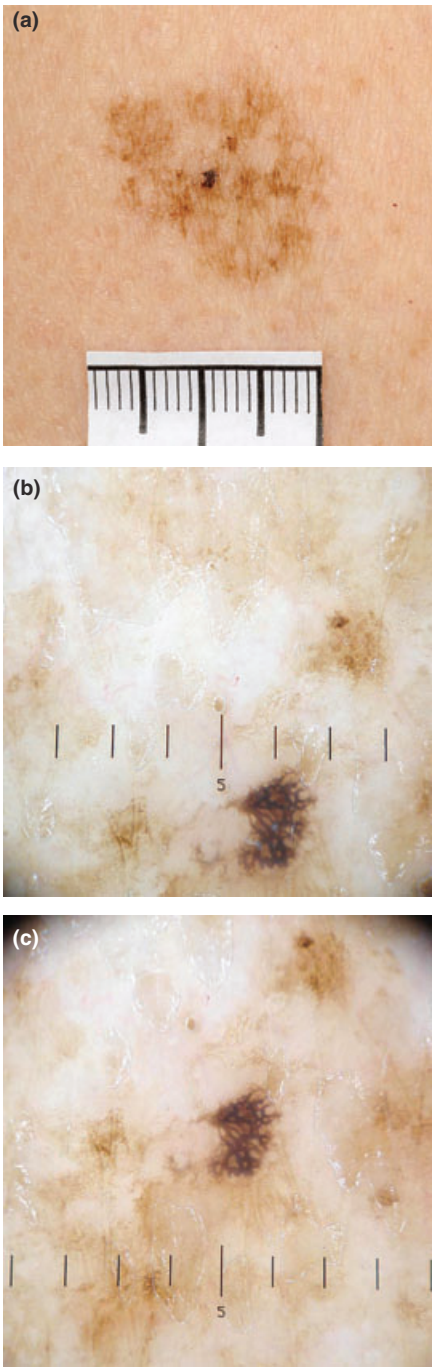


Figure 2 Patient 2: (a) lesion on right upper arm; (b) background solar lentigo-like pigmentation with dots and blotches, and (c) a heavily pigmented area with blotches, streaks and a dot pattern.

(Fig. 2a). She had previously lived for several years in the Far East. Clinically, the lesion appeared to be an atypical solar lentigo, but a biopsy was taken to exclude the differential diagnosis of extrafacial LM. On histological examination, a lentiginous proliferation of melanocytes was seen along elongated epidermal rete ridges. The cells showed moderate nuclear atypia, and there was no evidence of an invasive component, consistent with LM. Dermatoscopy showed a background of solar lentigo-like pigmentation with dots and

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blotches (Fig. 2b). A heavily pigmented area with blotches, streaks and dots (Fig. 2c) was also seen, and this dark pigmentation made dermatoscopic examination difficult.

Patient 3, a 45-year-old woman, presented with a new pigmented lesion on her left shoulder (Fig. 3a). The clinical differential diagnosis included SSM or extrafacial LM. The lesion was excised, and the histological findings confirmed LM. Using dermatoscopy, irregular dots and globules with early rhomboid-like structures were seen (Fig. 3b). An unusual double figure-of-eight configuration of irregular pigmentation, featuring dots and blotches and a fine reticulate pattern, was present (Fig. 3c).

Dermatoscopy is widely used as an adjunctive tool in assessing both pigmented and nonpigmented lesions. For pigmented lesions, a pigment network suggests a melanocytic origin. The dermatoscopic criteria of melanocytic lesions on the trunk and limbs mainly focuses on the rete ridge pattern at the junction of the epidermis and dermis. However, sun-exposed facial skin has flat rete ridges, devoid of network-derived features compared with other sites. The unique dermatoscopic features identified for LM include asymmetrical pigmented follicular openings, perifollicular slate-grey dots and granules, atypical blood vessels, linear pigmented lines forming rhomboidal structures,[2] and a zigzag pattern of angulated pigmented linear lines.[3] These features can be explained by the uneven descent of malignant cells within the hair follicles, giving rise to asymmetrical follicular openings, which progress to streaks and rhomboidal structures as the abnormal melanocytes proliferate and interweave around the follicles.[3] The slate-grey dots are representative of aggregates of melanin-loaded macrophages seen in the upper dermis. By contrast, the dermatoscopic features of SSM *in situ* are similar to those of invasive melanoma, and include a blue–white veil, grey–blue areas, black dots, brown globules, irregular extensions and branched streaks, irregular pigment network, pseudopods and depigmentation.[4]

To our knowledge, the dermatoscopic features of extrafacial LM have not been described previously. The differences in rete ridges and the concentration of hair follicles in facial skin compared with other body sites raises the question as to whether the dermatoscopic features of LM could be applied to extrafacial LM; from the three examples above, we suggest that this is so, as the dermatoscopic findings are similar to those in LM of the head and neck. These features, particularly the rhomboidal structures and asymmetrical perifollicular pigmentation were present in patients 1 and 3,

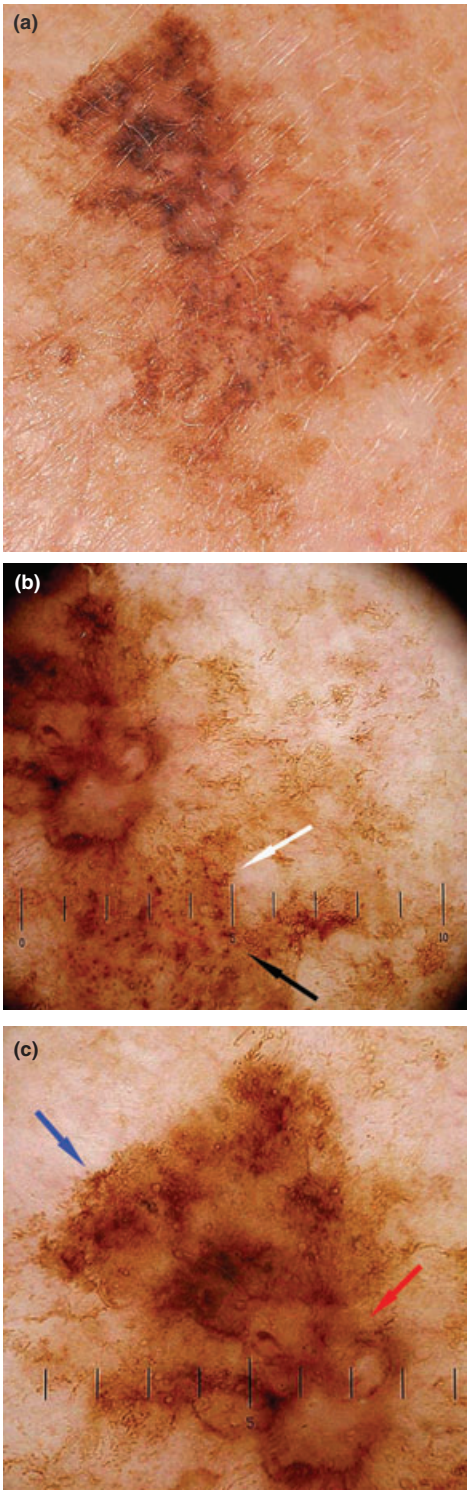


Figure 3 Patient 3: (a) lesion on the left shoulder; (b) irregular dots and globules (black arrow) and early rhomboid-like structures (white arrow); (c) unusual double figure-of-eight configuration of irregular pigmentation featuring dots and blotches (red arrow), and the presence of a fine reticulate background (blue arrow).

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but not in patient 2. The dermatoscopic features of SSM *in situ*, namely, irregular dots and streaks and an irregular pigment network, were seen in all three patients. Therefore, we propose that the dermatoscopic features of extrafacial LM can include a combination of features of LM and SSM *in situ*; however, the presence of rhomboidal structures and asymmetrical perifollicular pigmentation in extrafacial lesions makes the diagnosis of extrafacial LM more likely.

Histologically, LM is characterized by a linear and nested proliferation of atypical melanocytes along the dermoepidermal junction, and often down the walls of hair follicles and sweat ducts, in association with epidermal atrophy and solar elastosis.[5] One feature differentiating LMM from SSM is the absence of a pagetoid upward spread of melanocytes in LMM.[2]

There are biological differences between LM and SSM *in situ*. A previous immunohistochemical study showed that the melanocytes of SSM *in situ* have higher proliferative activity and higher rates of angiogenesis than those of LM.[6] These findings are consistent with the biological behaviour of these lesions, as the *in situ* phase of LM is longer than that of SSM.

In summary, extrafacial LM is rare, but should be included in the differential diagnosis of pigmented lesions on the trunk or limbs. Dermatoscopy may be a useful tool in distinguishing extrafacial LM from superficial spreading melanoma *in situ*, especially if there are features of rhomboidal structures and irregular perifollicular pigmentation. These dermatoscopic features correspond to the histological findings of atypical proliferation of melanocytes down hair follicles in extrafacial lentigo maligna. LM is a separate disease with specific clinical, dermatoscopic and histological features, which may help distinguish it from superficial spreading melanoma *in situ*.

Learning points

- The dermatoscopic features of facial LM include asymmetrical pigmented follicular openings, perifollicular slate-grey dots and granules, atypical blood vessels, linear pigmented lines forming rhomboidal structures, and a zigzag pattern of angulated pigmented linear lines.
- However, the dermatoscopic features of extrafacial LM have not been described previously.
- The differences in rete ridges and concentration of hair follicles between facial skin and other body sites raises the question as to whether dermatoscopic features of LM could be applied to extrafacial LM.
- We propose that the dermatoscopic features of extrafacial LM are a combination of some of the features of LM and SSM *in situ*, but the presence of rhomboidal structures and asymmetrical perifollicular pigmentation in extrafacial lesions implies extrafacial LM.

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CPD questions

Question 1

Which of the following dermatoscopic features suggest a diagnosis of lentigo maligna?

a) Blue–white veil.
b) Irregular pigment network.
c) Streaks and rhomboidal structures.
d) Depigmentation.

Question 2

Which of the following is true?

a) Superficial spreading melanoma has a prolonged *in situ* phase compared with extrafacial lentigo maligna.
b) The lesions of extrafacial lentigo maligna is usually located on the lower leg.
c) Extrafacial lentigo maligna is characterised histologically by a pagetoid upward spread of melanocytes.
d) The dermatoscopic feature of asymmetrical follicular openings corresponds to the uneven descent of malignant cells within hair follicles.

Instructions for answering questions

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- Reflect on the article
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