Diverse presentations of acral melanoma

Background
Acral melanoma (AM) is an uncommon melanoma subtype occurring on the palms, soles and nail apparatus. It often lacks the typical features of primary melanoma resulting in delayed diagnosis.

Objective
This article aims to raise awareness of AM and promote a high index of clinical suspicion to enable early diagnosis and improve outcomes for patients with AM.

Discussion
The diagnosis of AM is often delayed because its presentation mimics other benign conditions such as fungal infections and ulcers. When lesions that were thought to be benign fail to respond to appropriate therapies, biopsy is critically important to exclude AM or other malignant pathology. Clinician awareness of the diversity of AM presentations, maintaining AM as part of their differential diagnosis and facilitating early biopsy are essential for early diagnosis and improving outcomes in patients with AM.

Keywords
skin diseases; skin neoplasms; diagnosis, differential

Australia has the highest rate of melanoma in the world with individuals having a one in 17 lifetime risk of developing melanoma. Acral melanoma (AM) is an uncommon subtype of cutaneous melanoma that occurs on the soles and palms, as well as in the nail unit. AM is more common on the feet than on the hands. The incidence of AM is equal among races, but it is the most common melanoma subtype in dark-skinned populations due to the rarity of other melanoma subtypes. AM is estimated to account for only 1–3% of cutaneous melanomas in Australia and up to 36% in dark-skinned populations.

Several patients have recently presented with a delayed diagnosis of acral melanoma (AM) at the Melanoma Institute Australia (MIA). MIA treats more than 1500 new patients with melanoma each year and 1% of these have AM. AM often has a poor prognosis and the purpose of this article is to raise awareness of AM to facilitate early diagnosis and treatment with the ultimate aim of improving patient outcomes.

Case presentation
A 62-year-old man presented with a 2-year history of left foot symptoms. At presentation to MIA he had a reddish ulcerated lesion centered at the fourth toe and extending across the lateral three web spaces on both the plantar and dorsal surfaces of the foot. The lesion was amelanotic (Figure 1 I, J). His symptoms were of itch, pain and intermittent contact bleeding. He had seen multiple doctors and had been using topical antifungal treatments for almost 2 years. Partial biopsy demonstrated an intermediate thickness melanoma with a Breslow thickness of 2.5 mm. Adverse features included ulceration and a mitotic count of 1/mm². Clinically, there was no evidence of in-transit metastasis (metastases in the skin or subcutis between the primary tumor and the regional lymph nodes), no palpable regional lymphadenopathy and a staging positron emission tomography–computed tomography (PET–CT) scan showed no evidence of distant metastatic disease. Complete excision required forefoot amputation and the final pathology was similar to that of the biopsy. The regional lymph node field, identified as the left groin by...
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Clinical

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When these benign pathologies fail to resolve with appropriate management the diagnosis of AM must be excluded. Other patients have failed to appreciate the presence of pathology on their soles, which may not have been inspected recently.

The aetiology of AM is poorly understood. Risk factors such as fair skin type, ultraviolet (UV) exposure, the presence of acral nevi and trauma may be identified in a patient’s clinical history, but have not been consistently associated with AM.7–10

Clinically, AM can be pigmented and appear as a classical melanoma with asymmetry, border irregularity, colour variation, a diameter >6 mm and evolution of the lesion (using the ABCDEs) (Figure 1 A, B and C). However, a large proportion of AM is amelanotic, appearing pink in colour, and making diagnosis much more difficult. It can be as subtle as a change in skin texture (Figure 1 D, E), appear pink and nodular (Figure 1 G, H) or ulcerated (Figure 1 I, J). As a consequence, AM can easily be misdiagnosed and a high index of suspicion is needed when managing lesions of the feet and hands.

Subungual melanoma most commonly presents as longitudinal melanonychia (a brown-black

Discussion

Patients with AM most commonly present with lesions that have changed in size, colour or form, or that are bleeding, painful or itchy.7 The diagnosis of AM is often delayed because the patients have attributed their symptoms to much more common benign conditions, the most common being warts, fungal infections, haematomas and ulcers.7 When these benign pathologies fail to resolve with appropriate management the diagnosis of AM must be excluded. Other patients have failed to appreciate the presence of pathology on their soles, which may not have been inspected recently.

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Dermatcopy is an adjunct to clinical examination that can be used by clinicians experienced in the technique to assist with the diagnosis of AM. Specific dermatoscopic features are associated with pigmented lesions on the glabrous skin of palms and soles. Parallel ridge pattern and irregular diffuse pigmentation are findings that are highly specific for melanoma and can assist in differentiating AM from benign acral naevi. Dermatcopy has not been well validated in the diagnosis of non-pigmented AM and biopsy should be performed when there is clinical concern.

The prognosis of AM is generally poor and this has often been attributed to delayed diagnosis. However, in a recent series, AM patients were stage matched with patients with non-acral cutaneous melanoma (NACM) of the limb and AM patients were shown to have a poorer disease specific survival (5-year survival 70% for AM compared to 83% for NACM) suggesting that AM is biologically more aggressive. This also correlates with a different spectrum of genetic mutations in AM compared to NACM. BRAF mutations occur in approximately 50% of all melanomas but are less common in AM. In contrast, KIT mutations, which are rare in sun-damaged skin, occur in only 3% of all melanomas. Hence KIT mutations for AM patients have been proposed as a biomarker to facilitate early diagnosis and treatment so as to give AM patients their best chance of cure.

Conclusion

AM is an uncommon melanoma subtype. The clinical appearance of AM is often atypical and this can delay recognition and diagnosis. Clinicians need to maintain a high index of suspicion and arrange appropriate biopsy to facilitate early diagnosis and treatment so as to give AM patients their best chance of cure.

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