

Subungual amelanotic melanoma of the hallux: Review of the literature with a case report

MJ Tladi MBChB(UL Medunsa Campus), FCS (SA)Ortho, MMed(Orth Surg)(SMU)

Dr George Mukhari Academic Hospital, Ga-Rankuwa, Pretoria and Sefako Makgatho Health Science University,
Ga-Rankuwa, Pretoria, South Africa

NP Saragas MBBCh(Wits), FCS(SA)Ortho, MMed(Orth Surg)(Wits)

Netcare Linksfield Orthopaedic Sports & Rehabilitation Centre (Clinic), Johannesburg and Orthopaedic Department,
University of the Witwatersrand, Johannesburg, South Africa

PNF Ferrao MBChB(Pret), FCS(SA)Ortho

Netcare Linksfield Orthopaedic Sports & Rehabilitation Centre (Clinic), Johannesburg and Orthopaedic Department,
University of the Witwatersrand, Johannesburg, South Africa

A Strydom MBBCh(Wits), FCS(SA)Ortho, MMed(Orth Surg)(Wits)

Orthopaedic Department, University of the Witwatersrand, Johannesburg, South Africa

Corresponding author:

Dr MJ Tladi

Department of Orthopaedics

Sefako Makgatho Health Science University

PO Box 25

0204 Medunsa

Tel: 012 521 4049

Fax: 012 521 4284

Email: mphojohn@webmail.co.za

Abstract

Subungual melanoma of the hallux is a rare malignancy occurring mainly in elderly black Africans and Asians. It is often misdiagnosed as a benign lesion. We present a review of the English literature and report on a case of an 80-year-old Caucasian female patient who presented with a rare subungual amelanotic malignant melanoma of the hallux that was initially misdiagnosed as an ingrown toenail. Due to lack of pigmentation, amelanotic melanoma poses a huge dilemma in diagnosis. Upon comparing this case with the current literature, we propose that unlike most acral subungual melanomas, subungual amelanotic melanoma occurs mainly in elderly Caucasian females.

Level of study: IV case series

Key words: subungual melanoma, melanoma, amelanotic, acral lentiginous melanoma

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Introduction

Subungual melanoma was first fully described in 1886 by Sir Hutchinson,¹ yet in the 21st century this condition is still misdiagnosed resulting in a high morbidity and mortality. The famous reggae singer, Bob Marley, succumbed to an aggressive subungual malignant melanoma of the hallux at the age of 36 years.^{2,3} Lack of pigmentation in amelanotic subungual melanoma further complicates an already difficult diagnosis.^{4,6}

Case report

Clinical history

We present an 80-year-old Caucasian female who presented with an 18-month history of a lesion under her right hallux toenail. She consulted a podiatrist who initially diagnosed it as an ingrown toenail and managed it accordingly. The lesion did not resolve and she continued to have episodes of bleeding with closed shoes.



Figure 1. A granulomatous lesion of the hallux



Figure 2. X-ray showing no bony involvement

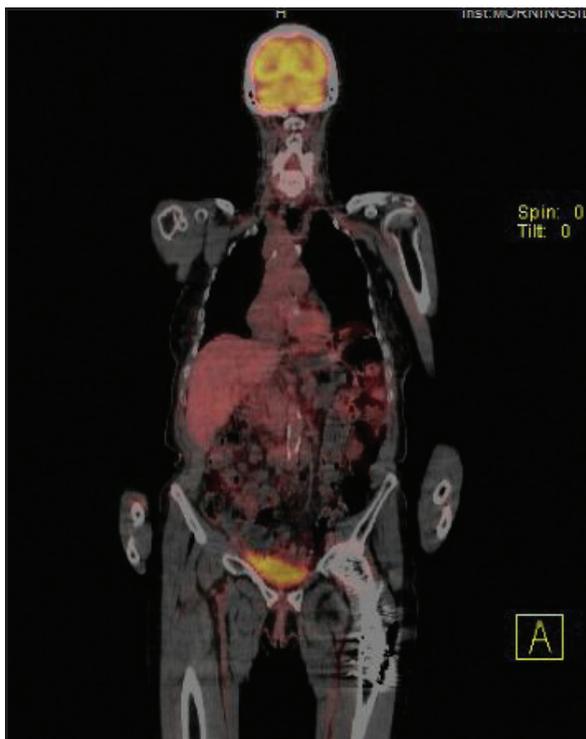


Figure 3. PET CT scan with no evidence of metastasis

The patient was referred to a dermatologist who performed a biopsy. The diagnosis of an amelanotic melanoma was made upon histology. There was no history of previous trauma to the nail or positive family history of melanoma (5%–10% reported incidence of a positive family history).⁷

Examination

Examination revealed a generally well elderly Caucasian female. A granulomatous type lesion, measuring 2 cm by 1.5 cm, was present overlying the nail bed of the right hallux (Figures 1a and 1b). The lesion was tender to palpation. No ipsilateral inguinal lymph nodes were palpable and the foot was neurovascularly intact.

Special investigations

No bony involvement was evident on plain radiographs (Figure 2). A PET CT scan (Figure 3) showed no metastases.

Management

Once the case was discussed with an oncologist, informed consent was obtained from the patient to perform a partial amputation of the hallux. The hallux was amputated at the level of the mid-shaft of the proximal phalanx, allowing for at least a 15mm clear margin from the tumour edge (Figures 4a and 4b). The histology report once again confirmed the amelanotic melanoma, stage IIb (Figure 5), with 20 mm clear margins. The patient was referred to the oncology unit for further treatment as required.



Figure 4a. Level of amputation



Figure 4b. Post-operative X-ray

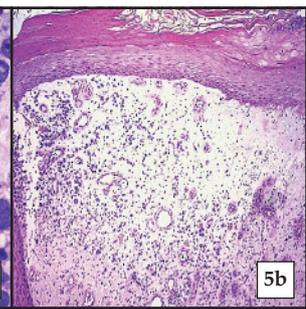
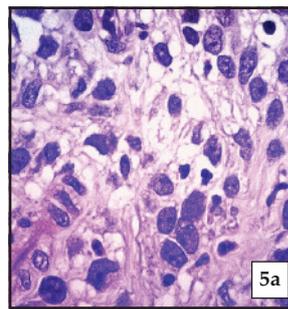


Figure 5. Histology showing amelanotic melanoma

Discussion

Pigmentation of the toenails can be due to benign, systemic disease manifestation or malignant conditions, with melanoma being one of the rare malignancies (3.2% of foot and ankle tumours).^{8,9} Generally there are four types of melanoma, with acral melanoma affecting the hands and feet. This usually occurs in black African and Asian populations contrary to the other types which are more common in fair-skinned populations.^{7,9} Delayed diagnosis of subungual melanoma is largely due to misdiagnosing the lesion as being benign (Table I).^{4,8-12} Table II shows all case reports of subungual amelanotic melanoma affecting the hallux, all of which had a delay in the diagnosis. Only five case reports were identified after an extensive search of the English literature.

Table I: Common benign conditions resembling subungual melanoma

Subungual haematoma	Subungual exostosis
Paronychia	Mucous cyst
Ingrown toenail	Subungual fibroma
Granuloma	Keratoacanthoma
Ethnic pigmentation	Dermatofibroma
Onychomycosis nigricans	Wart
Glomus tumour	
Benign naevus	

The incidence of subungual melanoma is high in black Africans and Asians between the ages of 50 to 70 years. Males and females are equally affected.^{4,7,8} Our case, however, was in an 80-year-old Caucasian female. Upon reviewing the literature reporting on subungual amelanotic melanoma of the hallux, all patients were Caucasian, with 60% being female. We thus propose that subungual amelanotic melanoma actually has a higher incidence in Caucasian females. Symptoms in most cases are vague and the lesion resembles other conditions. The ABCDEF rule was described by Levit *et al.* to reduce misdiagnosing subungual melanomas and improve early detection (Table III).¹⁶ Hutchinson's sign describes a black discoloration of the proximal nail fold which is suggestive of subungual melanoma. This sign has inherent flaws as it has no role in amelanotic melanoma due to absence of pigmentation, as was seen in our patient. Hutchinson's sign can also be associated with Laugier-Hunziker syndrome, ethnic pigmentation, infections or the use of certain medications. When present, however, it is indicative of a poor prognosis.^{8,16}

Management of subungual melanoma of the hallux is usually amputation of the digit, which can cause functional and emotional problems, such as in the case of Bob Marley who refused amputation due to religious reasons and only agreed to local excision of the tumour.^{24,17}

Table II: Literature case reports of hallux subungual amelanotic melanoma

Race	Age (years)	Sex	Initial diagnosis	Author	Surgical management
Caucasian	71	female	in-growing toenail	Cahill <i>et al.</i> ¹¹	amputation
Caucasian	35	female	in-growing toenail	Winslet <i>et al.</i> ¹³	amputation
Caucasian	31	male	mass	Gosselink <i>et al.</i> ⁶	amputation
Caucasian	61	male	pyogenic granuloma, squamous cell carcinoma, amelanotic malignant melanoma, deep fungal infection, verruca or cutaneous leishmaniasis	Arican <i>et al.</i> ¹⁴	amputation
Caucasian	72	female	fungal infection	Koch <i>et al.</i> ¹⁵	amputation

Table III: Steps to follow when suspecting subungual melanoma

A	Age: Range 20–90 y, peak 5th–7th decades Race: African-American, Native American, Asian
B	Band (nail band): pigment (Brown-Black) Breadth (≥ 3 mm) Border (irregular/blurred)
C	Change: Rapid increase in size/growth rate of nail band Lack of Change: Failure of nail dystrophy to improve despite adequate treatment
D	Digit involved: Thumb > hallux > index finger Single digit > multiple digits Dominant hand
E	Extension: Extension of pigment to involve proximal or lateral nail fold (Hutchinson's sign) or free edge of nail plate
F	Family or personal history: Of previous melanoma or dysplastic naevus syndrome

This ultimately resulted in metastasis and his demise. As the diagnosis is often delayed with resultant local invasion, amputation is the recommended management.¹⁷

The tissue must be sent for histology and the report should include: histological type, presence of ulceration, presence of infiltrative lymphocytes, regression, microsatellite lesions, margins, micro-staging (tumour thickness according to Breslow and level of invasion according to Clark) which are helpful with final staging.^{7,10} Melanoma is staged according to the American Joint Committee on Cancer (AJCC).⁷ A sentinel lymph node (SLN) biopsy is recommended for melanomas stage 1b and above.⁷ Recurrence of foot melanoma is 37% at approximately 3 years and survival rate after recurrence is shorter than 41 months.¹⁸ Prognosis of subungual melanoma in general is poor. The 5-year survival rate of subungual melanoma of the toe is 40% as compared to 72% in the finger.¹⁷

The 5-year survival rate for melanoma in general is 74.3% in the foot and 85.2% in the leg.¹⁸ The survival rate is directly related to the staging by the AJCC.

Conclusion

Subungual melanoma is rare, especially in the Caucasian population. However, the incidence of subungual amelanotic melanoma has only been reported in Caucasians, contrary to other subungual melanomas which occur mainly in black Africans and Asians. There is also a predilection for the female gender. We thus propose that subungual amelanotic melanoma occurs mainly in elderly Caucasian females. The diagnosis is often delayed, as it can mimic a variety of hallux nail conditions. Due to lack of pigmentation, amelanotic melanoma poses a huge dilemma in diagnosis. A high index of suspicion is thus imperative. The use of the ABCDEF rule can help in increasing awareness and reducing the delay in diagnosing subungual amelanotic melanoma. An incisional biopsy should be performed early. Once histological confirmation is made, amputation is the treatment of choice and the patient should be referred to an oncology unit for further management according to the AJCC staging.

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Compliance with ethics guidelines

Conflict of interest statement

The authors declare they have no conflict of interests in writing this article. No financial support was received for this study. The content of the article is the original work of the authors.

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