Case History: A Case of primary Amelanotic Malignant Melanoma

Imran Ahmad[1], Bilal Hasan Chaudhry[2]

(1) Consultant Family Medicine, Primary Healthcare Corporation, Doha Qatar
(2) Consultant Family Medicine, Primary Healthcare Corporation, Doha Qatar

Corresponding author:
Bilal Hasan Chaudhry,
Consultant Family Medicine, Primary Healthcare Corporation,
Doha, Qatar
Email: bchaudhry@phcc.gov.qa

Received: February 2023 Accepted: March 2023; Published: April 1, 2023.
Citation: Imran Ahmad, Bilal Hasan Chaudhry. Case History: A Case of primary Amelanotic Malignant Melanoma. World Family Medicine. March 2023; 21(3): 112-116 DOI: 10.5742/MEWFM.2023.95256082

Abstract

This case study discusses the atypical presentation of amelanotic melanoma in a 37-year-old Caucasian female patient and the challenges it poses for diagnosis and the consequent delay in treatment. 5-year survival rate is 93% for all melanomas but amelanotic melanoma survival is 88% and this is predominantly due to delayed diagnosis(1). Better patient and doctor education and readily available histopathological diagnosis confirmation can lead to earlier diagnosis and better outcome.

Key words: amelanotic malignant melanoma, case presentation

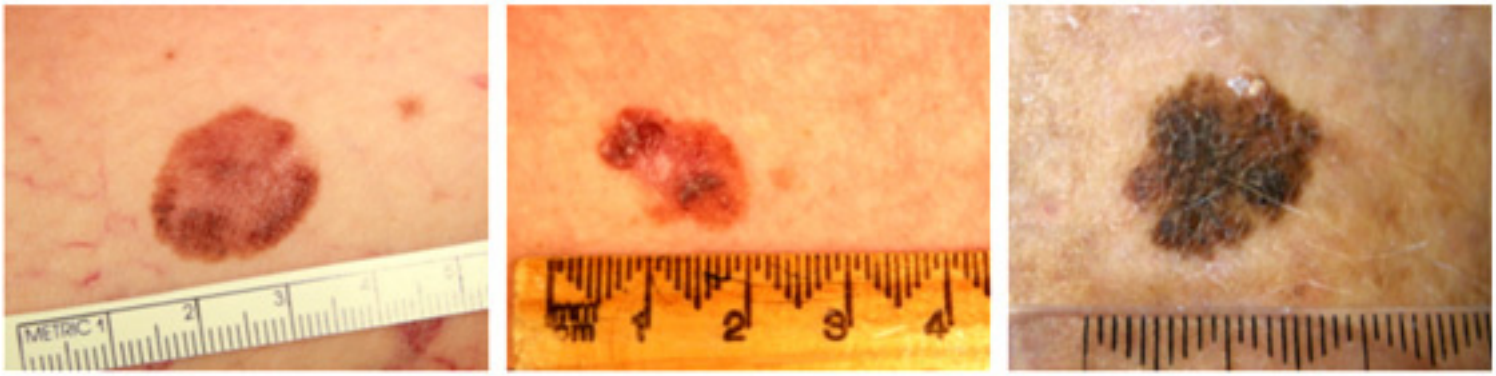


Figure 1 Superficial spreading melanoma; most common are superficial spreading which accountfor 70% of all malignant melanomas. They are commonly found in a young age group (20-40 years old). Common sites of incidence are backs for males and legs for females. They usually present with irregular edge and irregular pigmentation.(8)

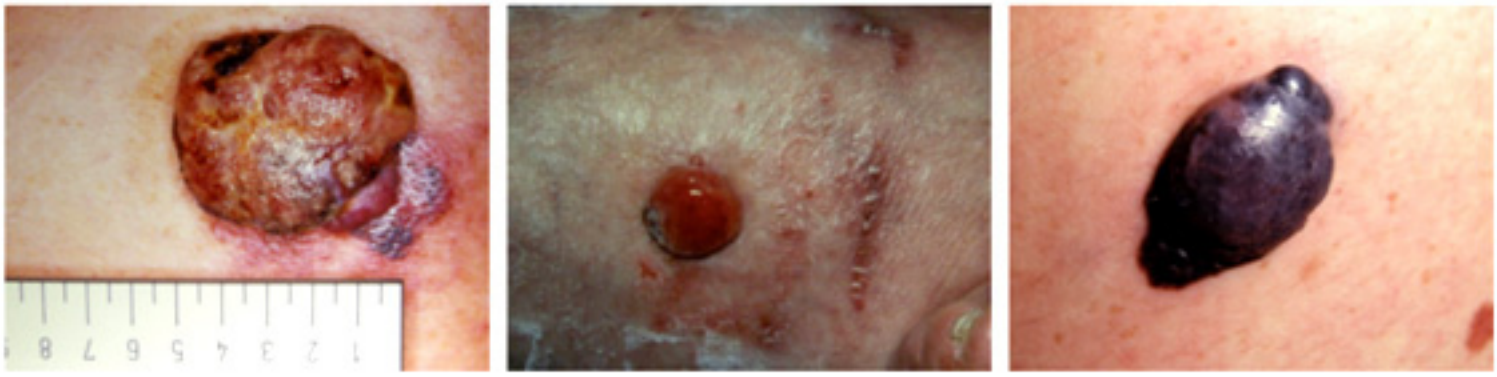


Figure 2 Nodular Melanoma; nodular melanomas constitute 20% of all malignant melanomas. They are commoner in older age group and appear mainly on the trunk. They present as elevated, dome shaped dark brown/black or even red/pink colour skin lesions(8).

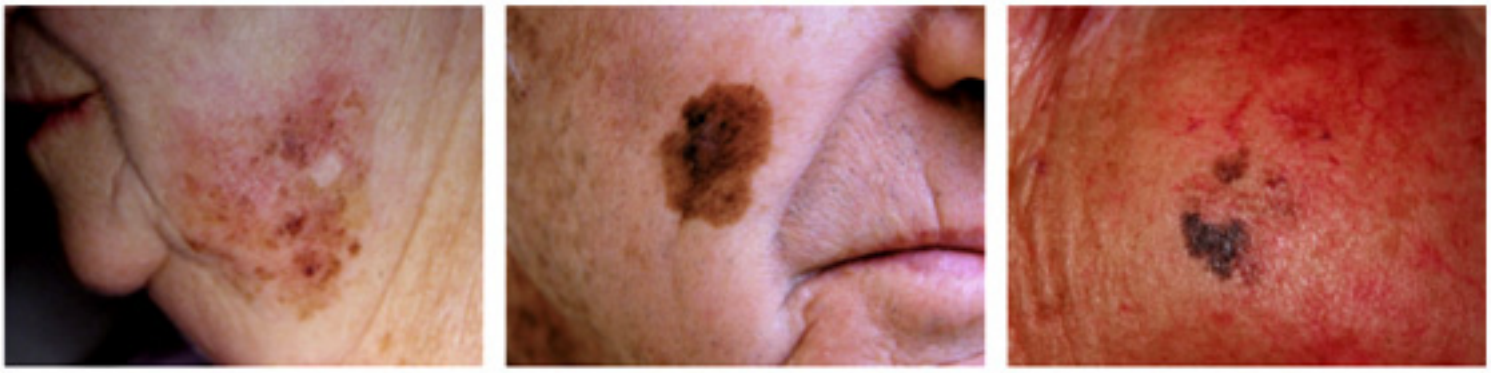


Figure 3 Lentigo malignant melanoma; mainly presents on face as a dark colour freckle and gradually increase to invade surface and develops into a nodule(8).

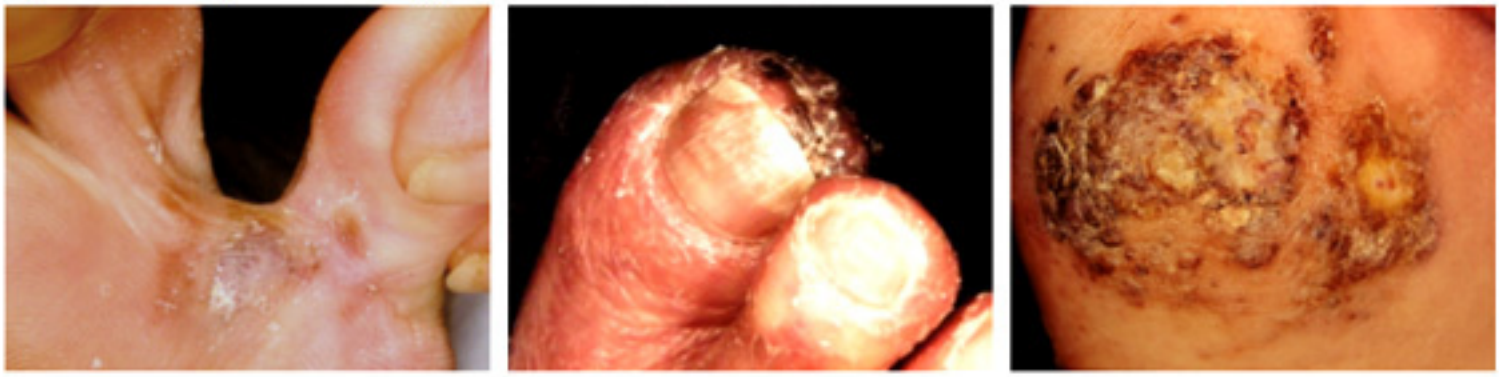


Figure 4 Acral lentiginous melanoma; accounts for approximately 10% of melanomas in the western world and are commonly found on the palms of hands, soles of feet and around nails(5,8).

Introduction

Malignant melanoma is a cutaneous and/or extra cutaneous tumour that arises from the embryological remnants of neural crest cells/melanocytes. While melanomas are commoner in skin they can occur in eyes and internal organs(2). There has been a rapid increase in the incidence of malignant melanoma over the last few decades in all western countries. There are approximately 16,700 new diagnosed cases of malignant melanoma each year in the UK making it the fifth commonest cancer overall in the UK, accounting for 4% of all new cancer cases(3). Over the last decade, melanoma skin cancer incidence rates have increased by around a third (32%) in the UK, and is higher in males compared to females(3).

The main aetiological factors in the development of malignant melanoma are UV radiation and sun exposure along with people of higher socio-economic status and the population with Type 1&2 skin phenotype, red or blonde hair and blue eyes(4,5). Genetics also play an important role in the development of malignant melanoma and approximately 2% of patients diagnosed with malignant melanoma have a positive family history(5). A literature review on melanoma genetics by A. Nelson et al found alteration in cyclin-dependent kinase inhibitor 2A (CDKN2A) gene is the main responsible factor in the development of malignant melanoma(6). Another small study demonstrated the possible role of Human growth hormone in the pathogenesis of malignant melanoma(7).

There are four main sub types of malignant melanoma, all with distinct clinical features. Superficial spreading; Nodular; Lentigo malignant and Acral lentiginous melanomas(5).

Treatment outcome is significantly dependant on the stage of disease at time of diagnosis, the earlier the better.

Presentation and Management

A 37-year-old Caucasian woman presented in routine family medicine clinic with a main complaint of ongoing mechanical back pain. At the end of consultation, she presented a skin lesion on her right second toe. As far as she could remember the lesion started as a small blister almost 7 months ago on the ventral surface of her right second toe. This gradually became bigger and broke the skin surface. Occasionally it wept with pinkish fluid and had a strong odour. It caused slight discomfort on walking. The lesion was not itching and there were no other systemic symptoms. She did not have other similar lesions elsewhere on the body. She was otherwise a healthy woman with no significant past medical and dermatological history. No family history of skin cancers. She lived with her husband and five children. Only surgical history was tubal sterilization after the birth of her youngest child six months ago.

At her initial consult with a General Practitioner (GP) the suspected diagnosis was superficial skin infection and she was given a one week course of Flucloxacillin. There was no improvement with antibiotics and the lesion continued to get bigger. She contacted another GP a month later about this and diagnosis was changed to fungal infection of foot. She was given topical treatment with regular dressings. She also had routine blood tests which revealed chronic mild anaemia and normal inflammatory markers and normal glucose. Thereafter there had been no change in the skin lesion which continued to stain her socks with mild discomfort on walking.

On examination she appeared well with mild restriction of her back movements. She was apyrexial with other vitals within normal range. The lesion in question was present on the plantar surface of right second toe. It was approximately 20 X 10mm in diameter, red coloured, symmetrical with regular raised borders, friable and a malodourous flat lesion with no pigmentation. The lesion appeared inflammatory and clean with no frank infection and bled easily on contact. There were no associated nail changes in any of the toes. The rest of the skin examination revealed several naevi which varied a little bit in size and shape, however all appeared benign.

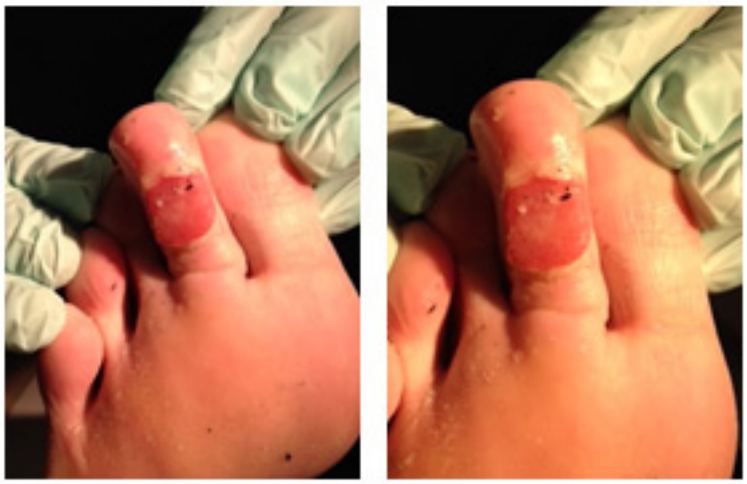
History and clinical presentation of the lesion was atypical and differentials at the time were bacterial skin infection, Fungal infection of foot, Eczema, Contact dermatitis and a Pyogenic granuloma.

Due to the chronic nature of the lesion, swab was sent for culture and sensitivity before initiating further treatment. The culture result was positive for Group C-G Streptococcus (S. dysgalactiae) which are prone to causing skin abscess. According to culture and sensitivity result the patient was treated with a 2 weeks course of Penicillin.

On review at end of antibiotics the skin lesion remained the same with no improvement in signs or symptoms. As per skin cancer guidelines, a persistent skin lesion which is unresponsive to treatment with uncertain diagnosis, she was referred to suspected cancer clinic(9).

Dermatologist initially treated this lesion as pyogenic granuloma with amelanotic melanoma and an eccrine poroma on differentials list. This was treated with topical Mupirocin. Subsequently she underwent urgent biopsy of the lesion which confirmed that it is a Malignant Melanoma of 2.7mm Breslow thickness with a mitotic count of 9 and is ulcerated. She underwent amputation of right second toe with sentinel node biopsy for tumour grading and suspected metastasis. It was found that she had developed various metastasis and unfortunately she passed away soon after with carcinomatosis.

Figure 5: Lesion as presented; (pictures were taken with the patients' consent)



Discussion

Amelanotic melanomas constitute only 2-8% of all melanomas(10). They are often missed due to lack of pigmentation. While criteria such as ABCDE (asymmetry, irregular borders, colour variation, diameter >6mm and evolution) are useful in diagnosis of cutaneous pigmented melanoma there is no explicit clinical appearance that is unique to the amelanotic variant(11). They are usually diagnosed at a very advanced stage when the lesion is nodular, vascular or ulcerated(12). N. Jaimes et al found in their study on 20 consecutively diagnosed amelanotic melanomas that all were erythematous, red, symmetric with regular borders and lacked clinical ABCD features commonly found in melanomas(10). Of those cases 70% exhibited a scaly appearance, which has been reported in numerous case reports of amelanotic melanoma (13–15). The report also identifies the use of dermatoscopy in correctly diagnosing these melanomas due to the presence of polymorphous vascular pattern(10). William V. Stoecker et al found that diagnostic sensitivity and specificity of amelanotic melanomas increased to 89% with use of dermatoscopy as compared to 65% without it(16). The dermatoscopic features of amelanotic melanomas include serpentine, dotted vessels throughout the lesion with central milky-red areas(17).

The case in discussion is likely Acral Lentiginous Amelanotic Malignant Melanoma due to its presence on the sole of the foot(5,18). It is a rare melanoma subtype which is mostly prevalent in dark skin population(5). Overall 5 and 10 year survival rate of acral malignant melanomas is worse than other cutaneous malignant melanomas(18,19).

Surgery is the mainstay treatment of primary cutaneous malignant melanoma (9) with margin of excision determined by the Breslow thickness(4,20). Melanomas measuring 2.01 to 4.0 mms, as in the above mentioned case, require 2-3cm margin for excision. Local recurrence of melanoma is about 5% by 2 years(9). The role of adjuvant treatment, chemotherapy and melanoma vaccines is controversial and so far show no evidence to suggest overall survival improvement in primary cutaneous malignant melanoma(4,9). Usually all patients diagnosed with malignant melanomas require specific follow up regimen on the basis of formal tumour grading(9).

Conclusion

The incidence of malignant melanoma has increased over the last few decades. While early recognition has improved prognosis of these life-threatening skin cancers there are rare forms which pose a diagnostic challenge. This case is a clear example of delayed diagnosis primarily because of the rare anatomical position and atypical presentation with lack of pigmentation.

This case was used as a learning tool to educate other physicians and nurses of the atypical presentation. Patient and family were provided with ongoing support and care as any cancer diagnoses has profound impact on patients quality of life(21).

Learning Points

- Early dermatoscopy can assist diagnosis and is often easier to access in community dermatology clinics.
- Do not try laser therapy for mole removal as it can alter vascular morphology hence hindering diagnosis.

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