

ATYPICAL CLINICAL APPEARANCE AND LOCATION OF AN ACRAL LENTIGINOUS MELANOMA IN A PREGNANT AFRICAN ALBINOS: A CASE REPORT

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ABSTRACT

Melanoma is a potentially serious skin cancer that arises from pigment cells (melanocytes). Acral lentiginous melanoma (ALM) is a kind of lentiginous skin melanoma. It is commonly observed on the palms, soles, under the nails and in mucous membranes. Unlike other forms of melanoma, ALM does not appear to be linked to sun exposure. This cancer is rare in Caucasians and people with lighter skin types; it is the most common subtype in people with darker skins. It is the most common form of melanoma diagnosed amongst Asian and Black ethnic groups. The average age at diagnosis is between sixty and seventy years. About 1 to 3% of melanomas in Australia and New Zealand are ALM. We do not have statistics on the prevalence of this tumor in the black African population, much less in African albinos. But the rarity of this cancer in African albinos is no doubt. Indeed, a histopathological retrospective review of 10 years in Tanzania has only found one case of this pathology. We report a case of ALM in an African albino patient with 4 particularities, an early onset age (25), a state of gestation (32 weeks), a location (orbital rim) and a clinical aspect (achromatic).

Keywords: Acral Lentiginous Melanoma - African Albinos Cance - Tumor and Gestation

INTRODUCTION

Reed [1] in 1976 reported the first case of ALM. Since ALM was considered as a distinct subtype of skin melanoma. It is considered much more aggressive than other melanoma subtypes [2]. It is commonly observed on the palms, soles, under the nails and in mucous membranes [3-5]. It occurs on non hair bearing surfaces of the body which may or may not be exposed to sunlight. Unlike other forms of skin melanoma, ALM does not appear to be linked to sun exposure. This cancer is rare in Caucasians and people with lighter skin types; it is the most common subtype in people with darker skins [2, 6, 7]. It is the most common form of melanoma diagnosed amongst Asian and Black ethnic groups. The average age at diagnosis is between sixty and seventy years. About 1 to 3% of melanomas in Australia and New Zealand are ALM [8].

We do not have statistics on the prevalence of this tumor in the black African population, much less in African albinos. But the rarity of this cancer in African albinos is no doubt. Indeed, a histopathological retrospective review of 10 years in a specialized center in Tanzania has only found one case of this pathology.

We report a case of peri orbital ALM in a 25 years old women African albino in state of gestation.

CASE PRESENTATION

Mrs. AH aged 25, suffering from albinism, was seen for a left periorbital mass evolving for nearly a year. This mass is located near the outer canthus, achromatic, highly vascularized, pie-shaped, about 5 X 6 cm sizes, painless, hard consistency, and mobile at its base (Figure 1).



There is no locoregional lymphadenopathy. Moreover, she was in state of gestation (32 weeks). The obstetrical examination was normal (monofoetal pregnancy, fetal active movements, uterine height, fetal heart sounds).

Faced with the risk of bleeding and obstetric stress, in agreement with obstetricians, we decided to carry in one time under general anesthesia wide surgical excision (margin of 1 cm on healthy skin) (Figure 2).



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Pathological examination showed a diffuse lentiginous pattern of melanocytes with focal atypism. Melanocytes were localized in both the basal and the spinous layers. The postoperative course was uneventful. Mrs. AH is restricted by a quarterly dermatological supervision for one year. She delivered vaginally a healthy black child.

DISCUSSION

Kiprono [3] in Tanzania shows that over 75% of skin tumors in African albinos are located at the face and neck. This is not surprising since most of these cancers are related to solar radiation. The special feature of this finding is due to the fact that the appearance of ALM has no direct link with exposure to sunlight. Secondly, ALM is commonly observed on the palms, soles, under the nails and in mucous membranes [9].

This Tanzanian study [3] also noted that in almost 99% of cases cancer in African albinos population are dominated by Squamous Cell Carcinoma followed by Basal Cell Carcinoma. Hormonal changes related to pregnancy can they explain this atypical presentation of ALM at Mrs AH? Unlikely, because the ALM is not a hormone cancer, and literature does not report cases distribution according to gender of this tumor [5, 9, 10].

ALM is generally described as a highly pigmented tumor [3, 9, 11]. Although we do not find in the literature a case of ALM unpigmented, the Color by itself does not suffice to rule out the diagnosis of melanoma. Authentic achromatic malignant melanoma of the choroid are described [12].

The average age of onset of ALM is situated around 60 [4-5, 9], well above that of Mrs AH (25). Albinism and pregnancy may be co factors behind this feature.

Early diagnosis of ALM is important for a better prognosis but its diagnosis is not always easy and it is often misdiagnosed. All the patients complained of a pre-existing lesion which is increasing in size or spreading to adjacent area. Most of the lesions located on the nail apparatus showed black to brown discoloration involving the whole nail plate, nail deformation, and Hutchinson's sign at the time of diagnosis [9, 11].

The use of a dermatoscope (Dermoscopy) by a Dermatologist or other doctor trained in its use can be very helpful in distinguishing ALM from other skin lesions [9, 13].

The most frequently observed dermoscopic features of acral lentiginous melanoma are:

- Asymmetrical structure and colours
- Parallel ridge pattern of pigment distribution
- Blue-grey structures

The pathologist's report should include a macroscopic description of the specimen and melanoma (the naked eye view), and a microscopic description. The report may also include comments about the cell type and its growth pattern, invasion of blood vessels or nerves, inflammatory response, regression and whether there is an associated naevus [9-11, 13]. The Breslow thickness is reported for invasive melanomas. It is measured vertically in millimeters from the top of the granular layer (or base of superficial ulceration) to the deepest point of tumor involvement. It is a strong predictor of outcome; the

thicker the melanoma, the more likely it is to metastasis. The Clark level [5, 9, 14] indicates the anatomic plane of invasion:

- Level 1 In situ melanoma
- Level 2 Melanoma has invaded papillary dermis
- Level 3 Melanoma has filled papillary dermis
- Level 4 Melanoma has invaded reticular dermis
- Level 5 Melanoma has invaded subcutaneous tissue

The deeper the Clark level, the greater the risk of metastasis. It is useful in predicting outcome in thin tumors and less useful for thicker ones in comparison to the value of the Breslow thickness.

Histopathological analyze showed a diffuse lentiginous pattern of melanocytic proliferation along the dermoepidermal junction with hyperplastic epidermis. In some cases there is a melanocytic hyperplasia with focal atypism but most cases revealed diffuse large atypical melanocytes with irregular shapes and hyperchromatic nuclei. The melanocytes can be localized in the basal and the spinous layers. When immunohistochemical study is performed, all lesions are positively stained with anti-HMB-45 but only weakly with anti-S-100 antibodies [9, 13, 15, 16].

The initial treatment of a primary melanoma is to cut it out; the lesion should be completely excised with a 2-3 mm margin of normal tissue. Further treatment depends mainly on the Breslow thickness of the lesion.

After initial excision biopsy; the radial excision margins, measured clinically from the edge of the melanoma, recommended in the Australian and New Zealand Guidelines [8] for the Management of Melanoma are shown in the table below (2008). This may necessitate flap or graft to close the wound. In the case of acral lentiginous and subungual melanoma, this may include partial amputation of a digit. Occasionally, the pathologist will report incomplete excision of the melanoma, despite wide margins. This means further surgery or radiotherapy will be recommended to ensure the tumor has been completely removed.

Thickness	Excision margin
Melanoma in situ	5mm
Melanoma < 1.0mm	1cm
Melanoma 1.0–2.0mm	1–2cm
Melanoma 2.0–4.0mm	1–2cm
Melanoma > 4.0mm	2cm

If the melanoma is widespread, other forms of treatment may be necessary, but are not always successful in eradicating the cancer. Immunotherapy, biologics such as Ipilimumab, Vemurafenib and Dabrafenib are showing promise [8, 9, 17]. Adjuvant chemotherapy is sometimes used by some authors (six cycles of Cisplatin, Vinblastine and Dexamethasone) [9, 18].

The main purpose of follow-up is to detect recurrences early. The Australian and New Zealand Guidelines [8] for the Management of Melanoma (2008) makes the following recommendations for follow-up for patients with invasive melanoma:

- Self skin examination
- Regular routine skin checks by patient's preferred

health professional

- Follow-up intervals are preferably from three-monthly to 12 months depending on the stage of the disease
- Individual patient's needs should be considered before appropriate follow-up is offered
- Provide education and support to help patient adjust to their illness

The follow-up appointments may be undertaken by the patient's general practitioner or specialist

CONCLUSION

Typically ALM occurs on the hairless skin of palms, soles or beneath the nail plate. Clinically it shows a brown to black macule or patch with color variation and irregular borders. However in the presence of an achromatic skin tumor of the face, the diagnosis of ALM should also mention.

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