

# A tragedy of albinism

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We would like to bring to your attention the plight of a 45-year-old epileptic, HIV-positive African male with oculo-cutaneous albinism (OCA). He presented to the dermatology department of a tertiary hospital in Gauteng in 2019 with a fungating mass on his left hand that had been progressively enlarging for a year. Surprisingly, a diagnosis of hyperkeratotic actinic keratosis with mild dysplasia was found on biopsy and he was treated conservatively. He was lost to follow-up during the COVID-19 pandemic. He re-presented in 2021 with marked expansion of the mass, left arm swelling and multiple foul-smelling lesions as shown in Figure 1a and 1b.

Magnetic resonance imagery (MRI) and computed tomography (CT) staging scans showed soft tissue, bone and cutaneous extension with left medial epitrochlear and axillary nodes and suspicious pulmonary nodules in his right upper lobe. A biopsy was done, and the histology revealed a pseudo-vascular variant of an invasive squamous cell carcinoma (SCC). In light of the extent of the disease and clinical evidence of ascending sepsis, a forequarter amputation was recommended. After discussion, the patient agreed to this course of action. A standard forequarter amputation was performed under general anaesthesia.

The patient had delays to follow-up owing to socio-economic and psychological constraints, which demotivated him from leaving his house to seek medical attention. He later presented with a postoperative wound that had complicated with a superficial surgical site infection related to retained skin clips. Despite clip removal and broad-

spectrum antibiotics, he re-presented a month later with a gaping wound secondary to an assault by individuals who accused him of having sold his arm for money. The wound required debridement and histology of the tissue removed confirmed local recurrence of the SCC. This was treated with wound dressings and irradiation. Despite the local recurrence, healing was eventually achieved (Figure 1c).

OCA is a condition that results in low melanin levels in the hair, skin, and eyes of affected individuals with a prevalence of 1/20 000 live births and a predilection for sub-Saharan Africa.<sup>1-5</sup> Individuals with albinism deal with many medical as well as psycho-social issues ranging from social rejection to physical abuse which, under extreme circumstances, has resulted in death.<sup>1,6</sup> The presence of an albino person in a community has an effect on both the affected individual as well as the community they live in. Specifically, in Africa where the population generally has pigmented skin, affected individuals stand out distinctively. Occurrences of unjustified discrimination, abuse and even killings often occur because affected individuals are seen as easy targets for abuse, just as we observed in this case.<sup>1,3,7</sup> Throughout Africa, the misunderstanding about the condition of albinism has developed into a culture of abuse with the practice of mutilating “albino” persons for their body parts, such as limbs and genitalia, which are deemed to have mystical powers.<sup>3,6,7</sup> In a study published in 2022, there had been more than 203 attacks, with 77 killings, 97 survivors, one abduction, and 26 other violations of the rights of the people living with albinism in Tanzania alone.<sup>8</sup> Moreover,



Figure 1: (a&b) Wound on presentation, and (c) eventual healing

witchcraft mutilations, murders, ritual attacking, infanticide and abandonment of children have been documented in anthropological literature as well.<sup>8</sup> People have even dug up graves of albino people in order to obtain their body parts.<sup>6</sup>

For late presenters with advanced stages of tumour disease and the absence of conservative options, amputations are indicated for the management of those tumours as was the situation in this patient.<sup>9,10</sup> A surgical amputation is a procedure that should only be performed in a clinical setting under surgically verified indications. It is important to emphasise that medico-legal issues arise at multiple points when treating people living with albinism. Hence it is extremely important that informed consent discussions are thorough.<sup>9</sup> Our patient had been accustomed to continual discrimination which impacted his early schooling experiences and his career opportunities. Somehow, he managed to overcome these setbacks by becoming a political activist and reintegrating into the community, only to once again be estranged from the community following the amputation.

In terms of progress regarding what is being done for people with albinism, the Southern African Development Community (SADC) region boasts a large population of persons living with albinism, yet they do not have a facility specifically dedicated to people living with albinism.<sup>8</sup> Some support is provided through the Cancer Association of South Africa (CANSA) through its mobile health clinics that are dedicated to all people at risk of developing cancer, persons living with albinism included.<sup>10</sup> Extending this type of support is important as without adequate medical care and primary health education on the benefits of sun protection, persons in Africa with albinism will continue to have a shortened life expectancy, rarely living past 40 years.<sup>8</sup> Outcomes are influenced by factors such as access to public health facilities, quality of care and patients' relationship with their disease, a situation that requires extensive and continuous counselling. Even though some interventions exist, there is a need for more to be done to protect this vulnerable group from the sequela of their condition and persecution by their communities.

### **Declaration**

The authors declare authorship of this article and that they have followed sound scientific research practice. This research is original and does not transgress any plagiarism policies.

### **Conflict of interest**


The authors declare no conflict of interest.

### **Ethical approval**

Ethical approval was obtained from the University of the Witwatersrand Human Research Ethics Committee (M220793).

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