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Epidemiological Study of Facial Malignancy Tumor in People with Albinism (PWA) in Togo

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Abstract

Introduction: Hypersensitivity of the skin of PWA to ultraviolet rays, resulting from insufficient melanin production, exposes them to precancerous and cancerous lesions. The head is highly exposed and remains the body part most prone to these skin lesions. **Objective:** To describe cancerous and precancerous skin lesions of the head in Togolese PWA through a retrospective study. **Patients and Methods:** Mobile screening and treatment campaigns for cancerous and precancerous skin lesions were conducted in 17 cities in Togo between 2019 and 2021. The medical records of PWA with at least one head lesion were included. Sociodemographic, clinical, histopathological, and therapeutic data were collected and processed. **Results:** Seventy-eight patients, including 28 women and 50 men, were involved, representing a sex ratio of 1.8. The mean age was 32.64 ± 14.64 years. The frequency of lesions of the head and neck was 66.10%. The face was affected in 68.7%, with the auriculotemporal region particularly affected in 24.3%. Clinically, nodular, ulcerated, budding, and crusting lesions represented 36.9%, 30.6%, 4.4%, and 24.8%, respectively. Histopathology revealed SCCs in 34.8% and BCCs in 26.1%. Actinic keratoses represented 14.8%. Excisional biopsy under local anesthesia was performed in 92.2%. **Discussion:** Skin tumors of the head and neck in Togolese PWA were dominated by squamous cell carcinomas. Raising awareness of good photoprotection practices could reduce the occurrence of skin tumors in African albinos.

Keywords

Albinism, Skin Tumors, Head and Neck, Togo

1. Introduction

Albinism encompasses a heterogeneous group of autosomal recessive genetic disorders [1]. It is characterized by hypopigmentation of the skin, hair, and eyes due to insufficient production of melanin pigments in the skin [1]-[3]. Albinism is a universal condition, with an overall incidence of all forms ranging from 1/17,000 to 1/20,000 births [4]. In the United States, its prevalence in the general population is approximately 1/36,000, higher among African Americans (1/12,000). In Spain, among Gypsies, the prevalence of albinism is 1/1200 [5]. In Africa, its prevalence ranges from 1 in 15,000 in Nigeria [2] to 1 in 1000 in Zimbabwe and Niger [3] [4]. The prevalence in Tanzania is estimated at 1 in 2500 inhabitants [6].

People with albinism (PWA) have sandy-colored hair, chalky-white skin, and light brown or blue eyes, making them more susceptible to the harmful effects of ultraviolet (UV) radiation [4] [7] [8]. This hypersensitivity to UV rays is therefore responsible for the high frequency of skin lesions in PWA [7] [9] [10].

Skin involvement is dominated by cancerous and precancerous lesions [11]-[14]. Management consists primarily of cryotherapy for actinic keratoses and surgical excision of lesions suspected of being cancerous, and generally of awareness and information on the use of sunscreen [1] [4] [15] [16].

The head and neck, often poorly protected due to the symbolism of the face, is the preferred site for these skin lesions [12] [14] [15] [17]. However, the involvement of the different areas of the head and neck is very little described in the literature. The present study aims to fill this gap by describing, through a retrospective study, the distribution of cancerous and precancerous lesions of the head and neck in PWA in Togo.

2. Patients and Method

This was a retrospective, descriptive study of medical records of people with albinism (PWA) who underwent surgery during street campaigns and in the maxillofacial surgery department of the Sylvanus Olympio University Hospital in Lomé for cancerous or suspicious skin lesions.

Screening campaigns for cancerous and precancerous skin lesions among PWA were conducted in 17 cities in Togo between 2019 and 2021. These campaigns were conducted by a multidisciplinary medical team composed of dermatologists and a maxillofacial surgeon.

We included in this study all patient records who presented with at least one skin lesion on the head and neck (face, neck, scalp) during the study period. We excluded incomplete and unusable records and patients who were lost to follow-up after treatment. Data collection was conducted using a pre-established survey form. The data collected came from the hospitalized patients' medical records and surgical report registers. Access to patients' medical records was subject to handwritten authorization from the head of the Stomatology and Maxillofacial Surgery (MFS) department of the Sylvanus Olympio University Teaching Hospital and the Director of the Sylvanus Olympio (SO) University Teaching Hospital. Patient an-

onymity was respected throughout the study. We obtained approval from the Ethics Committee of the Faculty of Health Sciences at the University of Lomé. The parameters studied were sociodemographic data (age, sex, occupation, city of origin), clinical features (number of lesions, their clinical appearance, and topography), histopathological features of the lesions, the type of surgical management and outcome data. Diagnostic confirmation was carried out on biopsies or excisional biopsies performed by the surgeon and analyzed by an anatomopathologist. Statistical analysis was performed using Excel 2019 and Epi info 7.2.5 software.

The topography of the lesions was described according to the aesthetic subunit diagram of the face and neck to facilitate comparison with data from the literature.

3. Result

3.1. Epidemiological Aspects

Of the 118 PWA operated on for skin lesions, 78 (66.10%) had at least one lesion on the head and neck (face, neck, scalp). Among the 78 PWA, 56 (71.8%) had at least one facial lesion, representing 47.5% of all PWA operated on. The sample included 28 women (35.9%) and 50 men (64.1%) (sex ratio (M/F) = 1.8) with a mean age of 32.64 ± 14.64 years and a range of 2 to 73 years.

Among the socio-professional groups involved, students represented 26.9%, shopkeepers 17.9%, farmers 16.7%, and housewives 11.5%.

3.2. Clinical Aspects

Skin lesions of the head were single in 67.9% of cases and double in 24.4%. The face was affected in 68.7% of cases; the neck in 22.6%; and the scalp in 8.7% of cases. Within the face, 31.6% of lesions were located in the auriculotemporal region, 22.8% in the oculopalpebral region, and 16.5% in the cheek region.

Clinically, nodular lesions (**Figure 1**) represented 35.8%, ulcerated lesions 30.7%, and budding lesions 4.4%. **Figure 2** summarizes the distribution according to the clinical type of lesions.

Figure 1. Nodular lesion of the inner canthus of the left eye.

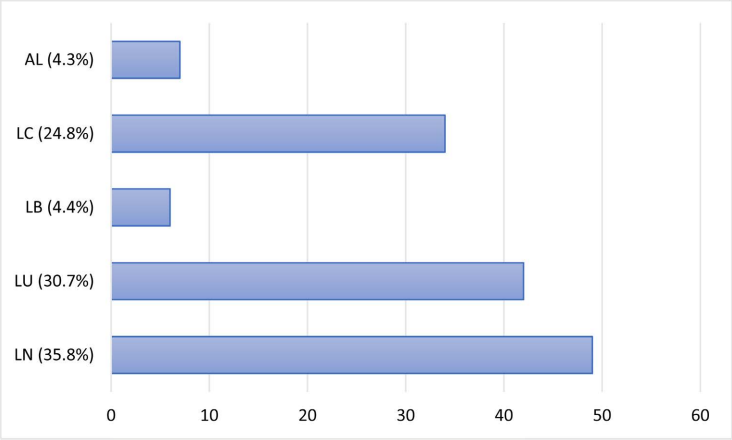

Figure 2. Distribution according to the clinical type of lesions. LN: Nodular lesion; LU: Ulcerated lesion; LB: Budding lesion; LC: Crusting lesion; AL: Other types of lesions.

3.3. Histopathological Aspects

Cancerous lesions accounted for 65.3% and precancerous lesions (actinic keratosis) for 14.7%. Among the cancerous lesions, invasive SCCs represented 50.3%, in situ SCCs 6.6%, and BCCs 43.1%.

3.4. Therapeutic Aspects

Excisional biopsy under local anesthesia was performed (92.2%), simple biopsy (4.3%), and excision under general anesthesia (3.5%). Excisional margins ranged from 2 to 6 mm (2 to 3 mm for BCCs and 5 to 6 mm for SCCs).

Depending on the topography and extent of the lesions, some lesions required simple suture, directed healing, local flap (**Figure 3**), locoregional flap, or skin graft (**Figure 4**). A loss of nasal substance (**Figure 5**) after surgical excision was noted.

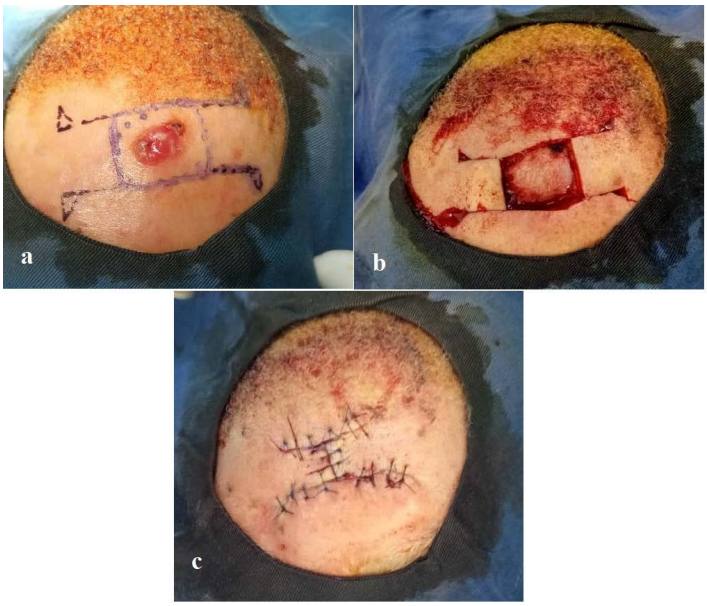

Figure 3. Frontal nodular lesion (a) resected (b) with local H-shaped flap closure (c).

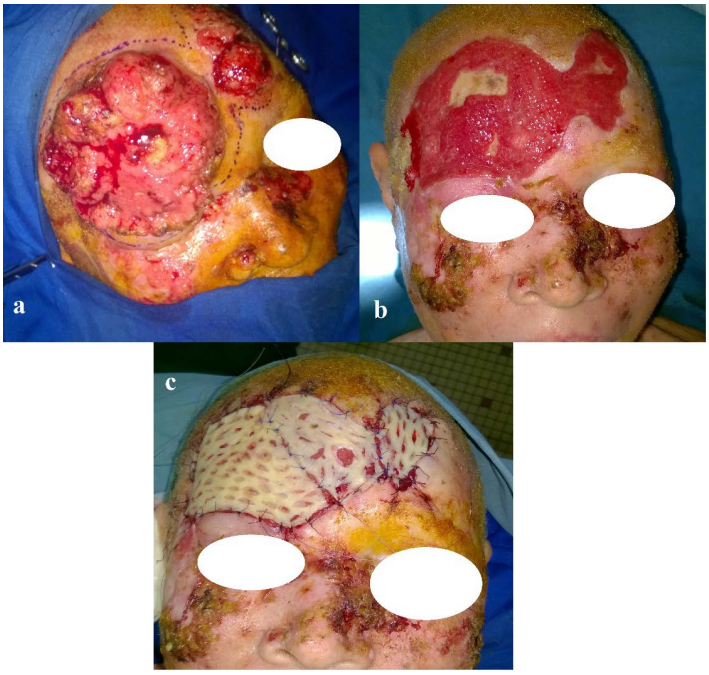

Figure 4. Frontal nodular budding and ulcerated lesion (a) resected and treated with skin graft (c).

Figure 5. Budding and crusting lesion of the nasal pyramid (a), amputation of the nasal pyramid (b).

4. Discussion

This study mapped cancerous and precancerous lesions of the head and neck in PWA operated on in Togo.

In this study, the head and neck (66.1%), particularly the face, was the predominant site of cancerous and precancerous skin lesions in PWA. Our results are consistent with those of other authors, particularly in Africa. In Kenya, a country located on both sides of the equator, Emadi *et al*. (2017) found 45.32% [7]. In Nigeria, Awé *et al*. (2018) found 63.6% of cancerous lesions to be located on the face and neck [14]. In Tanzania, in the study by Kiprono *et al*. (2014) involvement of the head and neck accounted for 56% [18]. In Mali, Gassama *et al*. (2021) reported a 70% incidence in a series of 20 cases [17]. Since UV exposure is very

detrimental to hypopigmented skin, the parts of the body most exposed to the sun, such as the face, ears, neck, and back of the hand, are the preferred sites for these lesions [1] [4] [7] [12] [15]. Moreover, the study by Kromberg *et al*. (1986) highlighted a higher frequency of skin lesions of the head and neck in albinos (81%) than in the general population (60%) [10].

Our study reveals a male predominance of skin lesions of the head and neck. This is consistent with the study by Gassama *et al*. [17]. This male predominance may be explained by negligence in observing photoprotection measures. The study by Lookingbill *et al*. (1995) noted non-compliance with photoprotection measures in men with the wearing of baseball hats [8]. Photoprotection of the head and neck essentially includes the wearing of wide-brimmed hats and the use of anti-UV sunscreen topicals [4]. The male predominance in therapeutic non-compliance in dermatology was reported by Baliou *et al*. (2012) [19].

The mean age in this study was 32.64 ± 14.64 years in agreement with the data in the literature [7] [12] [15] [18] [20]. This age appears earlier than the average age of onset of cancerous and precancerous skin lesions in the non-albino population [8] [10] [12]. Compliance with photoprotection measures is less rigorous in children and adolescents, which exposes them more to UV rays, with an earlier onset of skin lesions [4] [19] [21] [22]. Most of the patients in our sample were students. They are subject to daily exposure to UV rays in a relatively hot region like Togo. Furthermore, compliance with photoprotection measures requires financial resources. The ANAT (National Association of Albinos of Togo) occasionally distributes them free of charge during these awareness campaigns. However, these occasions are rare because they are subject to donations from various partners. The impact of compliance with photoprotection measures on reducing the appearance of skin lesions through therapeutic education of PWA has been demonstrated by several studies [16] [23].

In this study, the auriculotemporal region was the preferred site of skin lesions, followed by the occulopalpebral region and the frontal region. These results are in agreement with those of Gassama *et al*. (2021) [17]. However, the study by Gbéry *et al*. (2012) had shown a higher frontal localization of skin lesions [24]. The anatomical situation of this area requires a certain dexterity and meticulousness in the application of sunscreen creams. Histopathologically, cancerous lesions were predominant in the head and neck in our study, in agreement with several African studies [17] [24]-[26]. Exposure to UV rays remains a determining factor in the evolution of lesions towards a malignant process [6] [12]. Keratinocytes of basal progenitor cells in sun-exposed skin are at high risk of undergoing malignant transformation induced by sunlight. SCCs in albinos can appear immediately or from pre-malignant actinic lesions, in which keratinocytes have already undergone an initial transformation induced by sunlight. Keratinocytes of basal cells undergo DNA damage of varying degrees of severity depending on the intensity and duration of sun exposure [26]. Since the head and neck are more exposed to UV rays, it remains a preferred site for skin cancers in albinos [7]. Among skin cancers, squamous cell carcinomas were the most common, which is consistent with the results of several African studies, particularly in Mali, Ivory Coast, Kenya, and Nigeria [7] [17] [18] [21]

[24] [26]. However, the predominance of basal cell carcinomas has been reported in Brazil and Togo [11] [20]. These studies considered whole-body skin cancers.

Surgical excision of cancerous and precancerous skin lesions in albinos is the first-line option of choice [4] [9] [13] [15] [17] [24]. It was performed in almost all cases under local anesthesia with a margin of 2 to 6 mm in this study. The surgical techniques used in our study are consistent with the literature [25].

5. Limitations

The small number of lesions in certain regions did not allow us to identify the link between location and type of lesion. Our study had limitations due to the retrospective nature of any study, including the incompleteness of medical records and missing tracking data, which reduced the sample size. The lack of data (numbers) on surgical techniques is due to the retrospective nature of the study. The possible underrepresentation of lesions not treated surgically and the retrospective design can also explain the small size of our sample.

6. Conclusions

The head, particularly the face, an area more exposed to UV rays, remains the preferred region for cancerous and precancerous lesions in albinos.

Since therapeutic dermatology education is a pillar of therapeutic success, raising awareness and, above all, compliance with photoprotection measures, with emphasis on the auriculotemporal, frontal, and occulopalpebral regions, could significantly reduce skin cancers in people with albinism. An evaluation of this compliance would be useful to confirm the data from this study and further refine awareness.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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