

Case Report

Malignant skin tumors in patient with Oculo cutaneous AlbinismManmeet Kaur¹, Kanwardeep Kaur Tiwana², Sarita Nibhoria³, Nisha Singla⁴¹Assistant Professor (GGSMC & H, Faridkot), ²Associate Professor (GGSMC & H, Faridkot), ³Professor (GGSMC & H, Faridkot), ⁴Junior Resident (GGSMC & H, Faridkot)***Corresponding author**

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Abstract: Although squamous cell carcinoma are more in these patients presence of recurrent squamous cell carcinoma and basal cell carcinoma at different intervals are rare. Albinism is a genetic disorder characterized by lack of skin pigmentation. As a consequence these persons lack the protective effect of melanin against ultraviolet radiation damage. Oculo cutaneous albinism is an established risk factor for all the three major forms of skin cancers – Squamous cell carcinoma, Basal cell carcinoma and malignant melanoma. Simultaneous occurrence of Squamous cell carcinoma and Basal cell carcinoma is rare. This report highlights the metachronous presence of Squamous cell carcinoma and Basal cell carcinoma.

Keywords: Oculo cutaneous albinism, squamous cell carcinoma, basal cell carcinoma

INTRODUCTION

Albinism is a genetically inherited autosomal recessive disorder characterized by hypopigmentation of the skin, hair and eyes due to a reduced or lack of cutaneous melanin pigment production[1]. Albinism is broadly divided into two types: ocular albinism affecting eyes only and oculo cutaneous albinism affecting eyes, skin and hair[2]. Albinism has a worldwide distribution, but is said to be commoner in regions of the world closer to the equator, with greater penetration of the sun's ultraviolet radiation. It has an estimated frequency of 1 in 20000 in most populations with the highest incidence of 6.3 per 1000 reported among the Cuna Indians [3]. Albinism most commonly arises from mutations in genes encoding for proteins involved in the synthesis or transport of melanin by melanocytes [3]. Melanin is a photo protective pigment, protecting the skin from the harmful effects of ultraviolet radiation. The ocular problems faced by albinos are nystagmus, strabismus, photophobia, foveal hypoplasia and decreased visual acuity. The cutaneous problems seen with oculo cutaneous albinism are sun burns, basal cell carcinoma, malignant melanoma, dysplastic nevus syndrome and, most important and most common of all, actinic keratosis predisposing to squamous cell carcinoma [4].

CASE REPORT:

A 50 years, old male, who is a known case of albinism since birth present with multiple lesions on

face, first time in 2007 and was diagnosed as squamous cell carcinoma. 25 cycles of radiotherapy was given. Patient also gave the history of photosensitivity. After 3 years patient had developed multiple lesions on scalp, these lesions were excised and histopathological examination was done and diagnosed as poorly differentiated squamous cell carcinoma. After 1 year in 2011, patient presented with skin lesions near the right canthus of eye, histopathological examination showed the development of basal cell carcinoma. Chemotherapy was given. In 2012, biopsy was done from lesion on right temporal region and was diagnosed as basal cell carcinoma. Later after 1 year, patient again had developed lesions on right and left side of face, left preauricular region and was diagnosed as basal cell carcinoma. 2 year later, patient was diagnosed as squamous cell carcinoma in the neck lesion. Now, the patient presented with lesion on left side of neck, post auricular region and right thigh since 1 year. Excision was done and histopathological examination was done in pathology department, GGS & MC faridkot.

PATHOLOGICAL FINDINGS

Gross Examination: Received four skin covered soft tissue pieces labelled lesion on left side of neck measuring 3x1.5x0.3cm, post auricular lesion measuring 4x3.5x0.5cm with hypo pigmented raised area measuring 2x1.5cm, first lesion on right thigh measuring 4x1.5x0.2cm with hypo pigmented raised

area measuring 0.5x0.5cm and second lesion on thigh measuring 2x1x0.5cm with hyperpigmented raised area

measuring 0.3x0.2cm.



Fig 1: Albinism with white hairs and skin.

Microscopic Examination: Histological appearances from thigh lesion and post auricular lesion are suggestive of squamous cell carcinoma and from lesion

on left side neck is suggestive of Pseudo epitheliomatous hyperplasia.

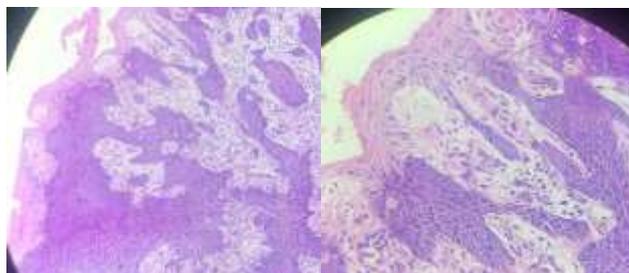


Fig 2: Squamous cell carcinoma

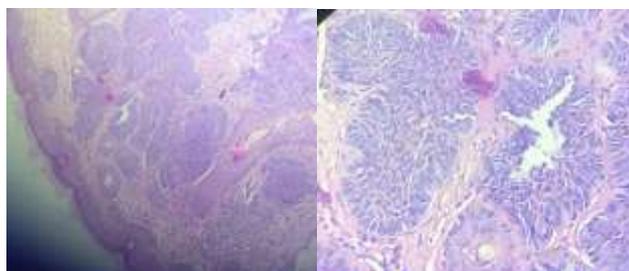


Fig 3: Basal cell carcinoma



Fig 4: Pseudo epitheliomatous hyperplasia.

DISCUSSION

Actinic keratosis was first described by Dubreuilh in 1904. The factors implicated in development of actinic keratoses are UV rays, fair skin, blue eyes, blond hair, immuno suppression and genetic

diseases like albinism and Xeroderma Pigmentosum [5]. Skin cancers in albinos accounted for 13.2% of total skin cancers [6]. In albinos there is a defect in the synthesis of tyrosinase, which catalyses hydroxylation of the melanin precursor tyrosine to

dioxyphenylalanine. As a consequence these persons lack the protective effect of melanin against ultraviolet radiation damage [7]. For the darker races, where melanin confers natural photo-protection, albinism poses as an established risk attribute for all three forms of cutaneous malignancies. Although squamous cell carcinoma (SCC) is usual, basal cell carcinomas have also been reported in albino [8]. Studies have shown that upto 60% of the squamous cell carcinomas begin as actinic keratoses and that there is histologic evidence of contiguous actinic keratoses in 97% of the squamous cell carcinoma lesions that arise on sun damaged skin [9]. Another fascinating entity that has emerged in recent times is baso-squamous cell carcinoma, where coexisting features of both SCC and BCC have been encountered.[10] Our case is a 50 years old male, who is a known case of albinism since birth now presented with lesion on left side of neck, post auricular region and right thigh since 1 year and is diagnosed as squamous cell carcinoma from thigh and post auricular lesion and Pseudo epitheliomatous hyperplasia from left side neck lesion.

CONCLUSION

Oculo cutaneous albinism is an established risk factor for all the three major forms of skin cancers, due to reduction and/or complete absence of melanin. Although SCC is common, the presence of BCC is not that infrequent and should be suspected.

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