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Case Report

Basal cell carcinoma in oculo-cutaneous albinism

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ABSTRACT

The basal cell carcinoma is the most common skin tumour especially affecting the white individuals worldwide. The exact incidence of basal cell carcinoma is not known from India but non melanoma skin cancers comprises about 1-2% of cutaneous tumour in India. The most common skin tumour is squamous cell carcinoma in albinism and the incidence of basal cell carcinoma is less. Hereby, we report a peculiar case of basal cell carcinoma in albinism to highlights the importance of early recognition and diagnosis of suspected lesions by performing histopathological examination in unusual circumstances.

Keywords: Basal cell carcinoma, Oculo-cutaneous albinism, Histopathological examination

INTRODUCTION

Basal cell carcinoma (BCC) is the most common cutaneous malignancy of humans arises from the cell similar to those of basal area of the epidermis and its appendages. It was first described in 1984 by Jacob.[1] It is the most common cutaneous tumour of white individuals and as albinism skin behaves as type 1 skin phenotype, so its occurrence in such skin deserves a special consideration. BCC contributes to 65-75% in whites and 20-30% in Asian Indians.[2] The intermittent, intense and infrequent exposure to ultraviolent radiations particularly ultraviolent B plays a major role in the causation of BCC unlike to squamous cell carcinoma.[3,4] It is a slow growing, locally invasive tumour that rarely metastasizes.

BCC is relatively found more in genodermatosis like oculo-cutaneous albinism (OCA), hermansky-pudlak syndrome, rombo syndrome, xenoderma pigmentosa, gorlin syndrome and bazex-dupre-christol syndrome.[5] The complete absence or reduction of photo protective melanin in these conditions may lead to increase susceptibility for the development of basal cell carcinoma. BCC is known to occur at specific sites at head and neck, most frequently above the line joining the angle of mouth and the tragus of ear, still the exact reason is not clear till today. It has also been reported from unusual sites namely scrotum, perianal, vulva, nipple and periungual areas.

Here, we report a case of basal cell carcinoma on the uncommon site in type-1 oculo-cutaneous albinism patient.

CASE REPORT

A 45 years old male, a case of type 1A oculo-cutaneous albinism, presented with a pea sized nodule over the lower back for the past 5 years. The lesion slowly progressed from pin head to pea size since then, with history of ulceration and bleeding on and off. There was no history of spontaneous regression. The patient had multiple lesions of actinic keratosis over the body (Figure 1). Some of the lesions become hard, darker and elevated above the skin surface with increasing age.

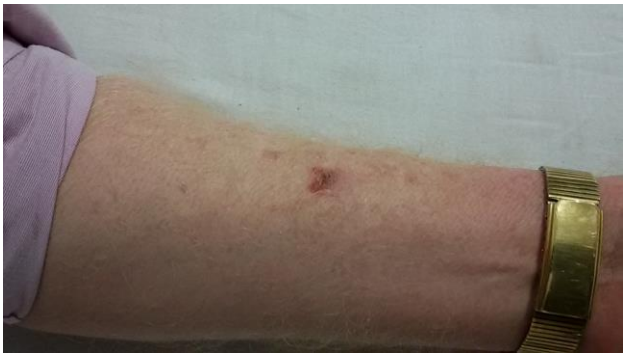

Figure 1: Actinic keratosis over the volar aspect of the left forearm.

The patient had history of photosensitivity, sun burning episodes, low vision, horizontal nystagmus, white hairs and chalky white skin. There was no history suggestive of neurological abnormality, bleeding tendency and recurrent infections. They were five brothers and two had type 1 OCA and one sister also had type 1A OCA. No history of consanguineous marriage in the family.

On local examination, over the lower back, there was well defined erythematous, indurated nodulo plaque with rolled out margins of size approximately 2x1.5x1.5 centimeters showing central ulceration with perilesional erythematous halo (Figure 2).

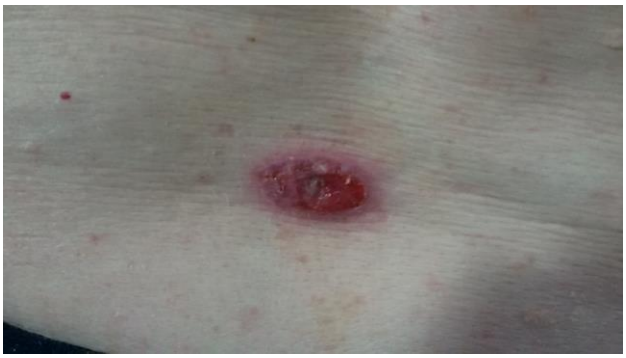

Figure 2: Well defined erythematous nodulo plaque over the lower back.

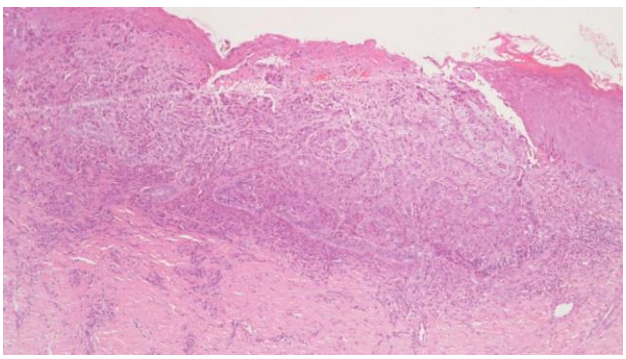

Figure 3: HPE showing basal cell carcinoma with basaloid cells, peripheral palisading with retraction artefacts.

The general physical and systemic examination was normal. No abnormality was detected in the routine blood investigations and radiological examination. The punch biopsy from lesion over the lower back was taken and histopathological report revealed ulcerated epidermis and the dermis was infiltrated by variably sized nodular/ basaloid cell showing peripheral palisading and cleft between the fibrotic epithelium suggestive of basal cell carcinoma (Figure 3).

DISCUSSION

The main factor determining the risk for skin cancer is the cutaneous pigmentation and the protective tanning ability in response to ultra violet radiation exposure which is in turn is controlled by number of genes or gene variants. The protective effects of pigmentation in darkly/intensely pigmented skin are underlined by the low occurrence of BCC in darkly pigmented skin.[6] The high risk patients are fair skinned with poor tanning ability and history of sun burning on sun exposure.[7] The albinism poses as an established risk for the development of all the three forms of cutaneous malignancies (actinic keratosis, squamous and basal cell carcinoma).[8]

As actinic keratosis is regarded as the precursor lesion for the squamous cell carcinoma (SCC), although the rate of progression to invasive SCC is low and usual occurrence in the albino skin. The cutaneous features of photo damage and morphological features favour the diagnosis of SCC, but histopathological report confirmed it as BCC. This case highlights the occurrence of basal cell carcinoma at unusual site with signs of photodamage and actinic keratosis, in the perspective of type-1 oculo-cutaneous albinism.

CONCLUSION

The white skin (type-1) phenotype and the complete absence or reduction of melanin in oculo-cutaneous albinism emphasizes the role of lifelong photo protective behaviour in decreasing the photo damage and prevention of cutaneous malignancies. The basal cell carcinoma should always be suspected in the background of cutaneous sign of photo damage and irrespective of clinical history, morphology and unusual site. The histopathological examination is crucial for correct diagnosis and early treatment.

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