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CLINICAL REPORT

Surgical Excision with Rotational Flap Reconstruction for Basosquamous Cell Carcinoma in Xeroderma Pigmentosa: A Case Report

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Abstract Xeroderma pigmentosa (XP) is a rare autosomal recessive disorder which is characterized by a defect in nucleotide excision repair of DNA following exposure to UV radiation. This leads to hypersensitivity to sunlight causing pigmented skin lesions, photophobia and a 1000-fold increase in risk of developing cutaneous malignancies like basal cell carcinomas, squamous cell carcinomas and melanomas of head and neck. We present an interesting case of a 28 year old man with basosquamous cell carcinoma secondary to XP treated successfully with surgical excision and reconstruction with rotational flap technique.

Keywords Rotational flap · Basal cell carcinoma · Xeroderma pigmentosa

Introduction

Xeroderma pigmentosa was first described by Moriz Kaposi [1]. It is a rare autosomal recessive disease with a prevalence rate of 1:250,000 in United States and 1:40,000 in Japan and can occur in all kinds of ethnic groups and races all over the world [2]. Most of the cases reported in Indian literature show evidence of Basal cell carcinoma being associated with Xeroderma pigmentosa [3, 4].

The most common skin cancer worldwide is Basal cell carcinoma (BCC) [5]. It is a locally invasive, slowly growing, non aggressive tumour arising from the basal layer of epidermis [6]. Around 75% of non-melanoma skin cancers include basal cell carcinoma. It occurs mostly in elderly males on face particularly in nasal area (25.5%) who are exposed to ultraviolet radiation [7]. Treatment modalities for Basal cell carcinoma (BCC) include surgical excision with reconstruction, radiotherapy, cryotherapy, electrodessication and curettage [8]. Local application of 5-fluorouracil (5-FU) ointment, sunscreen lotions and oral isotretinoin are other conservative methods [9]. Well defined basal cell carcinoma requires surgical excision with 2 mm wide margins to achieve 95% cure rate [10].

This paper discusses an interesting method in managing Basal cell carcinoma by excising and reconstructing the defect with rotational flap.

Case Report

A 28 year old male diagnosed with Xeroderma pigmentosa 15 years back presented to our outpatient clinic with a non healing ulcers over both sides nose and watering of eyes upon exposure to sunlight.

On examination he had dry atrophic skin with generalized freckles and multiple hyperpigmented macules distributed all over the face, trunk and extremities. A brownish black, circular lesion of 1 * 1 cm was present over the right side of nose (Fig. 1a) and a 1.5 * 1 cm circular ulcerated lesion with crusting and telangiectasia was present over the left side of nose (Fig. 1b). Conjunctiva was congested but cornea was non hazy. At the first visit patient had already received conservative management with multiple antibiotic creams, sunstop 19 lotion, oral isotretinoin for 6 weeks, and a surgery for left eyelid entropion. Systemic examination revealed no neurological defects. There was no history of radiation or chemotherapy.

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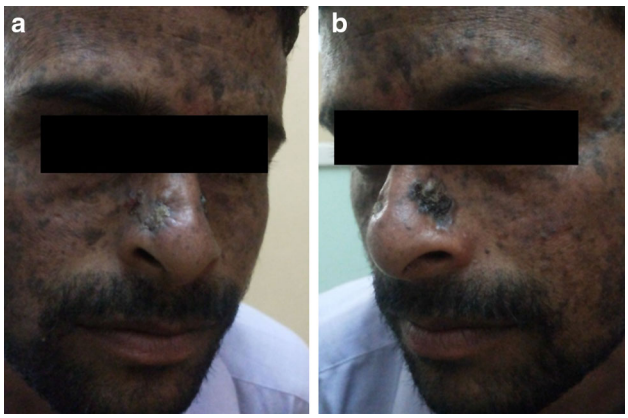

Fig. 1 Pre op photo with generalized freckles and hyperpigmented macules. Note the brownish black, circular ulcerated lesions on both sides of nose

Haemogram, biochemical studies and chest radiography were within normal limits.

Surgical excision with reconstruction was done on the right side of the nose and biopsy revealed basosquamous variant of basal cell carcinoma (Fig. 2). Patient was advised for surgical excision of tumour on the left side of nose but had refused for the procedure. 6 months later he again presented to the clinic with the unoperated lesion on the left side which has now increased to the size of 2.5 * 1 cm. Surgical excision of the tumour with wide margin of 2 mm was done and the defect was reconstructed with rotational flap harvested from left nasolabial fold (Fig. 3a, b) and biopsy confirmed pigmented basal cell carcinoma (Fig. 4). Skin edges were sutured with prolene 5.0 No adverse events occurred during the surgery and the entire tumor regressed at the end of 6-months (Fig. 5a, b).

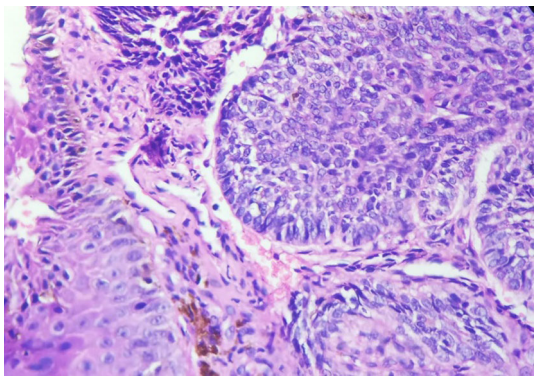

Fig. 2 Biospy of the Right sided lesion shows tissue lined by stratified squamous epithelium with round to oval tumor cells seen in sub-epithelium arranged in sheets and nests with containing hyperchromatic nuclei, moderate eosinophilic cytoplasm, peripheral palisading and retraction artifact. Mitotic cells 1–2/10HPF

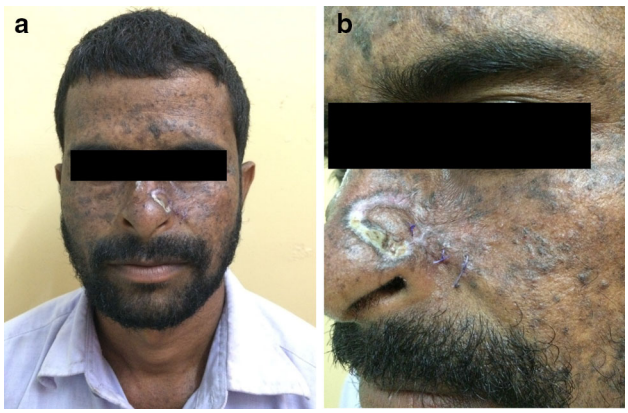

Fig. 3 Post op 8th day with surgical excision and rotational flap reconstruction of the left sided lesion

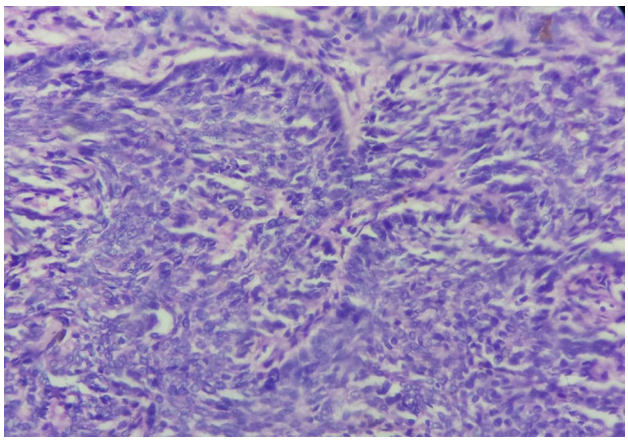

Fig. 4 Biopsy of left sided lesion showing tissue arranged in solid nests with peripheral palisading and retraction artifact. Individual tumor cells are pleomorphic with round to oval hyperchromatic nucleus and irregular nuclear membrane with scant cytoplasm. Melanin pigment incontinence at dermoepidermal junction seen

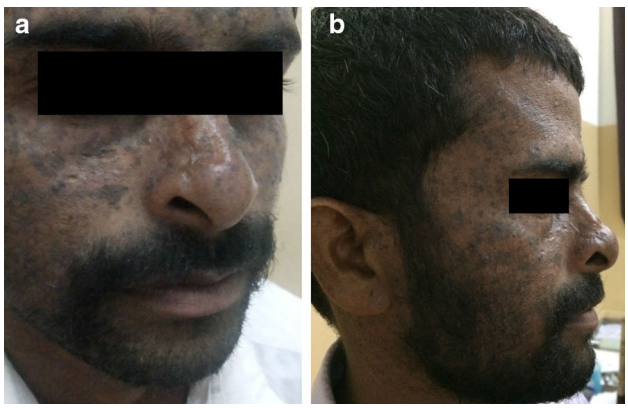

Fig. 5 6 months follow up picture after Right sided excision biopsy and reconstruction with rotational flap. Note that the margins are well healed and structural intricacy is maintained

Discussion

Xeroderma pigmentosa (XP) is an autosomal recessive disease characterized by inability to repair the DNA damage caused by UV radiation [11]. There are 10 genetic variations of XP. In one variant there is defect in the DNA repair post replication (XP variation) while the other nine variations are defective in excision repair (XP group A-I) [12]. Accumulation of damaged DNA causes chromosomal mutations, which results in cell death and neoplastic abnormalities. It causes a 1000-fold increase in skin cancers in homozygous individuals [13]. This genetic condition has been observed to be related with mutations in two tumor suppressor genes namely p53 and patched (PTCH). Mutations in (PTCH) gene are more aggressive in causing basal cell carcinoma [14].

Patients of Xeroderma pigmentosa with BCC require early diagnosis and excision of tumor as some of them behave aggressively particularly in the facial region. About 33% of all inadequate BCC excisions are found in the nasal area. The reoccurrence risk of BCC is 2.5 times higher in the nasal area after surgical excision [15]. Nevertheless, the cornerstone treatment for BCC of the nose is surgical excision with reconstruction ideally with rotational flaps because of close proximity of the flap to the defect, rapidity and quickness of method and angle of its rotation which makes it easier to reconstruct the defective margins resulting in excellent cosmetic result. Rotational flaps give the capacity to mobilise particular zone of tissue especially in the nasal area as they have excellent blood supply and good base for reconstruction [16].

A rotational flap is a crescent shaped or semicircular adjacent skin flap that is rotated into the deformity on a supportive fulcrum point. Rotational flap is harvested from the nasolabial or paranasal skin and rotated 90° with respect to the defect created after the surgical excision of the tumour. This rotational alar flap forms a redundant circular zone and recreates a "natural" nasal crease, by safeguarding the normal anatomical structures [17].

For a better result it is very important to differentiate between the histological variations and have good knowledge about the anatomic location of the tumour. Location on the nose is additionally viewed as an element of high-risk BCC due its anatomical characteristics and difficulty in precisely identifying the tumour margins in pre surgical evaluation. Proper preoperative counseling should be given to the patients regarding wound care and their final appearance after reconstruction.

As physiological functions of the nose play a key role in daily life, it is critical that the facial deformity reconstruction safeguards the integrity of facial expressions and symmetry. While planning for excision and reconstructive surgery, a specialist should precisely consider various attributes remarkable to the nose, including the size, depth and location of lesion, structural intricacy of the nose with arched and curved surfaces lying close to each other, the sebaceous structures and the constrained laxity of the skin [18]. The anatomic and physiological function of the nose must be preserved by safeguarding the bony and cartilaginous system and the mucosal covering without compromising the airway of the patient.

Conclusion

To conclude the main aim is to surgically remove the tumor with wide marginal excision and reconstruct the deformity with minimal scarring to achieve a better cosmetic result. The best reconstructive efforts fail when tumor reoccurs in the facial region. Rotational flap is one of the best approaches for reconstruction of facial defects because it helps in histological control of the lesion borders, decreases the traction arising from the borders of the surgical defect, provides efficient blood supply and gives good cosmetic result [19].

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