

Squamous Cell Carcinoma in Epidermolysis bullosa (dystrophic)

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Abstract Epidermolysis bullosa are heterogeneous group of rare diseases characterized by exacerbated skin and mucosal fragility and blister formation after minor trauma. This family of genodermatosis exposes patients to several complications, Cutaneous SCC being most severe. We present a case of 19 years old patient of Squamous cell carcinoma in Epidermolysis bullosa of right foot treated with surgery and radiotherapy.

Key words

Human papilloma Virus (HPV), Epidermolysis Bullosa(EB), Squamous cell carcinoma(SCC).

Introduction

Epidermolysis bullosa (EB) is a inherited heterogeneous group of skin disorders associated with widespread blisters, erosions and chronic wounds. Inherited EB is classified into three main subtypes: simplex, junctional and dystrophic, based on ultrastructural levels of skin cleavage. The epithelial fragility occurs at or close to the dermal–epidermal junction.¹ Over the past 10 years research has led to the identification and characterization of the genes for many of the structural proteins that secure adhesion between the epidermis and the underlying dermis. In addition to an increased understanding of the molecular basis of EB, recent years have also seen substantial clinical advances in patient management. However, one of the major clinical issues still afflicting EB sufferers is the complication of cutaneous squamous cell carcinoma (SCC).

Squamous cell carcinoma is a malignant tumor arises from keratinocytes of epidermis and contiguous mucous membranes. Risk factors are damaged skin, chronic exposure to sunlight, Fitzpatrick's skin type 1 and 2, scar, nonhealing ulcers, chronic granulomas and HPV. Common sites are photo exposed areas e.g. head, face, distal limbs and pina.²

Case report

A 19-years old male patient, known case of Epidermolysis bullosa dystrophic presented with non-healing ulcer on lateral side of right foot for 4 years. According to the patient, it was started as small growth which was increasing in size, initially it was painless then became very painful. It was not associated with itching, joint pain, fever and weight loss. Family history was insignificant. Patient had history of recurrent nail and dental infection (**Figure 1A,1B**). O/E A fungating ulcer on lateral side of right foot about 4cmx4cm in size, having well defined erythematous margins with serous discharge surrounded by hypopigmented atrophic skin and hyperpigmented at upper side (**Figure 2**).

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Figure 1 (A) Hypopigmented atrophic skin and hyperpigmented nails.



Figure 1 (B) Widened spaced teeth.



Figure 2 Ulcer on right foot.

Discussion

Non Melanoma skin cancer are most common tumors with SCC accounts for 1/4th cases. The cumulative risk of developing SCC in EB increases with age: for patients with RDEB-SG, the cumulative risk of having at least one SCC is

7.5% at age 20 years, 67.8% at 35 years and 90.1% by 55 years.¹ Clinical detection of SCCs In patients with EB on a background of chronic ulceration may be particularly challenging and therefore the possibility of malignancy should be borne in mind, with suspicious lesions biopsied for histological evaluation. SCC are typically well differentiated but they frequently show rapid growth and metastatic with an overall poor prognosis.¹⁻² In contrast to UV-induced SCCs, they are mostly located on the distal extremities.³ Unlike SCC occurs in general population due to chronic sun exposure, EB associated SCC arises at the site of blisters, scarring and wound formation. It behaves more aggressively than conventional.⁴⁻⁵ SCC's and is the most common cause of death.⁵ Treatment options includes Surgical excision, minimally invasive surgical techniques (Moh's micrographic surgery) rush paraffin sections ('slow Mohs'), chemosurgery, radiotherapy or chemotherapy. Topical modalities includes photodynamic therapy (with 5-aminolaevulinic acid).⁶

Chemotherapy agents include cisplatin, carboplatin, fluorouracil, doxorubicin and methotrexate. Surgical excision is the standard treatment for EB induced Squamous Cell Carcinoma.⁷

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