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SQUAMOUS CELL CARCINOMA OF LEFT HAND IN A PATIENT WITH EPIDERMOLYSIS BULLOSA DYSTROPHICA- A CASE REPORT

General Surgery

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KEYWORDS

INTRODUCTION :

Epidermolysis bullosa (EB) refers to a group of hereditary mechano-bullous disorders ,most of them are associated with chronic scarring. Several forms of this disease have been reported with cutaneous malignancy. Squamous cell carcinoma is a well recognised complication that develops from various cicatrising and ulcerative dermatoses. We report a case of Squamous cell carcinoma in a patient with Epidermolysis bullosa dystrophica.

CASE REPORT :

Mr. Thoufiq , a 32 year old male,who is a salesman came to our hospital with complaints of swelling in the left hand for the past 1 year. The swelling was insidious in onset , progressive in nature, associated with severe pain, not associated with any discharge. The patient is a known case of Epidermolysis Bullosa Dystrophica since childhood . On Examination, the patient was poorly built and nourished. Pallor was present. Fragile skin that peels off and blisters easily was seen all over the body especially over the back .Hand and Foot deformity was seen .Fingers and toes were underdeveloped and fused . Alopecia was present. On Local Examination of Left Hand , Ulceroproliferative growth of 6*5 cm was seen over the dorsal aspect , irregular in shape , irregular surface, well defined margins . The swelling was not mobile .Bleeding on touch and slough were present . MRI showed ulcerative soft tissue density thickening measuring 9 mm with ? malignant transformation of the lesion . Underlying bones were normal . Edge wedge biposy revealed Well differentiated Squamous cell carcinoma . His hemoglobin was 6 gm % . A series of blood transfusion was done till his Hb level reached 10 gm% . His cardiac status was normal . Anaesthesia fitness was obtained for surgery . Wide local excision was not feasible in this case as adequate margin could not be obtained . Hence, Below Elbow amputation was done and adequate proximal margin clearance and healthy flap closure was achieved . Post operative period was uneventful . He was treated with antibiotics, analgesics and other supportive medications . Patient was discharged on 5th post operative day and suture removal was done after two weeks . Histopathology revealed Well differentiated Squamous Cell Carcinoma (Low Grade). As it is a low grade tumour , and keeping in mind the general condition of the patient , Chemotherapy was not given post-operatively and the patient was advised regarding regular follow-up and chances of recurrence .

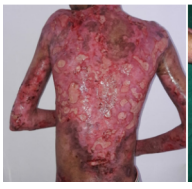

Figure (A)

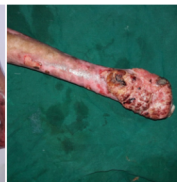

Figure (B)

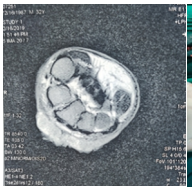

Figure (C)

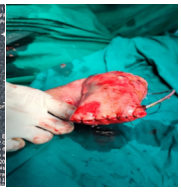

Figure (D)

Figure(A)- Epidermolysis Bullosa Skin lesions (Back), Figure(B) - Ulceroproliferative growth over Left hand , Figure(C)-MRI showing Soft tissue lesion ,underlying bones appear normal , Figure(D)-Below Elbow amputation Post-op pic showing healthy flap .

DISCUSSION :

Skin or mucous membrane malignancies are a rare but nevertheless significant complication of EB. . The majority of neoplasms are SCC occurring in cases of generaliscd recessive dystrophic EB. Other tumours reported include basal cell carcinomas , giant keratoacanthomas , Bowen's disease, and Kaposi's sarcoma. The peak age of onset for SCC in generalised recessive dystrophic EB is during the 20s and 30s compared to during the 50s for more localised forms of EB. Most of the reported histology is of well or moderately differentiated SCC. In comparison with other scarring dermatoses, it is uncertain why EB patients develop SCC. Studies to investigate growth activation and differentiation characteristics in epidermal keratinocytes in cases of recessive dystrophic EB (using immunohistochemical psi-3 expression) have shown increased growth-activated phenotype in keratinocytes of previously wounded skin that had been healed . It is possible that epidermis in this chronic growth activation state which subsequently fails to differentiate normally may be predisposed to future neoplastic change. In addition, a finding of defective innmune surveillance (and therefore an increased likelihood of neoplastic proliferation) based upon a reduced activity of circulating natural killer cell activity in patients with recessive dystrophic EB has also been reported . A possible explanation for the high occurrence of SCC in EB has been proposed by Goldberg et al. based upon a theory of "tissue stress" whereby the chronic processes of skin ulceration and tissue repair leads to a loss of cellular memory and a progressive lessening of differentiation in the region and persistent stimulus of wound healing is then directed towards tumour promotion. Early recognition of occult SCC is the most important aspect in management , since cutaneous examination of most patients shows evidence of a progressive range of keratinocyte de-differentiation. Assessment of a tumour or dysplastic cell-associated protein would be a potentially useful diagnostic tool, especially if its presence formed the basis for a cytological test to distinguish the chronic ulcers and skin crusting lesions that typify recessive dystrophic EB patients from some stage in the often insidious progression towards malignancy. It has been stated that the prognosis of even well differentiated SCC complicating dystrophic EB is poor, possibly due to a decreased structural resistance to the expanding tumour in the presence of abnormal basement membrane collagens or fibroblast-derived collagenases. It is doubtful whether more radical surgery with wider excision and lymph node clearance would add any benefit, particularly as the nature of dystrophic EB means that many of these patients already have severe physical constraints and loss of limb function.

CONCLUSION:

SCC is a significant threat to EB patients, especially those with generalised recessive dystrophic EB. At present there is no easily available clinical method to distinguish cutaneous SCC from non-neoplastic ulceration, hypertrophic granulation tissue, cutaneous horn-like lesions, or hyperkeratotic crusting, therefore, early and repetitive sampling of possible suspicious lesions for histological assessment

should be undertaken if metastatic disease is at all to be prevented.

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