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Case Report

Paraneoplastic pemphigus: a rare case

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ABSTRACT

Paraneoplastic pemphigus is an autoimmune disorder characterized by recalcitrant blistering, erosive mucocutaneous lesions, associated neoplasm and high titres of "pemphigus-like" anti-cell surface autoantibodies. A 55-years-old male presented to the OPD with fluid filled lesions over the chin region and few crusted lesions over the chin and chest since 10 days. There was a history of carcinoma tongue 6months back for which patient underwent 33 sessions of radiotherapy and chemotherapy. Various investigations, including hemogram, serum biochemistry, thyroid profile, lipid profile, viral markers, serum electrophoresis and histopathological examination, were conducted. Histopathology showed intraepidermal blister formation and DIF showed immune deposits in a linear deposition at DEJ and in a fishnet pattern over keratinised surface. Based on history and thorough examination, the case was diagnosed as Paraneoplastic Pemphigus. Patient was treated with oral corticosteroids, oral and topical antibiotics and complete resolution of all skin lesions and with residual pigmentary changes was seen after 4 weeks.

Keywords: Paraneoplastic pemphigus

INTRODUCTION

Paraneoplastic pemphigus is an autoimmune disorder characterized by recalcitrant blistering, erosive mucocutaneous lesions, associated neoplasm and high titres of "pemphigus-like" anti-cell surface autoantibodies.[1]

Cutaneous lesions may be polymorphous, resembling those of Pemphigus vulgaris, bullous pemphigoid, erythema multiforme, graft-versus-host disease or lichen planus.[2] We report a 55-years-old male who developed bullous lesions over head and neck region following treatment for carcinoma of tongue.

CASE REPORT

A 55-years-old male presented to the OPD with fluid filled lesions over the chin region and few crusted lesions over the chin and chest since, 10 days. He was apparently normal 10 days back when he developed fluid filled lesions initially over the chin, neck and upper trunk which later turned into crusts after 4 days. There was a history of carcinoma tongue 6 months back for which the patient underwent 33 sessions of radiotherapy and chemotherapy.

There was no history of weight loss, no history of trauma, no history of insect bite, no history of hemoptysis, no history of drug intake prior to the onset of lesions and no history of lesions over other areas of the body. Family history was non-contributory. Personal history was not significant.

On general examination no anemia, icterus, cyanosis, clubbing, pedal edema or lymphadenopathy were seen. On systemic examination cardiovascular system, respiratory system, central nervous system and per abdominal examination were normal. On Dermatological examination, eroded plaques with crust formation over surface was seen over the chin region and left side of the beard region (Figure 1). Two discrete crusted plaques

with haemorrhagic crusting were seen over the mandibular region (Figure 2) and similar erosion with crust formation were seen over the upper chest (Figure 3). Healed hypopigmented patch was seen over the retro auricular and infra-auricular area over the left side. Based on histopathology and clinical features a differential diagnosis of paraneoplastic pemphigus, Bullous pemphigoid and pemphigus vulgaris were considered.

Figure 1: Eroded plaques with crust formation over surface was seen over the chin region and left side of the beard region.

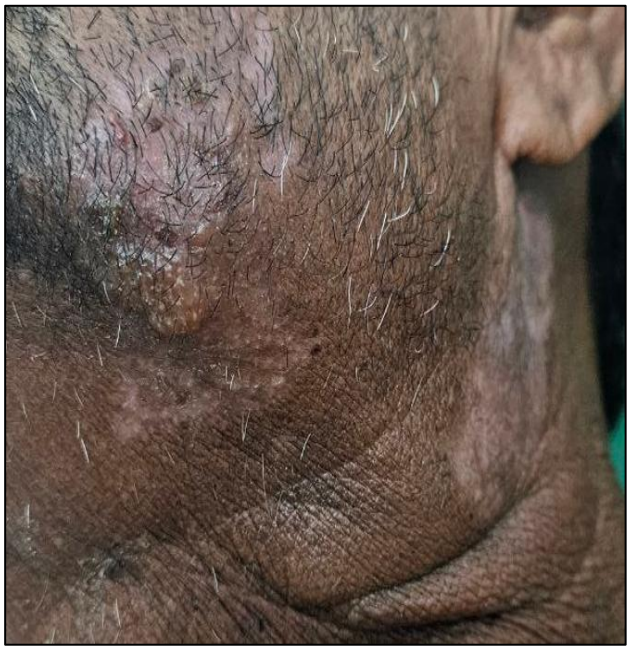

Figure 2: Two discrete crusted plaques with haemorrhagic crusting were seen over the mandibular region.

Figure 3: Erosion with crust formation were seen over the upper chest.

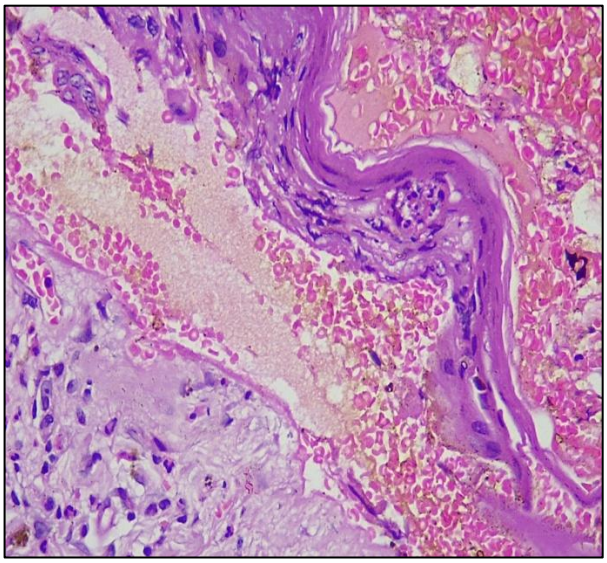

Figure 4: Histopathology-sub epidermal blister formation containing mainly haemorrhage.

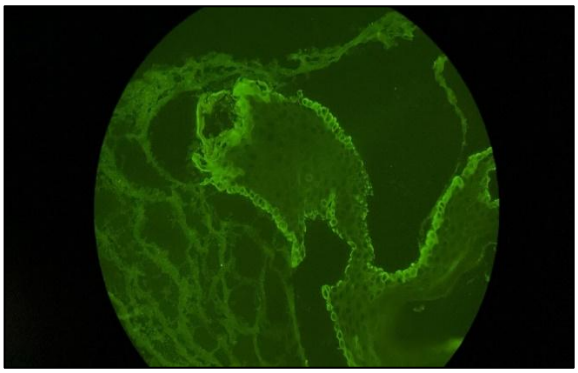

Figure 5: Direct immunofluorescence-IgG: positive (+3) lace-like pattern.

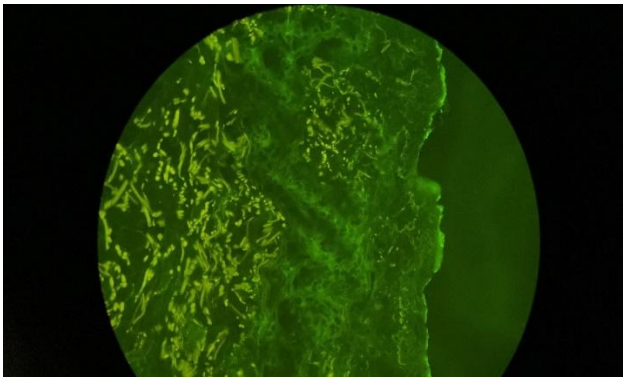

Figure 6: DIF-C3c: positive (+2) lace-like pattern.

Investigations and treatment

Routine investigations include hemogram, serum biochemistry, viral screening, chest X-ray, USG abdomen, ECG was within normal limits. For histopathology a Punch biopsy taken from small vesicle on beard region showed histopathological changes of sub epidermal blister formation containing mainly hemorrhage (Figure 4), the superficial dermis showed melanophages along with mild interstitial inflammatory infiltrate, composed of neutrophils and lymphocytes. A 3 mm punch biopsy was taken from perilesional skin.

Immunofluorescence: showed evidence of IgG: Positive (+3) (Figure 5), C3c: Positive (+2) lace-like pattern (Figure 6), in the intercellular squamous region. There is linear deposition of IgG along the basement membrane zone. Based on clinical features, histopathology and DIF findings the case was diagnosed as Paraneoplastic pemphigus. The patient was treated with oral prednisolone 30 mg/day for 2 weeks and gradually tapered over a period of 6 weeks. Oral antibiotic T. Cefuroxime axetil 500 mg twice daily was given for a week, Topical fusidic acid was given for 10days.Complete resolution of all skin lesions with residual pigmentary changes seen after 4 weeks.

DISCUSSION

PNP as an autoimmune disorder characterized by recalcitrant blistering, erosive mucocutaneous lesions, an associated neoplasm and high titres of “pemphigus like” anti-cell surface autoantibodies.[3] Later it was realized that the cutaneous lesions may be polymorphous, resembling those of PV, BP, erythema multiforme, graft-versus-host disease or lichen planus.[4] According to standard literature, paraneoplastic occurs concurrently with underlying malignancy in about 30% percent of patients. In the present case, patient developed skin lesions following 33 sessions of radiotherapy for squamous cell carcinoma.

The majority of patients are between the ages of 45 and 70 years. The common clinical feature, which is also usually the earliest presenting sign, is the recalcitrant oral stomatitis, seen as painful erosions and ulcerations of the oropharynx and vermilion border of the lips. It may resemble PV or herpetic stomatitis. The skin lesions are polymorphous and itchy and may resemble erythema multiforme (erythematous papules with central vesiculation), toxic epidermal necrolysis, BP (vesicles, bullae, erosions and crusting) or lichen planus. Conjunctival involvement, seen in approximately two-thirds of patients, may be severe, causing pseudomembranous conjunctivitis, symblepharon and significant visual impairment. Pulmonary involvement, sometimes severe enough to result in respiratory failure and death, has been reported to occur in 30% or more of patients in the form of bronchiolitis obliterans.[4] In the present case Patient developed flaccid bullae over previously irradiated area with no clinical evidence of eye and pulmonary involvement.

Although PNP shares some similarity with PV and PF, it differs from them in its clinical features, histological characteristics, immunofluorescent patterns, systemic involvement, association with neoplasms, poor response to therapy, poor prognosis and different pathophysiological characteristics. In two-thirds of cases, PNP occurs with an existing neoplasm. These may include non-Hodgkin’s lymphoma, chronic lymphocytic leukemia, castleman tumour (which is characterized by the growth of lymphoid tissue, usually in the mediastinum or retroperitoneum), Waldenström’s macroglobulinemia and, less commonly, thymoma (malignant and benign) and retroperitoneal sarcomas.[5] In the present case patient developed lesions after 33 sessions of Radiotherapy given for squamous cell carcinoma of tongue.

Histopathologic changes of intraepidermal acantholysis, keratinocyte necrosis and vacuolar interface dermatitis were seen. DIF observation of immunoreactions, typically IgG and C3, within the epidermal intercellular spaces as well as linear granular complement deposition along the epidermal BMZ are seen.[6] On indirect immunofluorescence (IIF), serum autoantibodies that bind not only to the cell surface of skin and mucosa in a pattern typical of PV but also to simple, columnar and transitional epithelium. In the present case subepidermal bullae formation along with inflammatory infiltration in upper dermis were in favour of PNP. Dual flouroscence with linear deposition of IgG along BMZ along with IgG and C3 deposits intercellularly in epidermis favoured a diagnosis of PNP.

PNP management involves a careful and multidisciplinary approach aiming to adequately treat the underlying neoplasm and control the mucocutaneous lesions. An ophthalmological evaluation is essential to determine the degree of ocular involvement and the appropriate treatment in order to prevent blindness. Corticosteroid eye drops, lubricants and artificial tears are often required. Successful management of cutaneous lesions with systemic steroids such as prednisone 1.0-1.5

mg/kg/day or in pulse therapy is possible, but mucosal improvement rarely occurs in monotherapy.[7] Azathioprine, Mycophenolate mofetil, The use of cyclosporine 5-7 mg/kg/day may improve cutaneous lesions, but with variable response of mucous involvement.[8] Rituximab is an immunobiological medication composed of chimeric anti-CD20 monoclonal antibodies that target both normal and tumoral cells expressing CD20.[9] Intravenous immunoglobulin (IVIg) is an alternative to counteract and decrease circulating pathogenic autoantibodies and to reduce inflammation through modulation of cytokines, complement, endothelial cells and lymphocytes.[10] In the present case because of limited cutaneous involvement we treated with only oral corticosteroids and other supportive drugs which resulted in complete resolution of all skin lesions within 2 to 3 weeks.

CONCLUSION

Our case highlights occurrence of paraneoplastic pemphigus over the localized areas of body in a patient after he recovered from carcinoma tongue. Satisfactory results were obtained with conservative treatment.

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