PEMPHIGOID IN IDENTICAL DIABETIC TWINS
A case report with review of literature

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Summary

Bullous pemphigoid developing at the same time in identical twin sisters with diabetes mellitus is reported.

Review of Literature

Bullous pemphigoid was first established as a distinct clinical and histopathological entity by Lever in 1953[1]. A new dimension was added to the study of bullous diseases by the development of immunopathology and immunofluorescence technique. The detection of serum antibody to the basement membrane of the skin has stimulated speculation that this distinct entity results from an autoimmune process[2]. Demonstration of Serum Ig G antibody fixing the complement at the exact site of pathologic change and predominance of polymorphonuclear leucocyte in the infiltrate are the evidences advanced in favour of this concept[3].

Although bullous pemphigoid is a disease associated with auto antibodies it rarely has been associated with other autoimmune diseases[4]. Diabetes mellitus is now recognised as a heterogenous disease and the evidences for a true autoimmune form of diabetes mellitus are accumulating. The clinical association of diabetes mellitus with diseases thought to be of autoimmune etiology such as idiopathic adrenal insufficiency, Grave's disease, Hashimoto's thyroiditis, pernicious anemia, myasthenia gravis and vitiligo is well recognised[5]. Bullous pemphigoid is also a disease of heterogenous nature. To the best of our knowledge diabetes mellitus has not been reported in frequent association with bullous pemphigoid. Rarity of this association and simultaneous development of diabetes mellitus and pemphigoid in identical twins prompted us to make this report.

Case Report

A fifty two year old married female, presented to the out patient department in February 1980 with multiple gradually progressive vesiculo bullous eruptions on the scalp, back, medial aspect of thighs, upperlimbs and face for one month. Lesions were mildly pruritic. Prior to her hospital visit, she had taken some indigenous medicines following which the lesions had worsened. Patient was a known diabetic for 6 years. She was on glybenclamide with which the diabetes was reported to be under control. Her twin sister who was married and residing at a place about 50 miles away was reported to be on treatment for similar skin complaints from January 1980. A dermatologist at a local hospital had diagnosed her to have pemphigoid.

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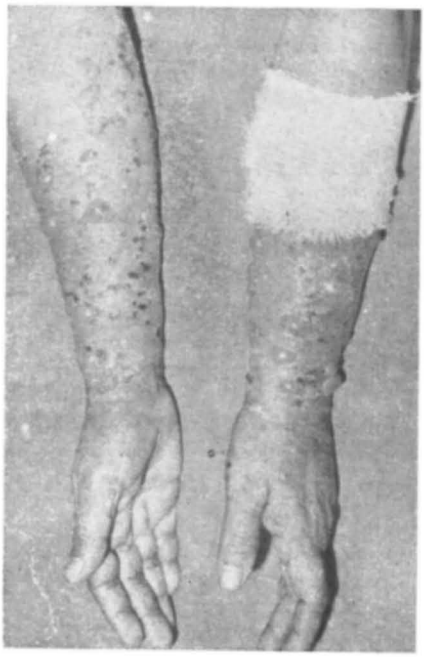

Fig. 1 Photograph showing vesico bullous eruptions on both upper limbs of patient (A)

She was also a known diabetic for 6 years and had been on treatment with glybenclamide for 2 years.

Personal, family and past history were non-contributory. General and systemic examination revealed nothing abnormal.

Local Examination

Multiple tense bullae were present over the face, scalp, upper limbs, flexural aspect of the fore arms (Fig 1), medial aspect of thighs and back. Palms and soles and mucous membranes were spared. Bullae were tense and on considerable pressure spread to the neighbouring area. The twin sister was also seen but her lesions has subsided with steroids. The scars which had resulted from the lesions had distribution similar to that of the patient's.

Routine laboratory examinations were within normal limits. Fasting blood sugar was 192 mg% and post prandial blood sugar 400 mg%. Serum proteins showed total protein of 6.8 gm% with albumin 3.5 gm% and globulin 3.15 gm%. Liver function tests were within normal limits and Serum amylase 96 units/100ml. X-ray chest was normal.

Histopathological examination showed sub-epidermal bullae (Fig 2) containing eosinophilic material, some polymorphs, pigmented macrophages, mononuclear cells and few old eosinophils. The base of the bullae showed infiltration by the same type of cells. No acantholytic cells were seen. Absence of acantholytic cells, sub-epidermal location of the bullae, presence of polymorpho nuclear leucocytes and eosinophils in the infiltrate and failure to respond to D.D.S. were features consistent with a diagnosis of pemphigoid.

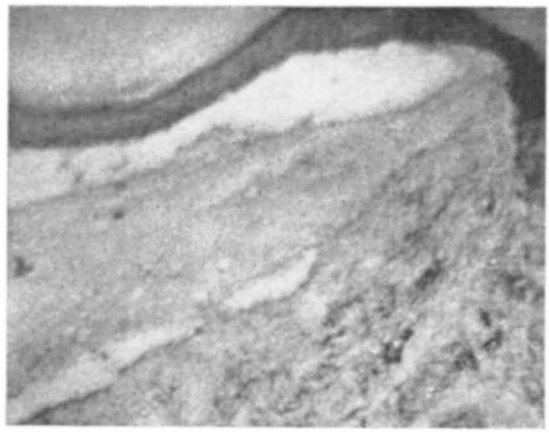

Fig. 2 Photomicrograph showing sub epidermal bullae of the skin containing eosinophilic material and the base showing infiltration with polymorphs and eosinophils

Discussion

Pathogenisis of bullous pemphigoid is unknown. Bean et al proposed that nonspecific insults to the dermoepidermal junction might result in the formation of tissue specific antibodies. These insults may be brought on by drugs, chemicals or ultraviolet rays, or may accompany other diseases like alopaecia areata, ankylosing spondylitis, benign and malignant neoplasms, inflammatory bowel diseases, lichen planus, membranous glomerulo nephropathy, multiple sclerosis, pancreatitis, polymyositis, psoriasis, rheumatoid arthritis, sarcoidosis, systemic lupus erythematosus and toxic goitre. The antibody formation may perpetuate the reaction in an autoimmune manner[5].

Drugs may assume a more than coincidental role in the induction of bullous pemphigoid as illustrated by several recent reports[8]. Fellner and Katz[9] reported the development of bullous pemphigoid in a 78 year old lady following furosemide therapy[9]. In our case possibility of induction of pemphigoid by the drug glybenclamide or non specific insults to the dermoepidermal junction due to diabetes mellitus in genetically predisposed individuals is to be considered.

Acknowledgement

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