PREGNANCY INDUCED PEMPHIGUS VULGARIS IN A YOUNG ADULT LADY

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Abstract

Pemphigus vulgaris is a cutaneous blistering disorder affecting mainly middle aged adult and is rarely observed in pregnancy and also below the age of 30 years. A 27 years old lady with 7 months pregnancy reported to Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka with oral and cutaneous lesions suggestive of pemphigus vulgaris. Tzanck smear, histological examination and direct immunofluorescence study confirmed the diagnosis and the case showed improvement with only oral steroid. The patient delivered a normal healthy child without complications. No clinical relapse was found after delivery and follow up period of six months.

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Key words: *Pemphigus vulgaris, Pregnancy, Direct Immunofluorescence study.*

Introduction

The term pemphigus (Greek pemphix meaning bubble) refers to a group of autoimmune intraepidermal blistering disorders of skin and mucous mucosae.It is characterized by mucosal erosion and thin walled, relatively flaccid, easily ruptured bullae that appear as apparently normal skin and mucous membrane or erythamatous bases. It affects mainly 5th to 6th decade and is rarely observed before 3rd decade and also rare in pregnancy. We report a 27 years old women having 7 months pregnancy with oral and cutaneous lesions suggestive for Pemphigus vulgaris. Diagnosis was made on the ground of clinical appearance, history, cytological, histological and immunofluorescence finding. Systemic steroid was effective in controlling the disease. Foetal mortality is high due to transplacental transmission of pemphigus antibody from mother to child. Risk of using teratogenic immuno suppressive drug may produce defect in the foetus.

Case report

A 27 years old lady having 7 months pregnancy presented to the Department of Dermatology and Venereology of Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, Bangladesh with 5 months history of oral lesion and 1 month history of vesicobullous skin lesions on different parts of the body. She had been treated with various drugs, like antibiotics,

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vitamins, antihistamine, analgesic and topical cream with out any improvement.

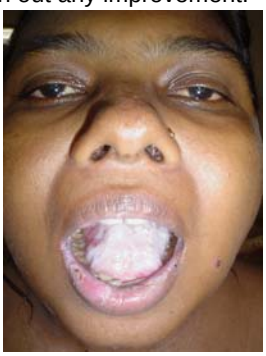

Fig 1: Erosive lesions on lips and oral cavity before treatment.

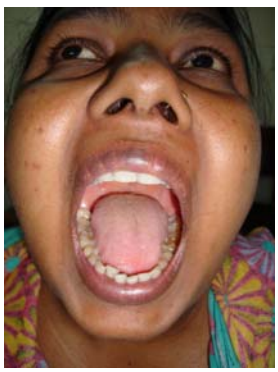

Fig 2: Same patient after treatment.

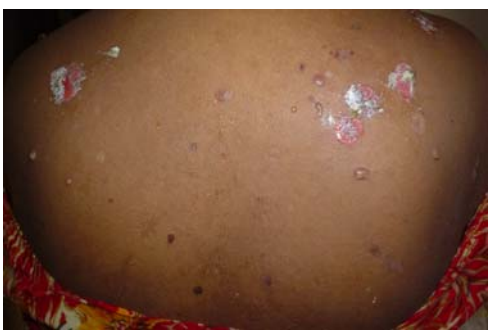

Fig 3: Bulla with eroded and crusted skin lesions before treatment.

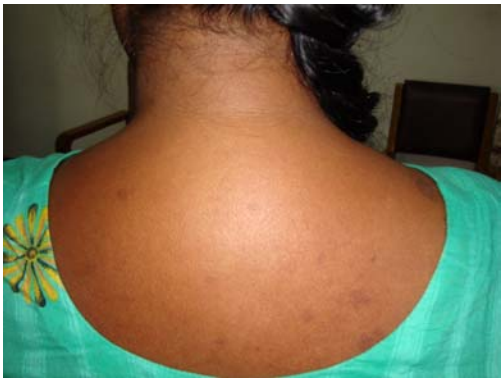

Fig 4: Same patient after treatment.

On examination we found multiple erosive lesions on lips and oral cavity, multiple flaccid vesicobullous lesions on different parts of the body with intermingly eroded area (Photograph-1 & 3). Nikolsky's sign and bulla spread sign are positive.

A Tzanck smear showed typical acantholytic cells. A skin biopsy taken from a bulla showed intraepidermal bullae in suprabasal location containing acantholytic cells. Direct immunofluorescence found deposition of IgG autoantibodies and C3 in the intercellular space. The histopathology and immunofluorescence was consistent with the diagnosis of Pemphigus vulgaris. Anti-desmoglein titre could not be done due to unavailability in our country. Other haematology and biochemical parameter were with in normal limit. Based on clinical and laboratory findings and the close relationship between the onset of mucocutaneous and cutaneous features the diagnosis of pregnancy induced pemphigus vulgaris was made which was her 3rd pregnancy but previously this never happened before.

Systemic treatment with prednisolone 1mg/kg body weight daily and topical treatment with clobetasone with neomycin and other supportive treatment resulted in regression of the disease. The dose of steroid was gradually tapered and stopped before delivery. The patient delivered a normal healthy child without complications. No clinical relapse was found after delivery and six months follow up period (Fig: 1b,2b,3b)

Discussion

Pemphigus vulgaris is characterized by mucosal lesions and thin walled flaccid, easily breakable bullae that appears on erythematous or normal appearing base distributed typically on mouth, groin, scalp, face, neck, axilla and genitalia.[1,2] In Pemphigus ,autoantibodies react with intercellular adhesion molecules between the epidermal keratinocytes that in turn results in separation of cells from each other(acantholysis) and thus produce blistering.The antibodies in Pemphigus vulgaris are most commonly directed against desmoglein-3. The course of the disease is usually severe and variably it responds to oral steroids, steroid plus steroid sparing agents(azathioprine, mycophenolate mofetil, cyclophosphamide), plasmapheresis, I/V Immunoglobulin, gold, tetracycline with nicotinamide and also newer biologics.[3,4,5]
The prognosis is variable according to severity of the disease. Several exogenous factors are capable of inducing PV in genetically predisposed people including drugs, viral infections and exposure to physical agents (heat, ultraviolet light and ionizing radiation, surgical and cosmetic procedure).[6,7,8]
The complex mechanism stimulates the keratinocyte to produce various cytokines and tumor necrosis factor. These cytokines regulate the synthesis of complement and proteases such as plasminogen activator, which has a pivotal role in the pathogenesis of acantholysis.[9]
Our patient presented with seven months pregnancy with 5 months duration of oral erosive lesion and 1 month duration of multiple flaccid, easily breakable bullae on different parts of the body.
A Tzanck smear showed acantholytic cells, on skin biopsy revealed intraepidermal bullae in suprabasal location containing acantholytic cells and direct immunofluorecence findings were deposition of IgG and C_3 in the intercellular space in epidermis which were consistent with pemphigus vulgaris. Systemic prednisolone 1mg/kg/day with topical clobetasone and neomycin and other supportive treatment resulted regression of the disease. The steroid was tapered gradually and stopped before delivery. No clinical relapse was seen during 6 months follow up period.
This report illustrates the importance of pregnancy induced pemphigus and hormone related to pregnancy should be added to the list of pemphigus inducer or pregnancy related dermatoses.

Conclusion

Pemphigus may be exacerbated during or after pregnancy, but often to a mild degree. Although the rate of stillbirth was not as high as previously reported, the rate of abortion was considerable. Pregnancy may have an uneventful course, especially in patients in clinical remission; nevertheless, careful monitoring of the high risk mother and fetus is mandatory.

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