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Gestational Pemphigus: A Case Report

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ABSTRACT

Gestational Pemphigus is the most common among the pemphigus group of disorders. Its incidence is variable across the globe and among various ethnicities. Many studies show that there exists a female preponderance of gestational pemphigus with an approximate female-to-male ratio of 1.4:1. Pemphigus could involve one or more mucosae, while Gestational pemphigus inflammatory skin lesions and severe pruritus. When it occurs in pregnancy, the condition becomes more complex. Early diagnosis and individually adjusted therapy are needed to avoid any risk for mother or child. Hence; we report the data of a patient who was diagnosed with gestational pemphigus in the department of OBG and Gyne, NKP Salve Institute of Medical Sciences and Research Centre.

Keywords: *Pemphigus, Gestational*

1. INTRODUCTION

The term pemphigus stems from the Greek ‘pemphix’, which means blister or bubble, and it describes a group of chronic blistering epithelial diseases in which the production of IgG autoantibodies against extracellular domains of cell membrane proteins of keratinocytes results in acantholysis (the loss of cell–cell adhesion between keratinocytes). In pemphigus, IgG autoantibodies are characteristically directed against desmogleins (desmoglein 1 and desmoglein 3), which are part of the cadherin family of cell–cell adhesion molecules that are found in desmosomes, which are the structures primarily responsible for maintaining intercellular adhesion in stratified squamous epithelia, such as the skin and oral mucosa.[1- 3]

Gestational pemphigus is the most common among the pemphigus group of disorders. Its incidence is variable across the globe and among various ethnicities. Many studies show that there exists a female preponderance of GP with an approximate female-to-male ratio of 1.4:1. The age of onset reported is between the fourth and sixth decades of life, with the exception of certain ethnicities like the Middle East and Asia that report disease onset earlier by about a decade. Pemphigus usually affects the elderly, and genetics play an important role in predisposition. When it occurs in pregnancy, the condition becomes more complex. Early diagnosis and individually adjusted therapy are needed to avoid any risk for mother or child.[4, 5]Hence; we report the data of a patient who was diagnosed with gestational pemphigus in the department of OBG and Gyne, NKP Salve Institute of Medical Sciences and Research Centre.

2. CASE REPORT

A 29 year old female, at 33 weeks gestation in her fourth pregnancy presented to the outpatient dermatology department with intensely itchy pruritic eruption since 15 days. The lesions were preceded by generalised itching, predominantly intense. Following which she developed multiple erythematous, areas that progressed to involve her arms, legs, oral cavity and neck. The lesions then developed a central dusky hue and peripheral rim of vesicles.The patient has been married for 7 years and previous pregnancies resulted in a full term LSCS.She had no family history of dermatological conditions or autoimmune diseases. There was no history of similar complaints in her previous pregnancy, no history of systemic illness, HSV infection and no history of drug intake other than multivitamins.Routine haematological and biochemical tests revealed dimorphic anaemia. Other parameters like bile acids, renal function were normal. Histopathological examination displayed supra basal bulla with plasma cells and few eosinophils. Dermis showed moderate degree of perivascular infiltration by lymphocytes and eosinophils. Based on the clinico-pathological correlation, diagnosis of gestational pemphigus was made.

Figure 1: Pemphigus affecting a) cavity, b) lips, c) hands, d) upper back, e) feet, f) neck and g) legs

3. DISCUSSION

Pemphigus in pregnancy is a special clinical scenario that has potential consequences on both maternal and fetal outcomes. Being an autoimmune disease with Th2 preponderance, pemphigus is expected to flare in pregnancy, especially in the first two trimesters. Fetal outcomes like stillbirth and neonatal pemphigus have been reported, the latter being a consequence of a transient transplacental transfer of autoantibodies. Management needs to be individualized keeping the risk/benefit ratios of therapies in mind while optimizing maternal and fetal health. It is crucial to have appropriate counseling regarding conception for women with pemphigus in the child-bearing period because the probability of adverse materno-fetal outcomes is higher if the disease is severe.[5- 7]

Pemphigus vulgaris can manifest as a mucosal-dominant, mucocutaneous or, less frequently, solely cutaneous type1. Lesions classically start in the oral mucosa and might then extend to the skin. Erosions of the intertriginous areas (where skin folds) might develop into vegetative lesions (abnormal growth of keratinocytes with granular papillomatous appearance) in the pemphigus vegetans variant. In pemphigus vulgaris and pemphigus foliaceus, Nikolsky sign might be positive in the active phase of the disease. A substantial number of patients with pemphigus vulgaris can relapse with a pemphigus foliaceus clinical and histological phenotype, especially when the relapses occur after a long remission.[7- 9]

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reviewed the literature, focusing on the relationship between pemphigus vulgaris and pregnancy. They conducted comprehensive searches in PubMed, Embase, Cochrane Library, and Web of Science databases, identifying 42 studies reporting the disease course, pregnancy outcomes, and management of both pregnancy and pemphigus vulgaris.A total of 57 cases were included in the analysis, categorized into three distinct forms: pemphigus vulgaris onset before pregnancy (n = 33), onset during pregnancy (n = 20), and onset during the postpartum period (n = 4). Fifty-four cases reported treatment strategies, among them, 44 cases (81.5%) initially received systemic corticosteroid therapy during pregnancy. Out of these cases, 7 (15.9%) did not achieve successful remission and required alternative treatment approaches. In terms of pregnancy outcomes, 23 out of 62 neonates (37.1%) exhibited skin lesions or tested positive for anti-dsg IgG in their serum, while 16 neonates (25.8%) experienced other complications.These findings highlighted the importance of effectively managing pemphigus vulgaris during pregnancy to ensure optimal outcomes.[10]Endemic pemphigus foliaceus (PF), also known as Fogo Selvagem (FS), is an organ-specific autoimmune disease mediated by autoantibodies. These autoantibodies are disease specific, predominantly restricted to the IgG4 subclass, and pathogenic, as demonstrated by passive transfer studies. In contrast to pemphigus vulgaris, neonatal skin disease does not appear to occur in babies born to mothers with non-endemic PF or FS. Rocha-Alvarez R et alhave examined 19 mother/neonate pairs where the mother had documented FS. Mothers and neonates were examined soon after delivery and tested by immunofluorescent (IF) techniques for FS autoantibodies either in circulation (mothers' sera or babies' cord blood) or bound to the neonatal epidermis. All neonates included in this study were born with normal skin. Twelve biopsies from 17 neonates showed negative direct IF using both FITC-antihuman IgG or monoclonal anti-IgG subclass antibodies. In five biopsies the epidermal ICS of the babies showed weak staining. In 10 of the 19 cord sera tested, FS IgG autoantibodies were undetectable; in nine, these autoantibodies were present in low titers (less than 1:40). The sera of the mothers showed higher titers of FS autoantibodies, and IgG4 was the predominant IgG subclass autoantibodies. It appears that human placenta may modulate the expression of disease in the newborn by operating as a "biologic immunoadsorbent" of pathogenic autoantibodies.[11]

4. CONCLUSION

Management needs to be individualized keeping the risk/benefit ratios of therapies in mind while optimizing maternal and fetal health.Our case report highlights the importance of effectively managing pemphigus vulgaris during pregnancy to ensure optimal outcomes.

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