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A Rare Case Report of Pemphigus Vulgaris in Pregnancy with Successful Pregnancy Outcome

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Abstract: ***Pemphigus vulgaris (PV) is a rare immune mediated bullous dermatosis which progresses to form a fragile blisters and extensive lesions on the skin and mucous membranes, During pregnancy, exacerbation of PV is observed due to changes in maternal hormones and there are adverse effects of the drug on both mother and the fetus, hence it becomes more challenging to diagnose and manage the disease during pregnancy. We report a case of a 23-year-old woman who was diagnosed with pemphigus vulgaris (PV) before 2nd trimester of pregnancy. The patient presented with widespread blistering dermatitis and associated with burning pain and pruritus. A skin biopsy revealed suprabasal acantholysis, diagnosis of PV confirmed and treated with corticosteroids, antibiotics, antihistaminics after explaining the chances of anomalies and other adverse effects of steroids on the fetus. The pregnancy progressed to Full term normal vaginal delivery of the neonate without skin lesions or apparent complications. Obstetrician should know the clinical presentation, complication and outcome of the disease to prevent the further complications.***

Keywords: Pemphigus vulgaris, pregnancy, autoimmune bullous dermatosis, corticosteroid

1. Introduction

Pemphigus vulgaris (PV) is an autoimmune bullous dermatosis affecting people of all races and showing equal prevalence in both men and women. [(1)]Autoantibodies for desmoglein 1 and desmoglein 3, members of the cadherin family involved in cellular adhesion, have been linked to the pathogenesis of PV. [(2)] These autoantibodies play a role in the lossof cell-to-cell adhesion in the basal and suprabasal layers of the deep epidermis while cellular adhesion in the superficial epidermis remains intact, leading to the clinical presentation of epidermal blistering and ulcerations most commonly found on the scalp, face, groin, and axillae. [(3)] The PVs cutaneous manifestations often include widespread vesicles, followed by erosions and ulcerations that heal without scarring, as these lesions are entirely epidermal [(4)] Nasal and oral mucosae are often involved in PV which may either be the only manifestation of PV or proceeded by cutaneous lesions. [(5)] Diagnosis typically is made based on skin biopsy and confirmed by direct immunofluorescence. Histologically, PV displays acantholysis and suprabasal cleft formation. Immunofluorescence may show IgG antibodies against the PV antigen in the epidermis. [(3)] Corticosteroids and immunosuppressive drugs are considered as effective treatments and have reduced the mortality rate of PV to5%-15%. [(6)] Pemphigus vulgaris during pregnancy may result in neonatal complications. Maternal antibodies may cross the placenta resulting in neonatal pemphigus. [(7, 8)] Use of corticosteroids during pregnancy can be potentially dangerous to the fetus, particularly if high doses are necessary to control maternal disease. [(9, 10)] corticosteroid treatment can be associated with low birth weight, prematurity, adrenal insufficiency, and infection in newborns. [(11)] We report a case of pregnancy-triggered pemphigus vulgaris before second trimester of pregnancy followed by the birth of a healthy neonate.

2. Case Report

A 23 years old g3p2l2 (gravida3, para2, living2) with 20 weeks of gestation was presented with chief complaints of generalized skin eruption with burning and pruritus, since 1 month, initially lesion appeared on oral cavity progressed to involve the trunk, upper limb, lower limb, axilla, genital region. Patient had no similar lesion in previous pregnancy.

Figure 1: Erosions over tongue, crusting of lips

On admission patient was conscious, oriented, gc: moderate, spo2: 99%offo2, p: 100bpm, bp: 110/60mmhg, cvs/rs: nad p/a: ut corresponding to 14 weeks, relaxed, external ballotment +, on L/E: multiple erosions with crusting involving oral mucosa, lips, scalp, face, trunk, bilateral upper and lower limbs, axilla, interdigital spaces. Patient had difficult in swallowing due to erosions of oral cavity and crusting of lips. she was started on inj. dexamethasone 8mg

iv od, inj. amoxycillin and clavulanic acid 1.2gm iv bd, antihistaminics, multivitamin, fusiwal cream, mucopain oral gel, candid mouth paint. Investigations revealed hb: 11gm%, wbc: 11400/mm^3, ESR: raised, RFT & LFT: Normal. vdrl: NR ultrasonography: single live fetus of 14 weeks gestation, changing lie, 45gm, liq: adequate with no anomalies, skin biopsy sent, rash didn't settle in spite of above mentioned treatment then the dose of dexamethasone is increased from 8 mg od to 8 mg bd patient showed response with improvement in lesion with no new lesions, dose tapered from 8 mg bd to 8 mg od and she was discharged on tablet. prednisolone 30 mg od for 7 days with biopsy report showing suprabasal cleft with few acantholytic cells and few polymorphs s/o pemphigus vulgaris diagnosis of PV was confirmed.

Figure 2: Erosions in interdigital spaces

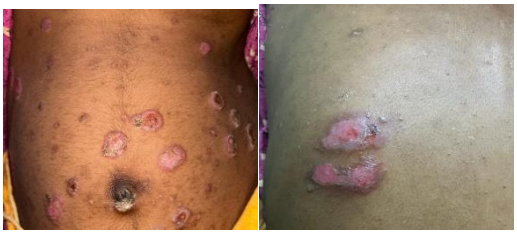

Figure 3: Erosions and crusting over front of abdomen and on back

Figure 4: Lesions in the scalp

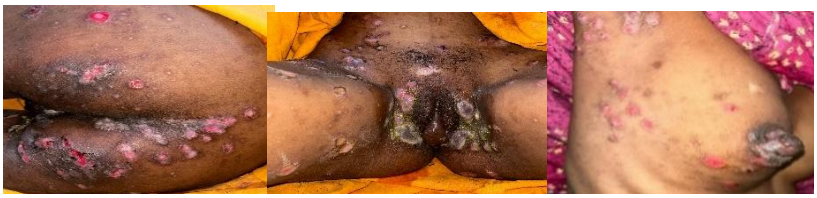

Figure 5: Erosions on the buttocks, groin, in axilla, nipples

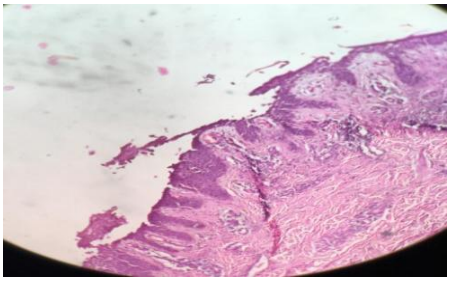

Figure 6: Biopsy report showing suprabasal cleft withacantholytic cells and few polymorphs

patient again presented after 1 month with similar lesions with fever with generalized weakness, all the investigations were done hb: 11.6gm% wbc: 18400/mm^3, es: raised, ps for mp: neg, dengue: neg, widal: positive with O: H 1: 160 titres, ultrasonography: single live fetus of 20 weeks gestation, changing lie, 450gm, liq: adequate with no anomalies. patient started on inj. Ceftriaxone 1 gm / iv /bd, tab. Prednisolone 15mg /od, clobetasole with fusidic acid cream for L/A, fever subsided, lesions started resolving, she was discharged on tab. Prednisolone 10 mg bd for 10 days tapered to 10 mg od for next 10 days, then to 5 mg od for 10 days. We have followed up her till delivery; she underwent spontaneous full term vaginal delivery of healthy baby of 2.7 kg, without any anomalies and complications.

3. Discussion

PV is an autoimmune, blistering, inflammatory disease of epidermal layer of the skin. Pemphigus vulgaris is associated with infertility in its active phase; therefore, PV during pregnancy is rare. [12] PV is commonly seen in 4-6th decade Pregnancy may exacerbate PV, which has been a similar finding in other well-documented autoimmune diseases. [10] One review of PV in pregnancy reported that 11 of 49 patients (22%) experienced an exacerbation of the disease. [12] Our patient presented during her second of pregnancy with blisters involving mucous membranes and skin of various parts of body from lips to limbs, trunk, genital area, axilla; lesions appeared and healed at different points in time. Transient skin lesions may occasionally appear in theneonate and seem to have an increased association with severe active PV in the mother; however, neonatal PV also has been present in mild cases in the mother. [10] but in our case neonate didn't show any complication. In pemphigus vulgaris the antibody titers or clinical presentation of mother do not predict the severity of disease in the neonate Antibody titers as low as 1: 20 in the neonate may produce disease. [13] outcome of PV in pregnancy could be normal live birth, stillbirth, spontaneous abortion, intrauterine fetal death, premature delivery, and neonatal pemphigus. One important differential diagnosis for PV in pregnancy ispemphigoid gestationis. Pemphigoid gestationis is a rare autoimmune disease during pregnancy and postpartum period. It is characterized by intense itching and erythematouspapules that later form vesicles. They appear on the trunk and abdomen and spare the face and mucosalsurfaces. Clinical manifestations (not involving mucosalsurfaces), histopathological findings (subepidermalvesicles with lymphocyte and eosinophil infiltration), and immunofluorescence findings (C3 and IgG

in a linear band along the basement membrane) of pemphigoidgestationis can differentiate it from PV in pregnancy. [14, 15] treatment with high dose steroid can cause complication in the newborn especially if used during organogenesis, but controlling the disease with aggressive treatment is necessary as our patient was having severe widespread disease. Inadequate treatment of pemphigusvulgaris can result in life threatening complication because of the severe infection. Thus it is necessary for the health of the patient and fetus to suppress the disease. prednisolone, is the drug of choice and the patient has shown drastic improvement after steroid use. Similar to prednisone, azathioprine is not recommended during pregnancy, but if use is necessary, it is suggested to keep the dose low to prevent fetal harm. [16] One alternative to treatment with steroids and immunosuppressants is plasma exchange, which has been successful in the clinical context of pregnancy. [17] The cons of plasma exchange are repeat procedures, the need to give the patient more immunosuppressants to prevent a rejection, and the return of the autoantibody. [10] Rituximab therapy proved to be useful in the clinical improvement of patients with refractory pemphigus vulgaris. Studies are currently under way to look at the effects of Rituximab on pregnancy and the fetus. ([18]

4. Conclusion

Although pemphigus vulgaris is primarily a dermatological disease, as obstetricians, we should be capable of early diagnosis to prevent further complications. The diagnosis was made after excluding other diseases and confirmed by skin biopsy. Initiating treatment with high dose steroid during the pregnancy was challenging and risky, but we had to start the treatment with steroid to control the disease. Ultimately healthy baby delivered by normal vaginal delivery was free of disease. Pemphigus vulgaris is a life threatening in some cases. So, we have to treat the disease aggressively to save the life of mother, though the life of fetus is compromised. Pemphigus vulgaris in the mother was efficiently controlled by low-dose prednisolone during the pregnancy.

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