

Case Report

Pemphigus vulgaris in pregnancy: Analysis of current data on the management and outcome

Rajalaxmi Kamath^{*1}, Shraddha Shetty² and Nanditha Shenoy³

¹Assistant Professor, Department. of OBG, Yenepoya Medical College, Mangalore, India

²Assistant Professor, Department of OBG, KMC Mangalore, India

³Associate Professor. Department of oral Medicine and Radiology. MCOODS Mangalore. India

***Correspondence Info:**

Dr. Rajalaxmi Kamath.
Assistant Professor.
Department of OBG,
Yenepoya Medical College, Deralakatte Mangalore. India.-
E-mail: gkbkrishna01@gmail.com

Abstract

Pemphigus vulgaris (PV) is an uncommon immune mediated bullous dermatosis, which, during its active phase, has been associated with infertility. PV is exceedingly rare during pregnancy. Only 26 cases with immunopathological confirmation have been reported. The disease may be associated with adverse neonatal outcome, including prematurity and foetal death. Transient skin lesions may occasionally appear in the neonate. We present a case of a patient who was diagnosed to have pemphigus since 3 years receiving corticosteroids and delivered a healthy female baby at 38 weeks of gestation.

Keywords: pemphigus vulgaris, pregnancy, autoimmune

1. Introduction

PV is a rare, autoimmune bullous dermatosis. The disease affects all races and both sexes, appears in middle age and rarely affects children. Genetic factors play an important role in predisposition to PV.¹ Certain major histocompatibility complex (MHC) class II molecules are more common with PV.² Joly et al stated that the pathogenesis is linked to the presence of autoantibodies directed against desmoglein 3, a desmosome trans-membrane glycoprotein belonging to the cadherin family. These autoantibodies cause blisters which result from loss of cell to cell adhesion in the basal and suprabasal layers of the deeper epidermis, with the keratinocytes in the superficial layers of the epidermis that maintain their cell adhesion.⁴

Clinical manifestations include numerous skin vesicles, most of which result in widespread erosions and ulcerations that heal without scarring, as the erosions are entirely epidermal. Involvement may be localised or generalised but the disease has a predilection for the scalp, face, axillae, groins and Pressure points. The oral and nasal mucosae are often involved (50 to 70% of patients). They may precede cutaneous lesions by months or maybe the only manifestations of disease. Diagnosis is based on lesion biopsy showing the presence of acantholysis, suprabasal cleft formation and deposition of immunoglobulin (Ig G) and complement in the intercellular spaces of the epidermis. IgG antibodies against the pemphigus antigen may be detected by indirect immunofluorescence in the serum.⁵

Before the availability of corticosteroid therapy the disease was usually fatal, due to secondary infection and sepsis or electrolyte imbalance.⁶ Treatment with systemic steroids has reduced mortality to 5 to 15%. Impairment of fertility is associated with various autoimmune disorders such as autoimmune premature ovarian failure, pernicious anemia, crohn's disease, systemic sclerosis, rheumatoid arthritis, IDDM. Recently an association of PV with infertility was described.⁷

2. Case Report

A thirty year old Gravida 2, Para1 living 1 who had been suffering from PV for the last three years came for antenatal checkup at 10 weeks of gestation. Her first delivery was a full term normal delivery 8 years back. She had delivered a healthy male baby. PV was suspected in her three years back when she developed bullous eruption in her mouth and then on her body. It was confirmed by Histopathology and demonstration of autoantibodies in serum by indirect immunofluorescence.

She was treated with corticosteroids. She had a flare up of disease with secondary infection two years back, for which she was admitted and treated. She had no menstrual problems and conceived while still on treatment. (Prednisolone 10 mg .O.D.) She had hyperemesis in first trimester. She developed lesions on thighs, forearm and abdomen associated with severe itching in second trimester. Oral mucosal and lesions in epiglottis developed later which resulted in dysphagia. The dose of prednisolone was increased to 30 mg/day and maintained for 15 days till she improved and further continued on a dose of 10 mg/day. Pregnancy follow up was otherwise normal regarding blood pressure examination, maternal weight gain and glucose tolerance test. The prednisolone was maintained at 10mg/day dosage till 38 weeks of gestation when she was admitted with premature rupture of membranes and oxytocin induction of labour resulted in vaginal delivery. Live female baby of 2.4 kg was born. The baby was healthy without manifestations of pemphigus. Both mother and baby did well in postnatal period.

Fig. 1 Buccal mucosa with area of erosion showing tissue



Fig. 2 Oral mucosal lesion in Pemphigus vulgaris



3. Discussion

PV is associated with difficulty in conceiving as other autoimmune disorders. In a retrospective study⁷ eight of nine patients suffering from PV failed to conceive. In contrast to these data, our patient conceived implying that the disease is not necessarily associated with infertility. The pregnancy may precipitate or aggravate PV as reported in autoimmune diseases such as SLE.⁸

Our patient had flare up during second trimester but improved in third trimester which can be well explained by using endogenous corticosteroids production by chorion and consequent immunosuppression.⁹ Transplacental transmission of PV IgG antibodies from mother to foetus may result in clinical manifestations in neonate.¹⁰ The baby in our case did not show any signs of the disease. PV was associated with more adverse neonatal outcomes such as growth restriction and preterm births. But, in our case we did not have any adverse outcome. The effect of PV varies during pregnancy, our patient had premature rupture of membranes which could be due to corticosteroid treatment. Their connection has been previously described where a significant increase in preterm births and premature rupture of membranes in women treated with prednisolone is more than those women treated with placebo.¹¹

Management in pregnancy should involve individual balancing of the risks of the disease and its complications against the potential adverse effects of therapy on both mother and child. According to Goldberg vaginal birth can result in worsening and spreading of pemphigus while corticosteroids can delay wound healing in patients undergoing caesarean section.¹² However, caesarean is indicated in patients with pemphigus lesions on genital mucosa. In our patient there was no involvement of genital area whereas skin of lower abdomen was involved making caesarean section a less appropriate mode of delivery.

4. Conclusion

In a case of PV it is essential to take preventive measures before conception to avoid complications. For example, efforts should be made to taper or stop any immunosuppressive agent. The dose of prednisolone should be reduced to the lowest effective dose. On the other hand, an adequate control of the disease is required before conception as it is expected that pregnancy can aggravate pre-existing PV as it does in other autoimmune diseases. However Lehman et al¹³ believe that adverse pregnancy outcome is more closely related to poor control of maternal disease and high titres of pemphigus antibodies. Corticosteroids are the first choice of treatment for PV. If disease is not controlled then steroid-sparing immunosuppressive agents may be added to therapy such as Azathioprine,¹⁴ but its use during pregnancy should be avoided. Plasmapheresis could be used as treatment during pregnancy but its use is still experimental.

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