

# Pemphigus vulgaris during pregnancy – a case report

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**Abstract** Pemphigus vulgaris (PV) is an uncommon immune-mediated bullous dermatosis which is very rare during pregnancy. Its management during pregnancy is a challenge and sometimes very difficult. Only few cases have been reported in literature so far. The disease may be associated with adverse fetal outcomes such as prematurity and fetal death. The neonate can develop transient skin lesions. We present a case of a patient who conceived during the active phase of PV required high doses of corticosteroids and delivered a preterm appropriate for gestation age newborn.

**Key words**

Pemphigus vulgaris, pregnancy.

## Introduction

Pemphigus vulgaris (PV) is a severe, potentially life-threatening autoimmune disease of the skin and mucous membranes. This cutaneous disorder is characterized by bullae in the skin and mucous membranes (mouth, upper airway, genitalia). The underlying mechanism involves the formation of autoantibodies against the cadherin-type adhesion molecules desmoglein 3 mainly.<sup>1</sup>

PV more frequently affects middle-aged women, including those in their childbearing years.<sup>1</sup> Compared to western countries it occurs at a younger age in our part of the world.<sup>2</sup> It is very rare to see this condition during pregnancy. Very few immunopathologically confirmed cases have been reported in literature so far.<sup>3</sup>

Pregnancy may precipitate or aggravate PV.<sup>3</sup> The disease may occur for the first time

during pregnancy or the disease may precede the pregnancy with or without the exacerbation of the disease.<sup>1</sup> The flaring is reported more during the first and second trimester and in the postpartum period.<sup>4</sup>

The effect of pemphigus varies during pregnancy, ranging from stillbirth, intrauterine growth retardation to preterm birth, as in our case. The clinical manifestations of the disease vary during pregnancy from mucosal lesions only in some patients to both mucosal and cutaneous lesions. While in a small number of patients only skin was seen to be affected, very few pregnant women presented with gingival lesions while some had no active lesions.

The reported perinatal mortality rate is 12%.<sup>1</sup> Different causes identified by various authors are intrauterine growth retardation, placental insufficiency, immune suppression, infections, and adverse drug reactions.<sup>1,3,5</sup>

In 30% to 45% of cases, there is some transfer of antibodies through the placenta from the mother to the fetus causing transient neonatal pemphigus which resolves completely within

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few months. However, in most cases the neonates are born completely free of disease.<sup>4</sup>

The management of PV during pregnancy is similar to that in the nonpregnant woman; however, it is more challenging and difficult during pregnancy. Steroids such as prednisolone are the first choice. Corticosteroids are considered safe during pregnancy, however, high dose or aggressive therapy may increase risk for having low-weight babies, infection and adrenal insufficiency, preterm premature rupture of membranes and preterm delivery.<sup>1</sup> Azathioprine and cyclosporine A which are used in non pregnant are not considered very safe during pregnancy. Breastfeeding is not contraindicated. Passive antibody transfer to the infant is a possible hazard.<sup>5</sup>

### **Case report**

A 22-year-old primigravida, who had been suffering from PV for the last four years, presented to our Obstetric clinic. PV was suspected when she developed bullous eruption in her mouth and then on her body. It was confirmed by histopathology and demonstration of autoantibodies in her serum by indirect immunofluorescence. She was treated with high doses of systemic corticosteroids tailored to the disease activity. She had no menstrual problem and she conceived while still on treatment without any difficulty.

She had early pregnancy bleeding and episodes of severe vomiting which continued throughout pregnancy. Pregnancy follow up was otherwise normal regarding blood pressure examination, maternal weight gain and glucose tolerance test. She was on oral prednisolone initially 120 mg per day gradually tapering to 60 mg per day by 34 weeks gestation. There was no flare of her disease during pregnancy. She was admitted at

35-week gestation with history of premature rupture of membrane and went into spontaneous labor delivering a small for gestation 2.2 kg alive male fetus. The baby was healthy, without manifestation of pemphigus. Both mother and baby did well in the postnatal period.

### **Discussion**

Pemphigus is associated with difficulty in conceiving as other autoimmune disorders. In contrast to these data, our patient conceived during the active phase of PV, which required high doses of prednisolone, thus implying that active disease is not necessarily associated with infertility.<sup>3</sup> The effect of pemphigus varies during pregnancy. Our patient had preterm labour at 34-week gestation and premature rupture of the membranes. Premature deliveries are mainly related to high doses of systemic corticosteroid treatment as reported by Fainaru *et al.*<sup>3</sup> It can cause transient neonatal pemphigus or may result in a delivery of a healthy neonate completely free of the disease. The baby in the case presented was although born preterm but was healthy with no sign of the disease.

Although our patient had a normal vaginal delivery there is no consensus on the choice of delivery type in PV mothers. According to Goldberg<sup>6</sup> vaginal birth can result in worsening and spreading of the pemphigus while corticosteroids can delay wound healing in patients undergoing caesarean section. However, caesarean section is indicated in patients with pemphigus lesions on the genital mucosa. Neonatal PV occurs due to transplacental transmission of PV IgG antibodies from mother to fetus. Despite high incidence of reported neonatal PV our neonate was normal without manifestation of clinical disease. Neonatal PV is not reported to progress to adulthood and the

lesions in neonate tend to improve spontaneously.

It is essential to take preventive measures before conception to avoid complications. For example, effort should be made to taper or stop any immunosuppressive agent. The dose of prednisone 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 preexisting PV as it does in other autoimmune diseases. However, Lehman *et al.*<sup>7</sup> believe that adverse pregnancy outcome is more closely related to poor control of maternal disease and high titers of pemphigus antibodies than to particular medication.

Corticosteroids are the first choice treatment for PV. But if disease is not controlled with it, then steroid-sparing immunosuppressive agents may be added to therapy such as azathioprine. But its use during pregnancy should be avoided. Plasmapheresis can be used as treatment during pregnancy but its use is still experimental.

## References

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