CASE REPORT

A Rare Case of Management of Pemphigus Vulgaris in Pregnancy and Newborn

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Abstract

Pemphigus vulgaris (PV) is a rare autoimmune disorder that causes flaccid vesiculobullous lesions and erosions on the skin, with an annual incidence of 0.09–1.8% in India. Its occurrence during pregnancy is even rarer, with only 47 cases reported in the literature over a 49 years. The condition can worsen during the first and second trimesters and postpartum, with a phase of waning during the third trimester, which may be associated with endogenous corticosteroid secretion. The condition can affect various parts of the body, including pressure points, skin-to-skin contact areas, and oral and nasal mucosae. It can also lead to a transient benign form of neonatal lesions called pemphigus neonatorum due to transplacental transmission. Complications such as infertility and stillbirth can occur in women suffering from PV. This is a case of ART with IVF conception and managed successfully by our hospital, it was well-controlled with oral steroids and had an uneventful course in pregnancy, transient neonatal lesions also appeared after birth, which were managed successfully. Exacerbation postpartum was also seen but occurred due to non-compliance with corticosteroid therapy by the patient.

Keywords: Antenatal care, Bad obstetrics history, Case report, Fetomaternal outcomes, High risk pregnancy, Neonatal pemphigus, Pemphigus vulgaris, Pregnancy.

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Introduction

Pemphigus vulgaris (PV) is a rare dermatological disorder of autoimmune origin with an annualized incidence of about 0.68 in 100,000 in the UK. Similar studies in India, peg the incidence at around 0.09–1.8% for all pemphigoid disorders, and around 75–92% of those being PV.[1,2] The presence of PV in pregnancy is even rarer, with only 47 cases being reported in recorded literature over a 49-year period.[3]

Most cases are detected before pregnancy, and exacerbation may occur during the first and second trimester and postpartum, with a phase of waning during the third trimester.[4] This appears to be associated with neuroendocrine mechanisms responsible for endogenous corticosteroid secretion, which reaches a peak in the third trimester, falling after delivery.

The autoantibodies directed against desmoglein are responsible for all pemphigus disorders, the subtype desmoglein-3 is in the one implicated in PV. Diagnosis is dependent on detecting the IgG group of antibodies against these proteins being detected in the serum. Clinical manifestations include numerous skin vesicles, mostly resulting in widespread erosions that heal without scarring, as the erosions are entirely epidermal. Involvement may be localized or generalized but the predilection for pressure points and areas of frequent skin-to-skin contact such as scalp, face, axillae, and groins has been noted. The oral and nasal mucosae are often involved and can precede cutaneous lesions by months or may be the only manifestation of the disease.[3] Due to IgG antibodies, transplacental transmission leads to pemphigus neonatorum, which is a transient benign form of neonatal lesions of similar distribution and appearance. Infertility and stillbirth are complications that can occur in women suffering from PV, hence discussion of such cases is important.

Here we present a case managed by our hospital, which was well controlled on oral steroids, and neonatal lesions appeared after birth. Exacerbation postpartum occurred in this case as well, but it was mostly due to non-compliance to therapy by the patient.

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Patient consent statement: The author(s) have obtained written informed consent from the patient for publication of the case report details and related images.

Case Description

A 32-year-old G2P1NND1 with previous full-term vaginal delivery, present pregnancy conceived by IVF presented to our OPD for antenatal registration at 26 weeks of gestation. She had received prior antenatal care at a private hospital.

As per her history, she had a previous neonatal death, which occurred in 2018 before her diagnosis of PV, but the patient did complain of similar lesions then as well, hence there could be a possible relation between the two.

She was diagnosed with PV in 2019 following a positive Immunohistochemistry report for IgG antibodies against

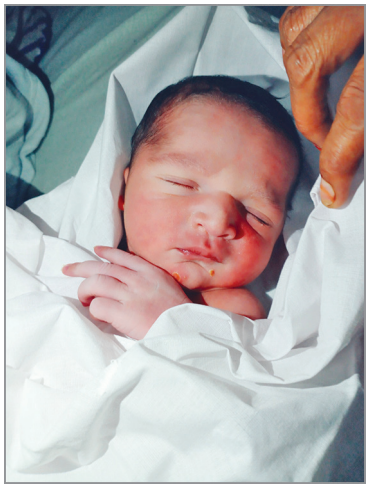

Fig. 1: Pemphigus in the neonate

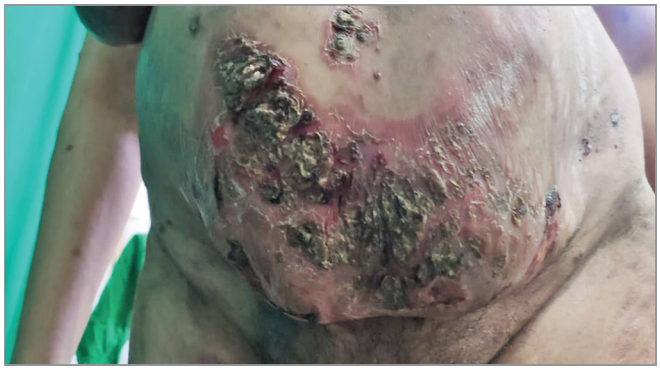

Fig. 2: Pemphigus vulgaris lesions in the mother's abdomen after flare up

desmoglein-3 and was on prednisolone 40 mg daily with good disease control.

On examination, the patient had flaccid vesiculobullous lesions predominantly in her oral mucosa with minor erosions covering the chest, back, and abdomen. She had a uterine size of 26–28 weeks, with a fetal heart rate of 136 beats per minute.

She was followed up weekly until 38 weeks, with regular home glucose monitoring and 3 monthly glycosylated hemoglobin (HbA1C), complete blood count (CBC), liver function tests, as well as prothrombin time (PT-INR). All parameters were normal during the follow-up (Fig. 1).

At 38 weeks, the patient was admitted to our hospital, and considering her past neonatal loss and her present IVF conception, we decided to offer her a short trial of labor by giving dinoprostone gel per vaginally, with hourly monitoring. Unfortunately, she underwent LSCS under GA due to meconium stained amniotic fluid associated with fetal distress. This resulted in the birth of a male neonate weighing 3405 gm (Fig. 2).

The patient was kept admitted for up to 14 days post-LSCS when suture removal was done and a healthy suture line was noted. Upon discharge, the patient was prescribed prednisolone 45 mg daily and potassium permanganate (KMnO4) gargles for her oral lesions to prevent infection. The neonate had transient neonatal pemphigus lesions that were treated with a combined fusidic acid and hydrocortisone cream.

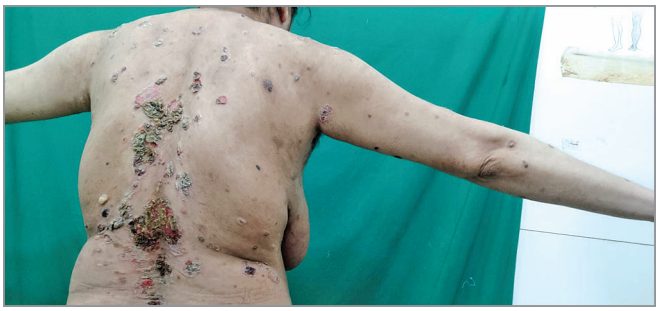

Fig. 3: Pemphigus vulgaris lesions in the mother's back after flare up

The patient was non-compliant with her steroid therapy due to fear of adverse effects of steroids through breastfeeding on her baby and had a PV flare-up that required rituximab therapy after breast suppression. The patient also had anxiety about her baby having PV in the future needing extensive treatments similar to her. She consulted a psychiatrist and was advised cognitive behavioral therapy (CBT) for the same. She is still undergoing follow-up treatment for PV in our hospital (Fig. 3).

Discussion

Pemphigus vulgaris is a rare autoimmune disease that affects the skin and mucous membranes and is characterized by the presence of intraepithelial blisters. Pemphigus vulgaris in pregnancy is a rare and complex condition, which presents unique challenges for both the mother and the fetus. The disease can worsen during pregnancy and may lead to a higher risk of fetal loss, preterm delivery, and neonatal complications.

Pemphigus can interact with pregnancy in the following manners:[4]

- Trans placental transfer of antibodies and pemphigus in the neonate
- Possible adverse effects of drugs transferred to the fetus
- Possible alteration of the course of pemphigus due to the immunologic and hormonal alterations seen in pregnancy
- The effects of the presence of a severe maternal disease on the fetus

In our case report, we presented a case of a 32-year-old pregnant woman with PV, who was treated with systemic steroids alone, prednisolone 45 mg. She had an IVF conception due to failure of ovulation induction and infertility associated with PV. She underwent a lower segment cesarean section, and the baby suffered from neonatal pemphigus, managed with local application of fusidic acid and hydrocortisone.

The mode of delivery in PV patients is a subject of debate. While some studies suggest that vaginal delivery is safe in stable PV patients, others recommend delivery by cesarean section due to the risk of fetal injury from maternal skin lesions and transfer of antibodies via the lesions. In our case, we opted for cesarean section due to the failure of induction of labor in a precious pregnancy and the need for close monitoring of the baby.

Neonatal pemphigus is a rare and potentially life-threatening condition that can occur in infants born to mothers with PV. The condition is caused by the trans placental transfer of maternal antibodies against desmoglein, which leads to the formation of

blisters and erosions on the infant's skin and mucous membranes. The condition can be mild or severe and may require treatment with topical or systemic corticosteroids.

In our case, the baby suffered from neonatal pemphigus, which was managed with local application of fusidic acid and hydrocortisone. Treatment of neonatal pemphigus is based on the severity of the condition and may include topical or systemic corticosteroids, as well as supportive care.

In conclusion, PV in pregnancy is a rare and challenging condition, which requires careful management to ensure optimal outcomes for both the mother and the fetus. Cesarean section may be considered in cases where there is a risk of fetal injury from maternal skin lesions, and close monitoring of the neonate is essential to detect and manage any neonatal complications, such as neonatal pemphigus.

Conclusion

In conclusion, unlike many cases and meta-analyses before this, the outcome of the pregnancy was positive. It is still imperative to closely monitor the patient and her fetus to prevent adverse outcomes. Early intervention with appropriate therapy may help control symptoms and achieve a near normal experience of pregnancy for such a patient.

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