A Rare Case of Recurrent Postpartum Pemphigoid Gestationis with Fetal Complications and Skip Pregnancy

Sir,

Pemphigoid gestationis (PG) is a rare, autoimmune, subepidermal bullous dermatosis, specific to pregnancy, which is clinically and pathogenetically similar to bullous pemphigoid, and occurs in approximately 1 in 50000 pregnancies.[1]

A 26-year-old female with obstetric code P5L2 (Para 5, Live 2), presented with fluid-filled lesions all over the body for more than a week. She had delivered a live baby boy of low birth weight (1.5 kgs) 15 days back. A four-year-old girl child born out of her third pregnancy was alive. Her birth weight was 2.6 kgs and the patient did not notice any skin lesions during that period. Other three previous pregnancies ended up in still births at around 7 months of gestation which were not attributable to any other particular cause. On examination, multiple well- to ill-defined urticarial plaques and fluid-filled blisters of 0.4 to 5 cm were seen over abdomen, chest, back, thighs, forearms, scalp, face, and hands [Figure 1a-d]. Nikolsky sign was negative and the Bulla spread sign was positive with rounded spread. Sheklakov sign was positive and there was no mucosal involvement. Tzanck smear showed predominantly eosinophils with few neutrophils. Histopathology showed subepidermal bulla with eosinophils [Figure 2a and b]. Direct immunofluorescence showed a strong linear band of C3 along dermoepidermal junction [Figure 3]. IgG was negative and the patient was started on oral prednisolone 40 mg with supportive measures. The lesions stopped appearing after 3 days. Prednisolone was gradually tapered and stopped after 4 weeks [Figure 4a-d].

PG is a rare immunobullous disease against BP180 in basement membranes of skin and amniotic epithelium of placental tissues, presents with intense itchy erythematous plaques and blisters. The possibility of recurrence in subsequent pregnancy is almost 90% and usually occurs earlier and more severe.[2] Studies have shown that PG may skip some pregnancies in 5% of patients. It was initially believed to be associated with a change in paternity or the expression of identical HLA-DR antigens by the mother and fetus.[3]

Another interesting finding is the high frequency of antibodies reactive to paternal MHC antigens in 85% to 100% of patients as opposed to 25% among normal multiparous women, whom were not associated with

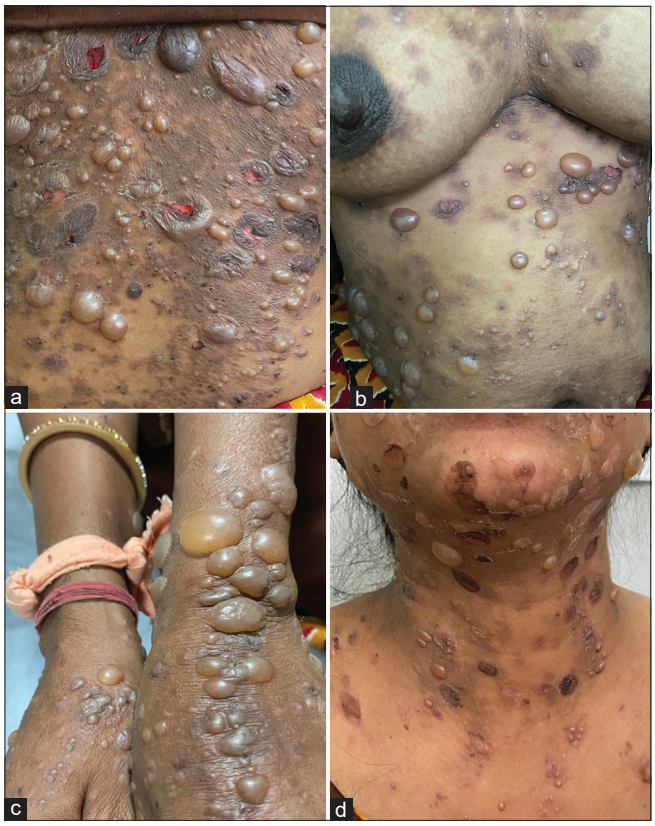

Figure 1: (a-d) Multiple fluid-filled blisters and urticarial plaques over abdomen, back, forearms and neck before treatment

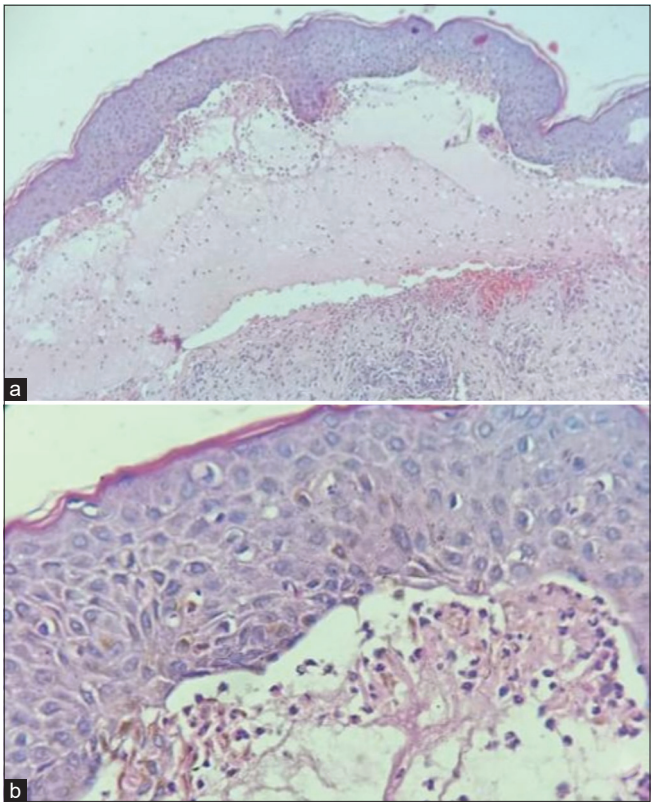

Figure 2: (a) Subepidermal split and eosinophils (H and E, 100X) (b) (H and E, 400X)

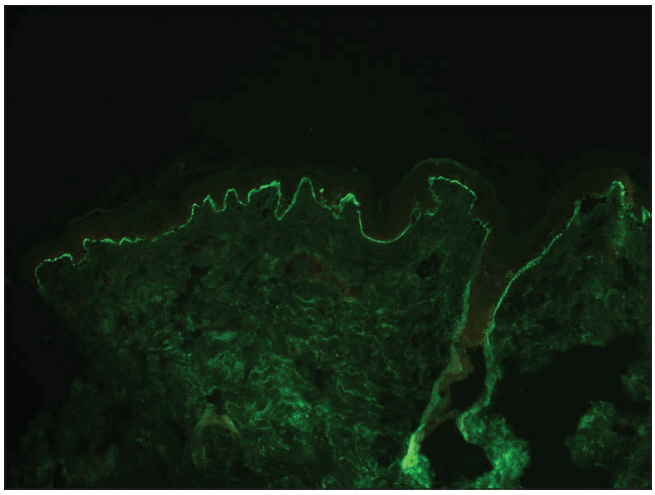

Figure 3: Direct immunofluorescence showing linear band of C3 along the dermoepidermal junction

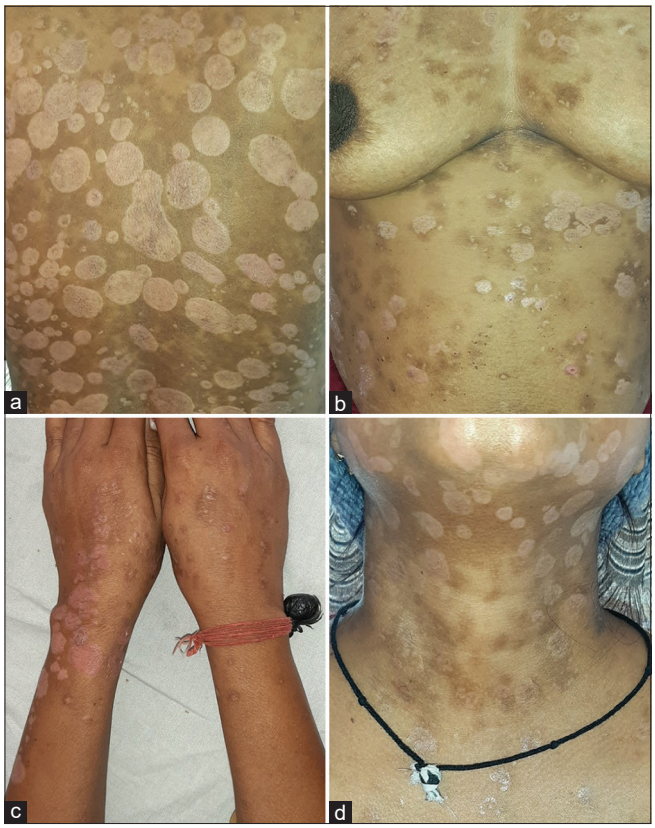

Figure 4: (a-d) Clinical photographs after treatment showing good improvement

any adverse pregnancy outcome.[4,5] In our patient, other causes of miscarriages like maternal smoking, diabetes, TORCH infection, and cholestasis were ruled out. Placental insufficiency due to preeclampsia could be the probable cause of stillbirths. Occurrence of PG in postpartum period is rare and its recurrence again in postpartum period is much rarer. Our patient had recurrent PG during postpartum period but was associated with fetal complications and hence being reported for its rarity.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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