

## Pemphigus Vulgaris: A unique clinical vignette in pregnancy

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### Introduction:

- Pemphigus is a group of immune-mediated bullous disorders that often cause fragile blisters and extensive skin or mucous membranes lesions.

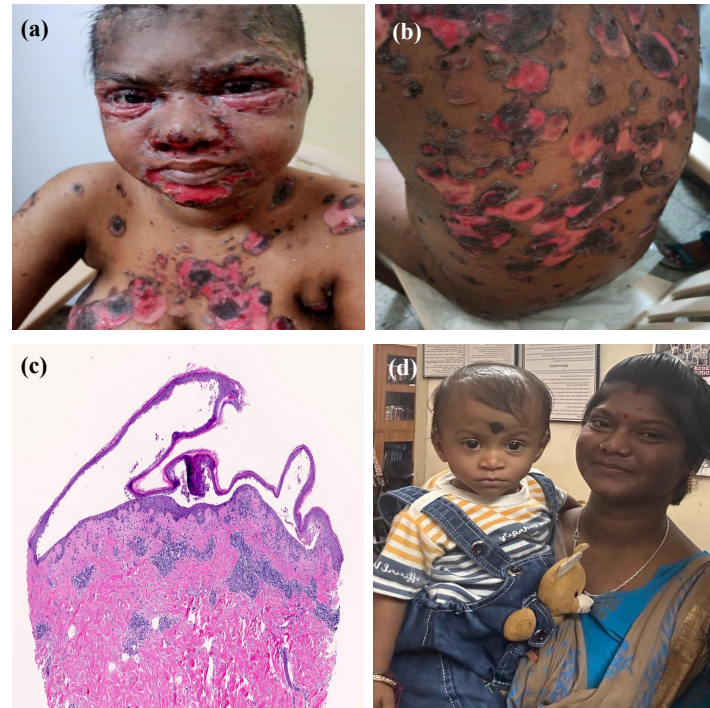
### Objectives:

- To outline the perinatal outcomes of active Pemphigus Vulgaris in pregnancy.

### Case Summary:

- 20 year primigravida, diagnosed elsewhere with Pemphigus Vulgaris after presenting with widespread fluid-filled blisters over her body.
- Had taken high doses of steroids at 8 to 12 weeks and MMF at 20 weeks due to aggressive disease.
- Presented to CMC at 30+1 wks with secondary infection of lesions – cultures grew Klebsiella and MSSA.
- Unresponsive to steroids → IVIg infusions with antibiotic cover.
- Developed steroid-induced hyperglycemia and hypertension, and eventually severe pre – eclampsia.

- Following MDT discussion with Derm, OBG, Endocrinology, Anaesthesia and Neonatology, she underwent Emergency LSCS (Ind: deteriorating maternal condition) at 31+5 weeks and delivered a 1.3 kg baby.
- She was discharged in remission with a healthy neonate.



(a) Large erosions with crusting seen over face and trunk (b) Similar appearance over back with post - inflammatory hyperpigmented macules (c) \*Histopathological image showing supra basal split (d) Patient and her baby at 6 months follow up

### Investigations:

- Skin biopsy - Suprabasal bullous formation
- Direct immunofluorescence – Positivity to IgG in the lower 2/3<sup>rd</sup> of the epidermis.

### Discussion:

- Being an autoimmune disease with dominant T Helper 2 cells activity, pemphigus is expected to flare in pregnancy, especially in the first two trimesters.
- Pemphigus in pregnancy is further complicated by the various drugs required in its treatment.
- Small possibility of transplacental transmission to the neonate.

### Conclusion:

Although Pemphigus Vulgaris in pregnancy is extremely rare, customized treatment options and care plans with a multidisciplinary approach can improve maternal and fetal outcomes.

### References:

De, Dipankar; et al.. Pemphigus and Pregnancy. Indian Dermatology Online Journal 15(5):p 749-757, Sep–Oct 2024.

\*<https://www.pathologyoutlines.com/topic/skinnontumorpemphigus.htm>

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