10.5005/jp-journals-10006-1202

CASE REPORT

Pemphigus Vulgaris with Pregnancy

G Nandini, K Umadevi, K Padma, Rajini Uday, R Sahana

ABSTRACT

Pemphigus vulgaris is an autoimmune bullous dermatosis affecting skin and mucous membrane.[2] It affects all races and both sexes equally. It is common during the 5th and 6th decade of life.[3] It is exceedingly rare in pregnancy[5] and is associated with increased maternal morbidity and poor neonatal outcome.[4] We do report a case of pemphigus vulgaris with pregnancy. She conceived during the active phase of the disease and treated with steroids throughout pregnancy. She delivered a live preterm, appropriate for gestational age and constitutionally small fetus.

Keywords: Pemphigus vulgaris, Autoimmune, Bullous dermatosis, Pregnancy.

How to cite this article: Nandini G, Umadevi K, Padma K, Uday R, Sahana R. Pemphigus Vulgaris with Pregnancy. J South Asian Feder Obst Gynae 2012;4(3):159-160.

Source of support: Nil

Conflict of interest: None declared

INTRODUCTION

Pemphigus vulgaris is an ultra rare autoimmune blistering disease. Though all races are affected, Jews are more susceptible.[1] Its common in eastern countries, like India, Malaysia and China and rare in west. In females, it may be associated with other autoimmune disorders and infertility.

Impact on the fetus may be due to the disease *per se* or because of the effect of immunosuppressive treatment given to the mother.

CASE REPORT

Mrs N aged 30 years referred to us from department of dermatology. She presented with history of multiple fluid-filled vesicles and blisters all over the body and mouth since 1 year. She gave history of amenorrhea of 8 months duration. She was gravida 3, para 2 with no living issues. She was married for 7 years and it was a nonconsanguineous marriage. In her first pregnancy, she had intrauterine fetal demise at 7th month and, in her second pregnancy, it was fetal demise at 8th month. The exact cause was not known.

Her menstrual history was regular with 3/30 day cycle. She was not able to recollect her last menstrual period date and no examination or ultrasound done in the first trimester.

She was diagnosed to have pemphigus vulgaris 1 year back. The diagnosis was confirmed by biopsy. She was on steroids, both oral and topical on and off.

On examination, her vitals were stable except for mild pallor. She was unable to lie down supine. Multiple vesicles and bullae over the axillae, chest, abdomen, groin, back and extremities were present. Whitish patches and erosions on tongue and buccal mucosa.

Per abdomen examination revealed uterus corresponding to 30 weeks of gestation and clinically liquor was adequate. All investigations were normal, including thyroid-stimulating hormone (TSH), GCT except for Hb% 10.2 gm% ultrasonography (USG) showed a live fetus with breech presentation corresponding to 33 weeks of gestation and adequate liquor.

She was on topical steroids. Dermatologist started her on parenteral steroids. She was discharged after 5 days with oral steroids and advised for regular follow-up. She came after 10 days with ruptured membranes and labor pains. On examination, she was having good uterine contractions with breech presentation. Per vaginal examination showed clear liquor draining and, on USG, amniotic fluid index (AFI) was 2 cm.

Emergency LSCS was done. There was difficulty in putting the incision as the lower abdomen was full of blisters. A live female baby was extracted weighing 1.9 kg with Apgar score 8, 10 and shifted to NICU. Baby had lesions over the legs which cleared after 4 days. She had a stormy postoperative period, gaping of wound and purulent discharge. She was treated with carbapenem broad-spectrum antibiotic after C/S report and steroids continued. Daily wound dressing done with biofil particles (sterile collagen type I in particle form sterilized by gamma irradiation which has a shelf life of 3 years). After complete wound healing, she and her baby discharged on 27th postoperative day in healthy condition (Figs 1A and B and 2A and B).

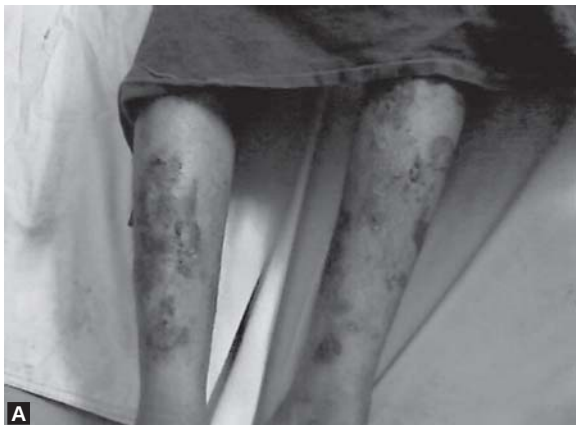
DISCUSSION

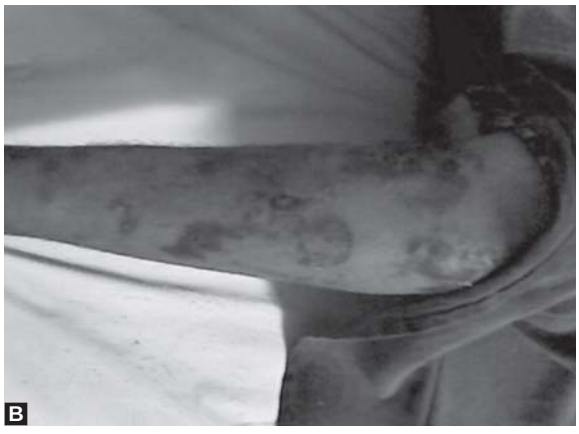
Pemphigus vulgaris is an autoimmune bullous dermatosis.[1] The autoantibodies are directed against desmoglein 3.[1] Desmoglein 3 is found in desmosomes and possibly on the cell membranes of keratinocytes. This leads to intercellular edema with loss of intercellular attachments in basal layer. Suprabasal epidermal cells separate from basal cells to form clefts and blisters.

Mucosal lesions precede cutaneous lesions. Most commonly, irregular, ill-defined buccal or palatal erosions occur and extend peripherally with shedding of epithelium. Cutaneous lesions are in the form of flaccid blisters. It may contain clear fluid and contents may become turbid or may rupture producing painful erosions which extend at the edges as more epidermis is lost. Healing occur without scarring.

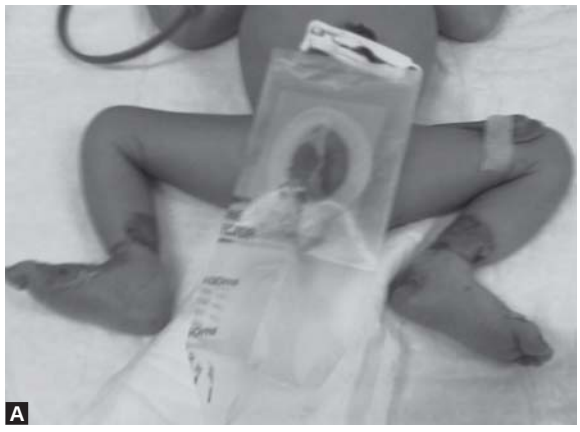
The dignosis is based on immunofluorescence and biopsy of the lesion. The severity and natural history of the disease are variable. Before the advent of steroids, the disease was fatal. Now, with steroids, the mortality has been reduced 5 to 15%. Complications in the mother include secondary infection, electrolyte imbalance and psychological stress as skin lesions are visible externally. Fetal complication may be prematurity and intrauterine fetal demise. This may be due to the disease *per se* or may be due to the immunosuppressive treatment given to mother.

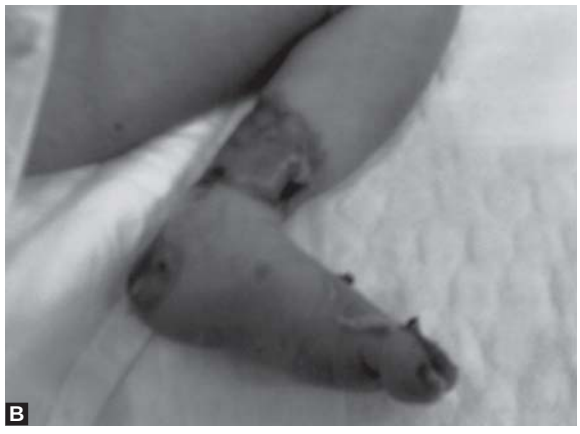
Transplacental transfer of maternal pemphigus vulgaris antibodies may cause transient blisters in the new born, which are short lived and will clear within days. The disease activity decrease with time and most relapses occur within 2 years of the diagnosis.





Figs 1A and B: Fourth postoperative day photographs, when the mother was psychologically better as baby was doing well in NICU: (A) Leg, (B) forearm (*Source:* MS Ramaiah Medical Teaching Hospital, Bengaluru, Karnataka, India)





Figs 2A and B: Second postoperative day photographs

Till now, about 40 cases are reported in literature, but immunopathologically proven cases are 28. Herpes gestationalis lesions resembles pemphigus vulgaris externally and hence immunopathological confirmation is a must.

CONCLUSION

Prognosis depends on the severity of lesions. Immunopathological confirmation helps to start immunosuppressive treatment. Conception during remission phase and close fetal surveillance may help in successful outcome.

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