University Journal of Medicine and Medical Sciences

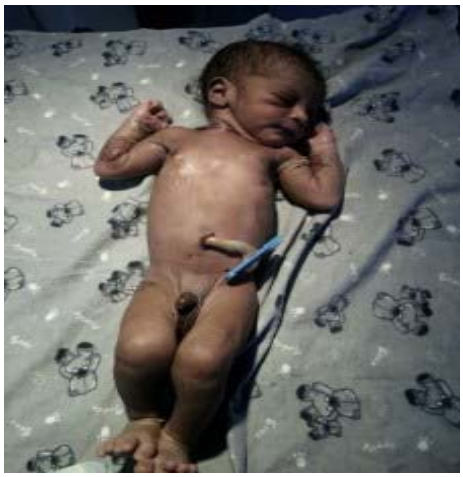
ISSN 2455- 2852 Volume 2 Issue 7 2016

Neonatal Pemphigus - A case report

ANITHA M
Department of Neonatology,
MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL

Abstract :
One of the rare differential diagnosis of blistering skin lesions occurring in newborns at birth is Neonatal Pemphigus. We report here a case of a male neonate born to a mother who was suffering from Pemphigus vulgaris, an autoimmune disorder during her second pregnancy. Mother had mucocutaneous lesions and mucosal involvement was more prominent whereas the infant had cutaneous manifestations. The disease in the mother flared up during the third trimester of pregnancy with increased oral mucosal involvement and was difficult to manage. The baby was growth restricted and developed fetal distress which needed resuscitation at birth.The newborn had multiple flaccid cutaneous bullae on the trunk and extremities which on rupture produced erosive lesions. There was no mucosal involvement. The lesions healed in 2-3 weeks time

Keyword :neonatal, pemphigus, autoimmune, desmoglein -3, India



Neonate at birth with lesions

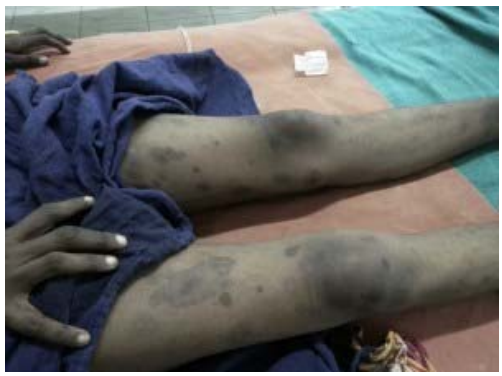
Introduction Neonatal Pemphigus is a rare cause of blistering conditions in newborns at birth. It occurs due to the transplacental transfer of maternal IgG autoantibodies formed against desmoglobin -3 and or desmoglobin-1. In a mother affected with pemphigus they are transferred through the placenta to the fetus who exhibit bullae at birth. There are two major types Pemphigus vulgaris and foliaceus.(1) Both types are said to be common among Jews, descendants from Middle east and Mediterranean with reported incidence of 0.1-0.5 per 1,00,000 people per year globally(2).

The Indian literature has reported more than 250 cases of adult pemphigus so far.(3,4)

Case report:

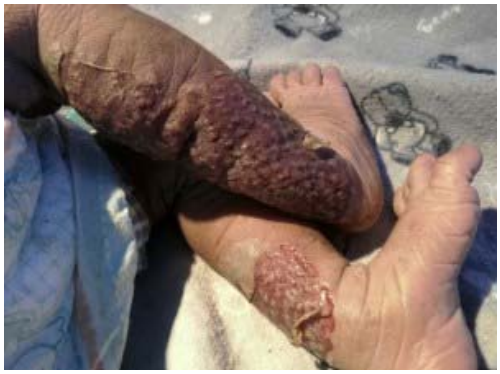
A term (38 weeks) male neonate delivered after an emergency cesarean section for fetal distress was noticed to have multiple flaccid cutaneous bullae spread over the trunk and both the extremities at birth. His mother had sudden onset of eruptions on the skin and mucus membranes during the third trimester of pregnancy. Her lesions were bullous with surrounding multiple papules and erosions and distributed over the skin of the face, chest, abdomen arms and legs. The lesions were varying in size. She was started on treatment with corticosteroids (Prednisolone 30mg per day). At 35 weeks of pregnancy her skin lesions flared up with vesicles and erosions in the face, buccal mucosa and lips and she was hospitalised. The disease was difficult to control and the dosage of corticosteroids was increased to 60mg per day. Her cutaneous lesions showed slow signs of healing and no new lesions appeared in the skin thereafter. But the oral mucosal lesions continued to increase inspite of escalating the dose of steroids

Maternal pemphigus - mucocutaneous involvement

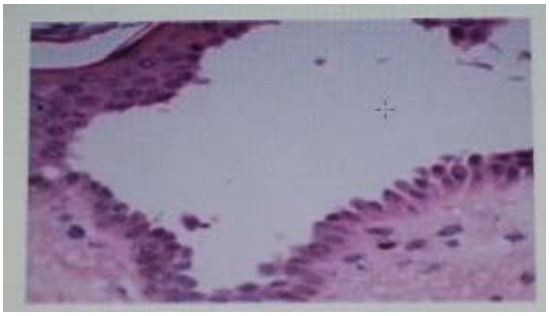


Maternal Pemphigus- cutaneous lesions

Towards term the neonate was born through meconium stained liquor and was non- vigorous at birth. He was resuscitated at birth with positive pressure ventilation. He was subsequently supported with oxygen and intravenous fluids. His anthropometric measurements revealed that he was small for gestational age and had features of in-utero growth restriction. The cutaneous blisters ruptured leading on to erosive areas. There was no involvement of the palms, soles or mucosa in the neonate.



Neonatal cutaneous manifestations



Skin biopsy showing intraepidermal bulla in the suprabasilar region and acantholysis

A differential diagnosis of herpes simplex, candidiasis, syphilis, infectious mononucleosis and epidermolysis bullosa were also considered. Tzanck smear was positive for acantholytic cells. Mother's VDRL test was negative. Her skin biopsy showed intraepidermal bullae and suprabasal clefts with irregular acanthosis based on which she was diagnosed to have Pemphigus vulgaris.

The infant was managed with warm saline compresses, topical antibiotics and breast feeds. Fluid input, output and electrolytes were monitored regularly. Neonatal skin biopsy also showed intraepidermal suprabasal bullae similar to maternal findings. All the lesions resolved at the end of the second week.

Healed lesions in the third week

Discussion

Pemphigus is an auto immune group of blistering disorders characterized by acantholysis (loss of adhesion between the keratinocytes) that results in the formation of intraepithelial blisters in mucous membranes and skin. In pemphigus IgG autoantibodies are directed against cell to cell adhesion molecules called desmosomes. Antibodies derived from PV patients bind both the desmosomes, predominantly desmoglein-isotype-3 (dsg3) and desmoglein-isotype-1 (dsg-1). These antibodies binding to the desmogleins disrupt cell to cell binding and cause acantholysis. The PV antigen, dsg3 is expressed predominately between cells of the deep, immediate suprabasilar region of the epidermis thus leading to the relatively deeper blister formation of PV. Additionally, mucosal sites have been shown to express significantly higher levels of dsg-3 relative to dsg-1, explaining the occurrence of mucosal blisters unique to PV.(5) Pregnancy is not uncommon among pemphigus patients in India as it occurs in a lower age group. (4) 8.5% of pemphigus patients became pregnant according to a retrospective study by Maryam Daneshpazhooh et al. Out of the 66 total pemphigus cases the diagnosis was made before pregnancy in 48 cases and during pregnancy in 18 cases. Pemphigus vulgaris was the most common type reported (85.4%). Exacerbation of the disease occurred during pregnancy in 54 % and the disease showed improvement in 17%. Postpartum flare occurred in 44% of cases. The disease was found to worsen predominantly during the first and second trimesters and the postpartum periods. (6)

The disease is said to be suppressed by the high endogenous chorionic steroids during the later gestation. In our patient the diagnosis was established based on the clinical features (mucocutaneous involvement) and histopathologic findings (intraepidermal cleft and acantholysis) during her last trimester of pregnancy. She was started on treatment with corticosteroids (Prednisolone 30mg/day) and required higher doses when the disease flared up later (60mg/day). Maternal pemphigus causes abortions, still births and premature births. In the recent English literature the rate of stillbirth was 10%, perinatal mortality 12% and abortions 9.7%. (6) This may be due to the disease per se or due to the immunosuppressive treatment given to the mother.

Neonatal pemphigus is a rare complication of pemphigus in pregnancy. Review of literature revealed that 40 cases of neonatal pemphigus have been reported so far. (7). In a large retrospective study on pregnancy outcomes in pemphigus patients the rate of neonatal pemphigus reported was 1.4% (single case).(6) In Indian literature three cases have been reported in the newborn period.(7,8,9) Majority of the infants have reported to have cutaneous manifestations. Desmoglein–3 is the predominant desmosome in the adult mucosa as well as neonatal skin. The desmoglein-1 is not present here to compensate for the destruction of the desmoglein-3 by its antibodies unlike the adult skin. Thus maternal pemphigus with predominant mucosal involvement due to the high titre of anti desmoglein–3 antibodies have higher incidence of neonatal pemphigus.(5, 10, 11, 12) The placental dysfunction either due to the disease per se or the treatment given for the disease is the usual manifestation seen during advanced pregnancy. Thus severe maternal disease predisposes to fetal growth restriction and poor neonatal outcomes.(13, 14)

The maternal pemphigus vulgaris antibodies transferred transplacentally may cause transient blisters in the newborn period. Unlike the maternal disease, these lesions have been reported to be short lived and found to clear within a few weeks. Thus the prognosis of neonatal pemphigus is very good. (9,15)

Conclusion:

In summary, we report a rare case of neonatal pemphigus born to a mother with an unusual course of this autoimmune disorder. Maternal pemphigus during the third trimester causes fetal growth restriction. Predominant mucosal manifestations in the mother predisposes to a higher incidence of neonatal pemphigus due to sharing of antigen between the adult mucosa and the fetal skin (desmoglein-3). The prognosis is very good with resolution of lesions completely by 3 weeks of life.

References:

1 Jacqueline P, Scott RF, Jason H, John Z, Kristin L, Shery V. Neonatal pemphigus in an infant born to a mother with serologic evidence of both pemphigus vulgaris and gestational pemphigoid. J Am Acad Dermatol. 2009; 60: 1057-62.

2 Michael H, Cassian S. Pathogenesis, Clinical Manifestations and Diagnosis of Pemphigus. Available from: URL: http//www.uptodate.com/store. Accessed July 7, 2013.

3 Sehgal VN. Pemphigus in India : A note. Indian J Dermatol Venereol. 1972; 18: 5-7.

4 Kanwar AJ, De D. Pemphigus in India. Indian J Dermatol Venerol Leprol. 2011; 77: 439-49.

5 Christopher A. D'Angelis. The Pathology of Pemphigus: A Mini-Review. North American Journal of Medical Sciences. 2013; 6(1):37-40.

6 Maryam Daneshpazhooh et al. Pemphigus and pregnancy – 23 years experience. Indian J Dermatol Venereol Leprol. 2011; 77: 534.

7 G Nandini, K Umadevi, K Padma, Rajini Uday, R Sahana. Pemphigus Vulgaris with Pregnancy. Journal of South Asian Federation of Obstetrics and Gynaecology. 2012; 4 (3): 159-160.

8 Sammera BKI, Yashodhara BM, Shashikiran U. Pemphigus vulgaris in a pregnant woman and her neonate. BMJ 2012; 20(13); 1-5.

9 Sheethal S Kodagali, SD Subbarao and Hiremagaloor Pemphigus Vulgaris in a Neonate and his Mother. Indian Paediatrics 2014; 51(15):316-17.

10 Dr. P. T. Chan. Review on Pathogenesis of Pemphigus Hong Kong Dermatology & Venereology. 2002;10 (2):62 -65.

11 Panko J, Florell SR, Hadley J, Zone J, Leiferman K, Vanderhooft. Neonatal pemphigus in an infant born to a mother with serologic evidence of both pemphigus vulgaris and gestational pemphigoid. J Am Acad Dermatol 2009; 60 (6): 1057-62.

12. Rafal Bialynicki-Birula, Marian Dmochowski, Joanna Maji,Justyna Gornowicz -Porowska. Pregnancy-Triggered Maternal Pemphigus Vulgaris with Persistent Gingival Lesions. Acta

13 Dermatovenerologica Croatalica 2011; 19(3):170-175.

Ruach M, Ohel G, Rahav D, samueloff A. Pemphigus and pregnancy. Obstet Gynecol Surv 1995; 50: 755-60.

14 Ofer Fainaru, Roy Mashiach, Michael Kupferminc et al. Pemphigus vularis in pregnancy: A case report and review of literature. Human reproduction 2000; 15(5): 1195-97.

15 Gushi M, Yamamoto Y, Mine Y, Awazawa R, Nonaka K,Taira K, et al. Neonatal pemphigus vulgaris. J Dermatol 2008 ;35: 529-35.