International Journal of Research in Dermatology
Singh SK et al. Int J Res Dermatol. 2018 Aug;4(3):441-443
http://www.ijord.com

DOI: http://dx.doi.org/10.18203/issn.2455-4529.IntJResDermatol20183173

Case Report

Neonatal pemphigus vulgaris

Santosh Kumar Singh, Aayushi Mohan*, Anil Kumar Gupta, Amit Kumar Pandey

Department of Dermatology, B.R.D. Medical College, Gorakhpur, Uttar Pradesh, India

Received: 02 November 2017
Accepted: 04 May 2018

***Correspondence:**
Dr. Aayushi Mohan,
E-mail: mohanaayushi@yahoo.in

ABSTRACT

Neonatal pemphigus is a transitory autoimmune blistering disorder caused by transplacental passage of maternal Auto antibodies to desmoglein to neonate through an affected mother. The association of maternal pemphigus vulgaris with neonate disease had been rarely reported. Herein, we report a case of neonatal pemphigus vulagris born to mother who conceived during active phase of disease.

Keywords: Neonatal pemphigus, Desmoglein, Acantholysis, Self limiting

INTRODUCTION

Neonatal pemphigus is a self limiting bullous disorder seen at time at birth. It is clinically characterized by transient flaccid blisters and erosions on skin and mucous membrane. It has an excellent prognosis as lesions are transitory and resolve within 3 weeks of birth.[1] Therefore it has no long term significance since after birth no new lesions appear.

CASE REPORT

A 1 day old male child, after 38 weeks of gestation, weighing 1.9 kg was born to a mother with pemphigus vulgaris. The child was reported to have multiple flaccid bullae filled with clear fluid on forehead, neck, elbow, groin, thigh and knees at the time of birth. The mother was diagnosed with pemphigus vulgaris 3 months before pregnancy by Tzanck smear and histopathological examination. She was treated for same but she discontinued the treatment on her own. The patient then came with active lesions and ANC 4 months. On admission the patient had multiple flaccid bullae on chest and extremities. She was managed on oral corticosteroid throughout the pregnancy by our side and monitored regularly by obstetrics department. The patient delivered a male child with active lesion by normal vaginal delivery. The infant was admitted in NICU for further management.

On general examination the child was lethargic febrile with HR 110 /min RR 45/min without signs of apnoea with feeble cry with absence of jaundice, without any bleeding from skin, mouth, rectum and without any apparent congenital malformation.

On cutaneous examination there were multiple flaccid bullae and erosions on forehead, neck, elbow, groin, thigh and knee which were well defined having normal skin in between. The lesions showed raw oozy surface with seropurulent discharge. The bullae were flaccid and filled with clear fluid (Figure 1-3). The scalp, hair, nail, palm and soles were normal. The conjunctiva, oral mucosa and genital mucosa were normal. On investigation Hb was 12.9 gm% TLC 14,400 cells/cc DLC P 64% l 34% e1% M 1%. Chest X ray was normal.

0n histopathological examination section showed intraepidermal blister that has formed in the upper spinous zone and containing few acantholytic

keratinocyte with few neutrophils. The epidermis adjacent to the blister showed mild incipient acantholysis a moderate to dense infiltrate of lymphocytes with few neutrophils present in upper dermis. Diagnosis of intraepidermal blister with acantholysis/neonatal pemphigus was made (Figure 4, 5).

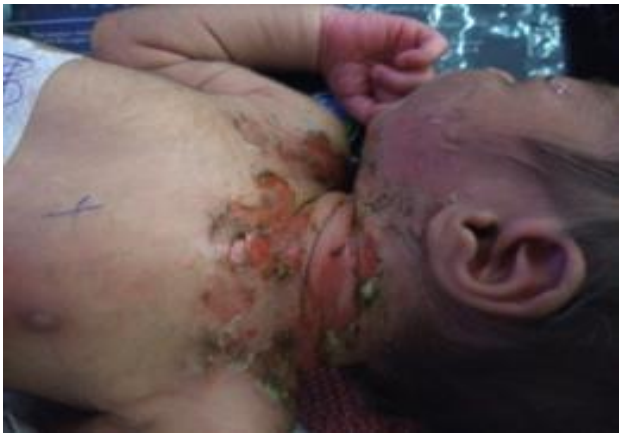

Figure 1: Showing neonatal pemphigus vulgaris lesions on the neck.

Figure 2: Showing neonatal pemphigus vulgaris lesions on the knee.

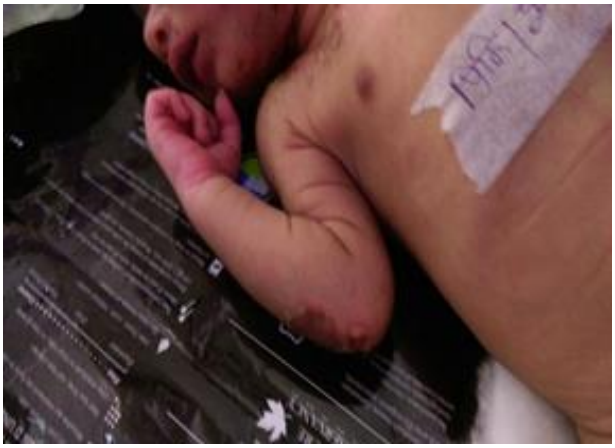

Figure 3: Showing neonatal pemphigus vulgaris lesions on the elbow.

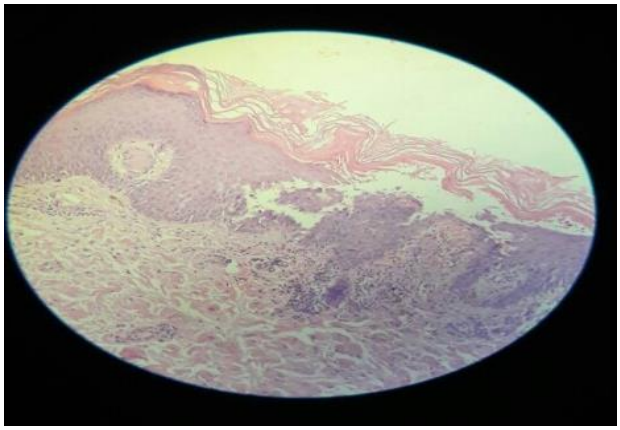

Figure 4: HPE showing intraepidermal blisters formed in the upper spinous zone and containing few acantholytic keratinocyte with neutrophils.

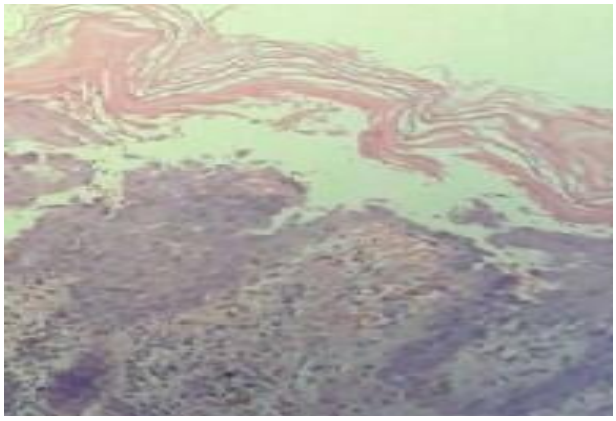

Figure 5: HPE showing intraepidermal blisters formed in the upper spinous zone and containing few acantholytic keratinocyte with neutrophils.

The child was suspected to have neonatal pemphigus based upon the distribution of lesions, maternal history and biopsy finding.

The child was managed on oxygen inhalation, barrier nursing, warm saline compresses and injectable antibiotics. Fluid input, output and electrolytes were monitored regularly.

DISCUSSION

Transplacental transmission of pemphigus vulgaris Ig-G antibodies from mother to foetus may result in clinical manifestations in the neonate.[2] The physiological variation in desmoglein in neonatal skin(dsg1 and dsg3 are present in upper layers while dsg3 is prominent in suprabasal layer) as compared to adult skin accounts for greater chance in pregnant women with pemphigus vulgaris delivering affected child then a pregnant women with pemphigus foliaceous.[3,4] Pemphigus vulgaris in pregnancy may result in abortion, still birth, foetal growth retardation, intrauterine death, premature delivery, and in approximately 30% neonatal PV of the newborn.[5,6]

Poor prognostic factor for foetal outcome include high ante partum maternal titre of autoantibody, increased maternal activity, maternal disease that requires high dose of corticosteroids and use of combined therapy.[7]

Diagnosis is based on lesion biopsy showing the presence of acantholysis and suprabasal cleft formation and deposition of immunoglobulin (Ig)-G and complement in the intercellular spaces of the epidermis. Immunoglobulin-G antibodies against the pemphigus antigen may be detected by indirect immunofluorescence in the serum.[8] Direct immunofluorescence shows IgG antibodies predominantly seen in intercellular spaces in epidermis giving green fish net appearance.[9]

The differential diagnosis of neonatal pemphigus congenital syphilis, herpes simplex, epidermolysis bullosa, and candidiasis was considered. The mother was TPHA and VDRL negative and IgG for HSV was negative. Therefore, the child was suspected to have neonatal pemphigus based upon the distribution of skin lesions, maternal history and skin biopsy.

Management of PV in pregnancy is similar to that in non-pregnant women. High dose prednisone (60–360 mg/day) for several weeks and gradual tapering to a maintenance dose is usually successful.[10] Although no increase in congenital malformations has been reported, corticosteroid treatment in pregnancy has been associated, with PROM and preterm births. Neonatal PV has not been reported to progress to adulthood, and if the lesions do appear on the neonate they tend to improve spontaneously within 3 weeks.[11]

Neonatal pemphigus is a rare complication of pregnancy in pemphigus patients. To our knowledge only 29 cases of neonatal pemphigus have been reported in the literature.[12-14]

Neonatal pemphigus is an uncommon presentation in neonates, hence a rarely found neonatal autoimmune bullous disorder.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Ian AG. Maternal Medicine: Medical Problems in Pregnancy, Dermatological disease in pregnancy, 1st ed. UK: Elsevier Health Sciences. 2007.
2. Wasserstrum N, Laros RK. Transplacental transmission of pemphigus. JAMA. 1983;249:1480–2.
3. Payne AS, Stanley JR. Fitzpatrick Dermatology in General Medicine. 8th edition. Chapter 54. 2012: 589.
4. Hilario-Vargas J, Vitorio IB, Stamey C, Culton DA, Prisayanh P, Rivitti EA, et al. Analysis of Anti-desmoglein 1 Autoantibodies in 68 Healthy Mother/Neonate Pairs from a Highly Endemic Region of Fogo Selvagem in Brazil. J Clin Exp Dermatol Res. 2014;5:209.
5. Shahir AK, Briggs N, Katsoulis J, Levidiotis V. An observational outcomes study from 1966-2008, examining pregnancy and neonatal outcomes from dialysed women using data from the ANZDATA Registry. Nephrology (Carlton). 2013;18:276–84.
6. Meurer M. Pemphigus diseases in children and adolescents. Hautarzt. 2009;60:208–16.
7. Lehman JS, Mueller KK, Schraith DF. Do safe and effective treatment options exist for patients with active Pemphigus vulgaris who plan conception and pregnancy? Arch Dermatol. 2008;144:783–5.
8. Daniel Y, Shenhav M, Botchan AM, Peyser R, Lessing JB. Pregnancy associated with pemphigus. Br J Obstet Gynecol. 1995;102:667–9.
9. Darling MR, Daley T. Blistering mucocutaneous diseases of the oral mucosa-a review: Part 2. Pemphigus vulgaris. J Can Dent Assoc. 2006;72:63-6.
10. Hertl M, Eming R, Veldman C. T cell control in autoimmune bullous skin disorders. J Clin Invest. 2006;116:1159–66.
11. Chowdhury MM, Natarajan S. Neonatal pemphigus vulgaris associated with mild oral pemphigus vulgaris in the mother during pregnancy. Br J Dermatol. 1998;139:500–3.
12. Gushi M, Yamamoto Y, Mine Y, Awazawa R, Nonaka K, Taira K, et al. Neonatal pemphigus vulgaris. J Dermatol. 2008;35:529-35.
13. Amer YB, Al Ajroush W. Pemphigus vulgaris in a neonate. Ann Saudi Med. 2007;27:453-5.
14. Panko J, Florell SR, Hadley J, Zone J, Leiferman K, Vanderhooft S. 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.

Cite this article as: Singh SK, Mohan A, Gupta AK, Pandey AK. Neonatal pemphigus vulgaris. Int J Res Dermatol 2018;4:441-3.