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Dermatology Case Reports

Case Report

Neonatal Pemphigus in Homozygous Twins: A Rarity

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ABSTRACT

Pemphigus Vulgaris (PV) is a chronic immunobullous disorder which affects the skin and mucous membranes. A 30-year-old lady, a proven case of oral pemphigus which was well controlled with corticosteroids gave birth to monozygotic twins at 28 weeks of gestation, delivered through Lower Segment Caesarean Section (LSCS).

Keywords: Neonatal; Pemphigus; Homozygous

INTRODUCTION

Pemphigus Vulgaris (PV) is a chronic immunobullous disorder which affects the skin and mucous membranes. Though **Pemphigus Vulgaris** is the most common form among the pemphigus group of disorders, neonatal pemphigus is rare. There are only a handful of case reports in the literature regarding neonatal pemphigus [1,2]. Here we are presenting a case of neonatal pemphigus, in monozygotic twins.

CASE REPORT

- A 30-year-old lady, a proven case of oral pemphigus which was well controlled with corticosteroids gave birth to monozygotic twins at 28 weeks of gestation, delivered through Lower Segment Caesarean Section (LSCS). Dermatologists were called immediately after delivery in view of skin lesions in the neonates.
- Twin I, weighing 1.4 kg had areas of raw moist erosions with crusting over anterior aspect of neck, right post auricular area, abdomen above the umbilicus and posterior aspect of left leg and twin II weighing 1.3 kg had areas of raw moist erosions over left temporal area of the scalp. Perilesional Nikolsky sign was positive in both. Hair, nail and mucosa were normal. No other abnormalities were noted on physical and systemic examination.
- Tzanck smear from the erosions showed a few acantholytic cells. Histopathological examination from one of the representative skin specimens showed eosinophilic spongiosis with intraepidermal vesicle containing a few acantholytic cells. Direct immunofluorescence showed intercellular deposits of IgG in the lower epidermis in a lace like pattern. Indirect immunofluorescence showed circulating IgG antibodies with a titre of 1:10.

Both of them were treated symptomatically with a course of antibiotics, saline compress and combination of topical antibiotic with corticosteroid cream. The lesions regressed in a span of 2 weeks and healed with milia formation. Babies are on regular follow up with no evidence of new skin lesions (Figures 1-12).

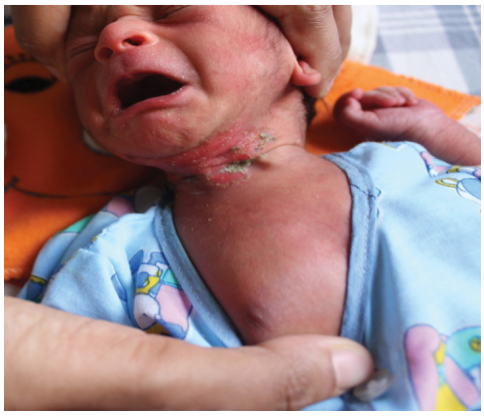

Figure 1: Twin 1 erosions over neck.

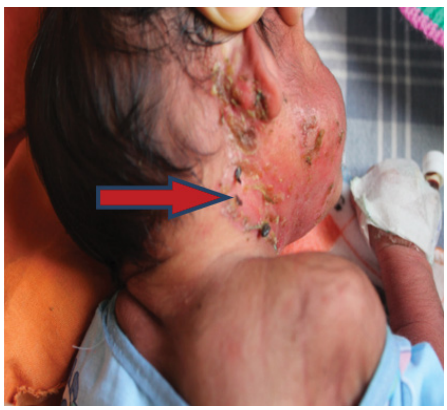

Figure 2: Twin 1 crusted plaque over neck.

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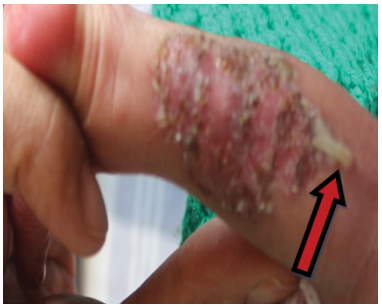

Figure 3: Twin 1 large crusted plaque with hypopyon on posterior aspect of leg.

Figure 4: Twin 1 large crusted plaque with hypopyon on abdomen.

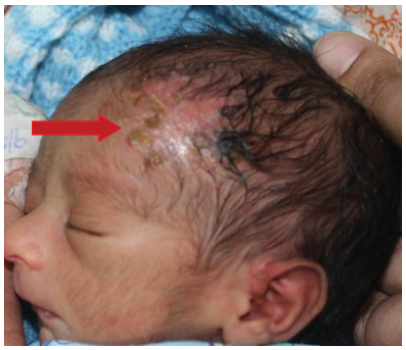

Figure 5: Twin 2 crusted plaque with an erosion on scalp.

Figure 6: Twin 1 and twin 2.

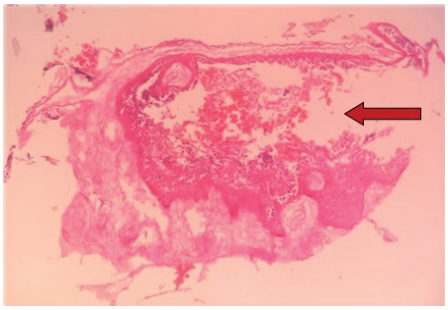

Figure 7: Scanner view showing intraepidermal split.

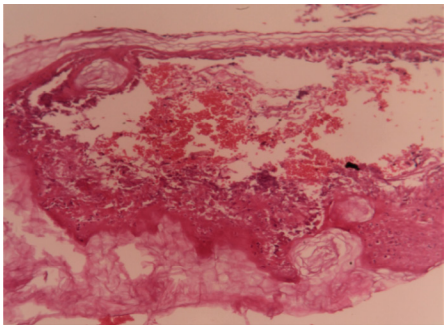

Figure 8: Low power view showing intraepidermal split with eosinophilic spongiosis.

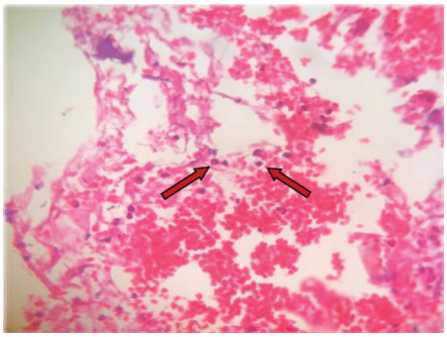

Figure 9: High power view showing acantholytic cells.

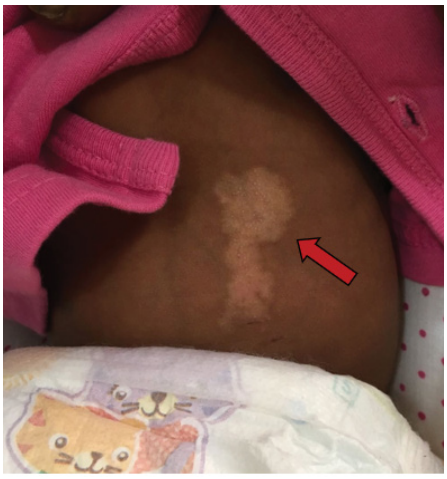

Figure 10: Healed with post inflammatory hypopigmentation with milia formation over abdomen.

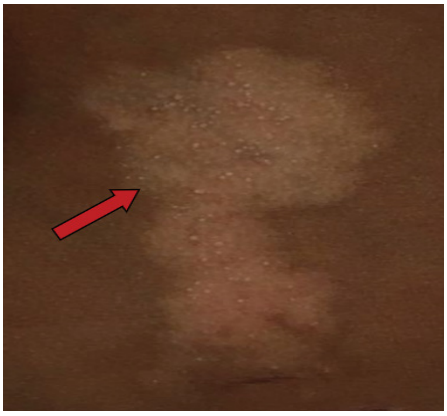

Figure 11: Healed with post inflammatory hypopigmentation with milia formation over abdomen (Closer view).

DISCUSSION

Neonatal pemphigus is caused by transplacental transfer of IgG antibodies from mother suffering from PV to her foetus [2,3]. These

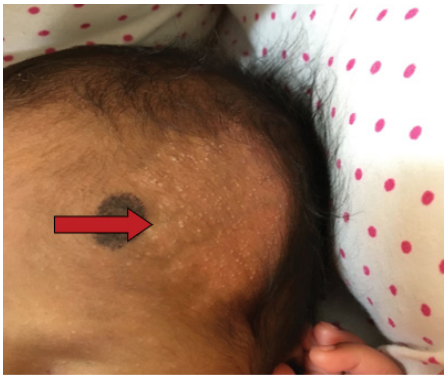

Figure 12: Healed with post inflammatory hypopigmentation with milia formation on scalp.

antibodies bind to neonatal skin after crossing the placenta, and may cause blisters. Neonatal PV has a varied clinical manifestation ranging from mild to widespread cutaneous involvement [1,2]. The lesions may heal spontaneously, as it is due to transplacental transfer of antibodies which gets degraded [4]. The sub corneal keratinocytes in newborn infants contain both Dsg1 and Dsg3, whereas the suprabasal layers contain much lower levels of Dsg1, with Dsg3 constituting most of the strength of the intercellular desmosomal bridge. So, in a pregnant woman with PV there are higher chances of delivering an affected child than pemphigus foliaceous [3]. Mother to child transmission resulting in neonatal pemphigus can be as high as 30% [5]. Histopathology and direct immunofluorescence findings of neonatal pemphigus is similar to pemphigus vulgaris [6] as seen in adults.

There are very few case reports regarding this entity showing varied clinical manifestations ranging from only oral involvement to widespread denuded skin, which can be observed at birth [3]. The clinical and histological variations in presentations can be explained by desmoglein compensation theory, however some atypical presentations can be attributed to the pathogenic strength of immunoglobulin IgG against Dsg3 [7]. The degree of involvement may vary from case to case and may lead to premature deliveries, still birth and/or intrauterine death [1-4]. Antibody titres in the mother or newborn at the time of clinical presentation do not correlate with the severity of neonatal PV [6]. There are also reports describing skin lesions in neonates with antibody titres as low as 1:20 or undetectable levels as seen in our case [8]. Indirect Immuno Fluorescence (IIF) positivity depends on both the quantities of anti-Dsg1 and anti-Dsg3 antibodies in the test serum and the relative expression of Dsg1 and Dsg3 in the epithelial substrate [9].

The immunoassays available for the serological diagnosis of PV include DIF, IIF, ELISA, immunoblotting and new biochip mosaic indirect immunofluorescence. The new biochip mosaic IIF technique combines the screening of autoantibodies and target antigen-specific substrates in a single miniature incubation field and has the highest sensitivity and specificity with all autoimmune bullous disorders unlike traditional IIF [10-13]. This new biochip mosaic technique might help in diagnosing challenging cases. There are reports of neonatal pemphigus born to mother exhibiting features of mucocutaneous pemphigus and having antibodies to antigens responsible for pemphigus as well as those responsible for gestational pemphigoid [14].

The lesions usually heal within 2-3 weeks and are self-limiting, topical antibiotics can be considered if clinically indicated [1,3,4]. Careful handling of the baby is utmost important. Progression of disease to adult PV has not been reported so far [2,5].

CONCLUSION

Neonatal pemphigus is a rare entity and here we are describing neonatal pemphigus in monozygotic twins which has never been described so far in the literature.

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