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Pemphigus Vulgaris in a Neonate and his Mother

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Background: Neonatal pemphigus is a rare, transient blistering condition due to transplacental transfer of maternal autoantibodies. **Case characteristics**: A male neonate born to a mother with oral pemphigus was noticed to have multiple lesions. **Observation**: Multiple flaccid bullae were noticed on the face, scalp, trunk and extremities with clear fluid and few areas of erosions. **Outcome**: All lesions resolved at the end of one week with conservative management. **Message**: Maternal pamphigus may rarely involve her newborn infant; it resolves on its own.

Keywords: *Autoimmunity, Neonatal pemphigus vulgaris, Transplacental transfer.*

Pemphigus is a group of autoimmune blistering disease of skin and mucous membranes [1]. Incidence rates between 0.1 and 0.5 per 100,000 people per year have been reported [2]. Neonatal pemphigus is a transient autoimmune blistering disease caused by transfer of maternal IgG autoantibodies to desmoglein-3 through the placenta when the mother is affected with pemphigus [3,4].

CASE REPORT

A 1-day-old boy weighing 2900 grams, born after full term pregnancy, was noticed to have multiple flaccid bullae on the face, scalp, trunk and extremities with clear fluid and few areas of erosions. All lesions showed a rim of erythema and abrupt demarcation from the surrounding normal skin (***Fig.* 1**). These lesions were not restricted to trauma-prone areas. A few lesions had profuse serous discharge. Nails and oral mucosa were not involved. Mother of this infant was diagnosed to have oral pemphigus vulgaris 8 months before conception, documented by incisional biopsy from buccal mucosa and direct immunofluorescence test (DIF), and was on daily oral steroids. The child was suspected to have neonatal pemphigus vulgaris based on the morphology

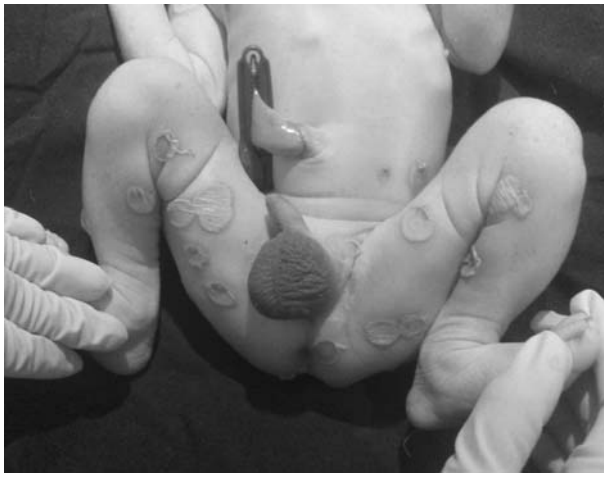

FIG.1 *Flaccid bullae and crusted erosions with an erythematous rim distributed over the groin area in the neonate.*

and distribution of skin lesions, and maternal history. A differential diagnosis of herpes simplex, candidosis, syphilis, infectious mononucleosis and epidermolysis bullosa were also considered. Tzanck smear was negative for multinucleated giant cells and acantholytic cells. Mother's VDRL test was negative. Skin biopsy was deferred as the lesions were drying up on second day. Child was managed with warm saline compresses, barrier nursing, topical antibiotics and breast feeds supplemented with formula feeds. Intravenous fluids were not required. Fluid input, output and electrolytes were monitored regularly. All the lesions resolved at the end of one week.

DISCUSSION

Pemphigus is defined as a group of life-threatening blistering disorders characterised by acantholysis (loss of keratinocyte to keratinocyte adhesion) that results in the formation of intraepithelial blisters in mucous membranes and skin. The process of acantholysis is induced by circulating autoantibodies to intracellular adhesion molecules [2]. Patients with pemphigus develop mucosal erosions and/or flaccid bullae, erosions, or pustules on skin. Neonatal pemphigus is a very rare transient form which occurs as a consequence of placental transmission of autoantibodies to the fetus from the mother. Maternal Pemphigus causes premature births and still births, with rare occurrence of neonatal pemphigus. The prognosis is very good with resolution of lesions completely by 3 weeks of life [5].

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