PASSIVELY TRANSFERRED PEMPHIGUS IN A NEWBORN

J S Pasricha, K A Seetharam and Meharban Singh

A 24-year-old patient having pemphigus for the last 1 year delivered a female baby after 36 weeks of gestation. The baby had pemphigus-like lesions on the skin and a single ulcer on the hard palate. The serum antibody titres of intercellular antibodies in the mother and the baby at the time of delivery were 1:80 and 1:40 respectively. The lesions in the baby healed completely by the 9th day after birth, without any specific treatment. Three months later, the antibody titre in the mother was 1:40, while in the baby, the test became negative. The baby has since shown no recurrence of the lesions.

Key words: Pemphigus, Pregnancy, Trans-placental transfer, Neonatal pemphigus.

Pemphigus is now well accepted to be an autoimmune disease, mediated by IgG antibodies directed against the epidermal intercellular substance.1 The titres of these antibodies show a good correlation with the clinical activity of the disease² and these are capable of inducing acantholysis on the epidermal cell cultures.⁸ Passive transfer of these antibodies in neonatal mice has been shown to result in pemphigus-like disease.4 Since IgG can readily pass through the placenta, causation pemphigus-like lesions in the newborn of mothers having pemphigus is a unique natural situation indicating the pathogenic nature of the autoantibodies seen in pemphigus. Recently, we followed a pregnant pemphigus patient, whose newborn baby had pemphigus-like lesions on her skin at birth.

Case Report

A 24-year-old lady had been having pemphigus vulgaris for the last 1 year. In addition, she was pregnant for the last 8 months. This was her second pregnancy. Her first pregnancy had ended in an abortion at 10 weeks of gestation, one year ago. During the current antenatal period, her pemphigus activity was kept under control with a daily oral dose of 10 to 20 mg of

From the Departments of Dermato-Venereology, and Pediatrics, All India Institute of Medical Sciences, New Delhi-110 029, India.

Address correspondence to : Dr J S Pasricha.



Fig. 1. Multiple, superficial erosions on the scalp, ₹ face, neck and upper chest.

prednisolone. At the end of 36 weeks, she gave birth to a 2.3 kg female baby, with one-minute Apgar Score of 9. The placenta was normal and did not show any calcification. The baby at birth had multiple erosions on the scalp, face, neck and upper chest (Fig. 1). Distal ends of both the feet were macerated. In addition, there was an erosion on the hard palate. Nikolsky's sign was positive. Tzanck smear from an erosion showed acantholytic cells. Skin biopsy, unfortunately missed the split, and direct immunofluorescence could not be done. The antibody titres in the mother and the baby (cord-blood) were 1:80 and 1:40 respectively. The baby was given oral prophylactic antibiotic therapy. Corticosteroids were not given. There was no evidence of cortisol

withdrawal symptoms or Addisonian crisis in the baby. Within 12 hours, the erosions became dry, and healing started by the 3rd day. All the lesions healed completely by the 9th day, and there has been no recurrence of the skin lesions in the baby during a follow-up of 6 months. Titres of intercellular antibodies in the mother and the baby repeated 3 months after the delivery were 1:40 in the mother, and negative in the baby.

Comments

Manifestations of the maternal autoimmune diseases in the newborn have been reported in lupus erythematosus, myasthenia gravis, thyrotoxicosis and idiopathic thrombocytopenic purpura, in addition to pemphigus. The clinical lesions in all these cases are because of the passive trans-placental transfer of the maternal autoantibodies during the antenatal period. So far, there have been 10 reports of pemphigus vulgaris and pregnancy, involving 13 babies.⁵-⁷ Of these, 8 babies had skin lesions, one had oral lesions only, while 4 were unaffected. Of the 9 babies with pemphigus lesions, 3 were stillbirths, while the remaining 6 were born live. The clinical lesions in the 6 live born babies subsided by 1-2 weeks. In 3 of these, the serum antibody titres were done (1:20 in 2 cases and 1:80 in 1) and these became negative by 3 to 8 weeks. We have had two previous experiences with pemphigus and pregnancy, one of these resulted in a still-birth, while in the other case, the baby was clinically normal. The present case was the first patient where clinical lesions were demonstrable at birth. It has been reported that active pemphigus during pregnancy is not necessarily associated with pemphigus in the newborn, because some of the children born to pemphigus patients were unaffected. Though the deciding factor(s) for the development of the clinical lesions in the newborn are not clearly known, the severity of the disease and the maternal antibody titre may be one of the important factors. After the delivery, as the passively transferred maternal antibody titre falls, the baby with the lesions improves rapidly even without any specific treatment. Transplacentally acquired pemphigus has not been reported to recur at a later age.

References

- Anhalt GJ, Patel H and Diaz LA: Pemphigus vulgaris, in: Pathogenesis of Skin Disease, Editors, Thiers BH and Dobson RL: Churchill Livingstone. New York, 1986.
- Sams WJ Jr and Jordon RE: Pemphigus antibodies: their role in disease, J Invest Dermatol, 1971; 56: 474-479.
- Schiltz JR and Michel B: Production of epidermal acantholysis in normal human skin in vitro by the IgG fraction from pemphigus serum, J Invest Dermatol, 1976; 67: 254-260.
- 4. Anhalt GJ, Labib RS, Voorhees JJ et al: Induction of pemphigus in neonatal mice by passive transfer of IgG from patients with the disease, New Eng J Med, 1982; 306: 1189-1196.
- Moncada B, Kettelson S, Hernandes-Montezume JL et al: Neonatal pemphigus vulgaris: Role of passively transferred pemphigus antibodies, Clin Res, 1980; 28: 251A.
- Storer JS, Galen WK, Nesbitt LT et al: Neonatal pemphigus vulgaris, J Amer Acad Dermatol, 1982; 6: 929-932.
- Merlob P, Metzker A, Hazaz B et al: Neonatal pemphigus vulgaris, Pediatrics, 1986; 78: 1102-1105.