Pemphigus Vulgaris Associated with Pregnancy

A Case Report from Japan

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We present a case of pemphigus vulgaris which developed during pregnancy. The newborn infant was normal. Bullous lesions were successfully treated by pulse therapy with high-dose corticosteroids. This is, to our knowledge, the first report in English from Japan describing pemphigus vulgaris associated with pregnancy.

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Pemphigus vulgaris is an uncommon autoimmune bullous dermatosis occurring most frequently during the fifth and sixth decades of life (1). The diagnosis is confirmed histopathologically by suprabasal cleft formation and immunopathologically by IgG fluorescence in the intercellular spaces of the epidermis (2). Blistering is suppressed by adequate corticosteroid therapy.

Although pregnancy is accompanied by profound metabolic and hormonal changes, occasionally associated with a variety of skin disorders, coexistence of pregnancy and bullous dermatosis is unusual (3).

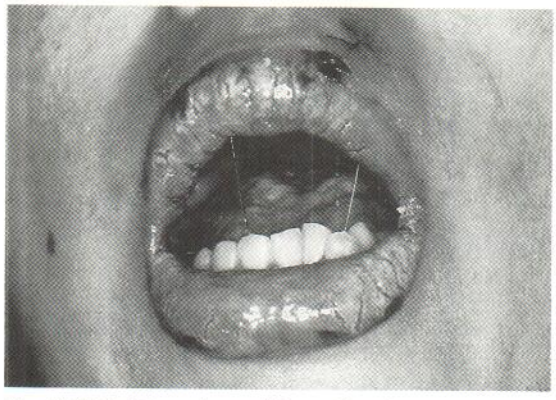

Fig. 1. Whiteish erosions of lips and oral mucosa.

Consequently, there have been only a few reports of pemphigus vulgaris during pregnancy (4). We describe here a case of pemphigus vulgaris that developed late during pregnancy, but was cured with pulse therapy with a large dose of corticosteroids.

CASE REPORT

A 37-year-old pregnant woman, gravida IV, para III, developed painful erosions of the lips and the oral mucosa (Fig. 1) at 8 months' gestation. She had no serious past history except for hydatidiform mole. Blistering erythema and erosions extended not only to the axillae, perineum, hands and feet, but also to other areas, including the scalp, nipples, umbilicus and back. The patient was referred to us at 9 months' gestation and was immediately hospitalized. Nikolsky's sign was positive. A biopsy specimen of the oral lesions revealed suprabasal acantholytic bullae formation (Fig. 2). Direct immunofluorescence microscopy revealed IgG and C_3 in the intercellular space (ICS) of the epidermis (Fig. 3). An indirect immunofluorescence study also showed IgG ICS antibody deposition in a titre of 1:20. Routine laboratory examinations disclosed no abnormal values, while HLA-A10 was not detected. A diagnosis of pemphigus vulgaris was established.

The patient subsequently underwent induction of labour, and a normal female infant was born. Four days after delivery the mother began treatment with oral prednisolone, 60 mg daily to counteract new bullae formation. Improvement in the lesions, however, was obtained within 2 weeks with subsequent pulse therapy with 1000 mg/day of methylprednisolone i.v. for 3 days, followed by a daily oral dose of 40 mg prednisolone. Gradual reduction of corticosteroid administration has not provoked any flare-up of pemphigus vulgaris to date for at least 2 years.

DISCUSSION

Thirteen maternal pemphigus cases have been recorded since 1969 (4–11). Of these, nine newborns had skin or oral mucosal lesions and three of the affected babies were stillborn. The Japanese literature contains a case of maternal pemphigus in which the infant developed bullous lesions (12). In all of the 13 reported cases, maternal pemphigus developed during the last trimester. Wasserstrum & Laros

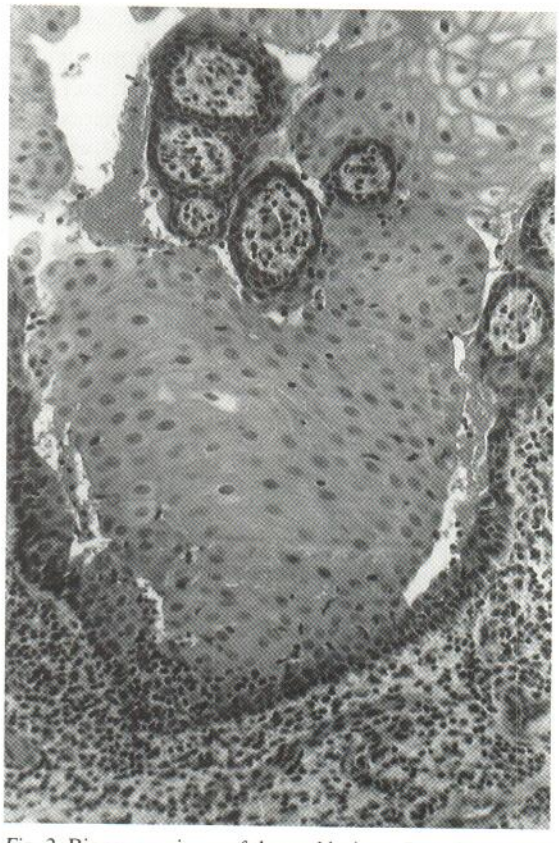

Fig. 2. Biopsy specimen of the oral lesions. Suprabasal cleft is evident (hematoxylin-eosin, ×200).

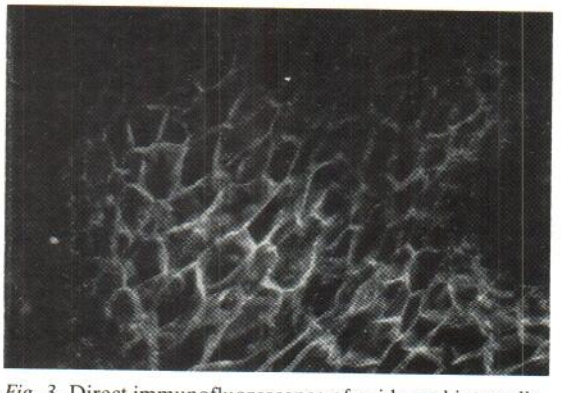

Fig. 3. Direct immunofluorescence of epidermal intercellular IgG deposits (×200).

(10) reported a pregnancy complicated by maternal pemphigus vulgaris, in which indirect immunofluorescence showed serum from the umbilical cord blood to be positive for intercellular substance antibody. Furthermore, several reports have proved, using an immunofluorescence technique, that pemphigus IgG can be transplacentally transmitted to fetal circulation and attack the fetal epidermis (8, 9, 11, 13). Kelly et al. (14, 15) studied antigen binding by using human amnion as the substrate in indirect immunofluorescence, and demonstrated linear BMZ staining with sera from patients with herpes gestationis, the most common bullous disorder in pregnancy (16). This phenomenon was observed in both full-term and second-trimester (but not in first-trimester) placentae. These findings show that maternal pemphigus vulgaris as well as herpes gestationis tends to develop during the late pregnancy stage and can be transplacentally transmitted in the manner of an autoimmune bullous dermatosis.

Our patient had a healthy infant. She was HLA-A10 negative, despite a significant increase in HLA-A10 positive Japanese pemphigus patients (13). Her clinical course improved as a result of pulse therapy with a large dose of corticosteroids. Consequently, this case shows the effectiveness of this therapy for inveterate pemphigus vulgaris.

We also think that pulse therapy may be useful for managing active pemphigus vulgaris, and should be introduced whenever the disease becomes serious, even during the gestational period, in view of the fetal and maternal risks posed by pemphigus vulgaris.

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