

Case Report

Transient Neonatal Autoimmune Blistering in Pemphigus Complicating Pregnancy

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ABSTRACT

Maternal pemphigus can result in a spectrum of clinical manifestations in her offspring, including prematurity and still-birth. Neonatal pemphigus is a rare transient blistering condition due to transplacental transfer of

maternal antibodies. It seldom requires specific treatment. The case of a baby with large skin erosion which healed spontaneously within six days is reported.

KEYWORDS: autoimmunity, neonate, pemphigus vulgaris

INTRODUCTION

A number of chronic dermatological disorders may complicate pregnancy. Pemphigus vulgaris (PV) is one such blistering disorder of autoimmune etiology. The disease is generally rare in childhood.

Because pemphigus is usually a disease of middle age, its occurrence during pregnancy is a rare event. Mothers with pemphigus can occasionally produce offsprings with erosive blisters over large areas of the skin. A case of a baby born with widespread erosion over the chest is reported. The entire lesion subsided in six days without treatment with topical or systemic steroids.

CASE REPORT

A term appropriate for date female baby was born of a non-consanguineous marriage by vaginal delivery. Her mother was 24 years old. She was diagnosed to have PV at 19 years of age and had been on continuous treatment with oral prednisolone 10 mg twice daily since then. She was free of skin lesions throughout pregnancy.

The baby had extensive erosion of the skin over the entire anterior chest (Fig. 1). The lesion had an erythematous base and an abrupt demarcation from the surrounding normal skin. There was profuse serous discharge from the surface. The erosion measured 14 cm and 12 cm in the maximal horizontal and vertical dimensions respectively. Mucosal surfaces and nails were normal. Her scalp, hair and the skin over other body parts also appeared normal.

A biopsy of the lesion with surrounding normal skin demonstrated scattered areas of acantholysis.

Indirect immunofluorescence test of serum was negative. She was treated with supportive measures alone. Meticulous attention was paid to fluid balance and prevention of infections. Topical mupirocin cream was applied twice daily for a week. The erosion healed rapidly and completely by the sixth day of life (Fig. 2). She is under follow-up and doing well.

DISCUSSION

PV is a rare intraepidermal blistering disease characterized by autoimmunity to specific proteins^[1]. It has a worldwide annual incidence between 0.1 and 0.5 per 100,000 in the general population^[2]. The disease principally affects patients in the fourth, fifth and sixth decades^[3]. A predilection of the disease for women is observed in patients under the age of twenty^[4]. Severe maternal pemphigus is known to be associated with fetal prematurity and death^[5]. Rarely, babies born to mothers with PV may display clinical, histological and immunopathological features of the disease, as in the present case^[6].

Transplacental passive transfer of maternal anti-intercellular antibodies to the fetus is a well established, albeit rare, phenomenon^[3]. All four classes of IgG are known to cross the placenta. Because only 5% of circulating IgG in the neonate is produced by its own immune system, the majority of antibodies in such babies are of maternal origin^[7]. These antibodies are thought to be responsible for the neonatal blisters and erosions. The blisters are usually flaccid and occasionally large. Erosions seldom occur in adults with pemphigus. The

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Fig. 1: Extensive erosion over the entire anterior chest noted at birth



Fig. 2: Complete healing of the lesion by the sixth day of life

extensive erosion over the entire anterior chest wall in the present case is possibly due to the effect of mechanical trauma of vaginal delivery resulting in the rupture of a large flaccid blister.

The diagnosis is usually obvious because of concurrent disease or a positive history in the mother. The mother in our report had a history of blisters from 19 years of age and was on continuous steroids. Very few instances of transient neonatal autoimmune blistering have been reported in the literature. Tope *et al* described a baby born with PV to a mother who was in complete remission throughout pregnancy^[8]. They reviewed ten reported cases and found that there were three stillbirths. All seven neonates who survived showed healing of the lesions within a period of three weeks with either no specific treatment or use of topical antibiotics. In contrast, Goldenberg *et al* documented a mother with active disease and an antibody titer of 1:640 whose baby was free of lesions^[9].

Circulating anti-intercellular antibodies in the serum of neonates as well as a positive immunofluorescence have been identified in such babies^[6]. However, the latter may frequently yield a negative result, as in the present case. Both these were positive in the present case as well. Biopsies of lesional skin reveal rounding up of epidermal cells with loss of cell adhesions, a pattern referred to as acantholysis. Biopsy specimen of our patient also revealed a mild form of this typical picture.

The prognosis of neonatal pemphigus is variable^[3]. Ross *et al* reviewed the literature in 1986 and found a 14% still-birth rate in neonatal pemphigus, compared with 1% in controls^[10]. In neonates who develop blisters, the lesions are known to resolve within a few weeks of birth, with one study showing normalization of immunofluorescence by six weeks^[6]. In our baby, the entire lesions healed within a period of six days.

Unlike the case with maternal PV, use of topical or systemic steroids is not indicated in neonatal blisters, as the latter is a transient phenomenon. Tackling of fluid loss and prevention of infections are the two most important aspects in the management of such babies. The role of prophylactic topical antibiotics is controversial. Hence we decided to give it the benefit of doubt and used topical mupirocin in our patient till the lesion healed completely. Whether it had any positive influence on the rapid healing of the erosion or prevention of infection remains a debatable point especially as the literature reports neonatal lesions to be transient.

The fact that disease activity in the mother does not correlate well with the severity of neonatal involvement makes counseling affected mothers difficult^[3]. However, the rarer instances of fetal deaths and prematurity seem to carry a lower risk rate when compared to most reports which have documented neonatal disease to have a mild and uneventful course. The purpose of this report is to emphasize upon the self-limiting nature of this apparently alarming condition in neonates. Clinicians must be aware of this rare entity and unnecessary steroid use must be avoided in such babies.

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