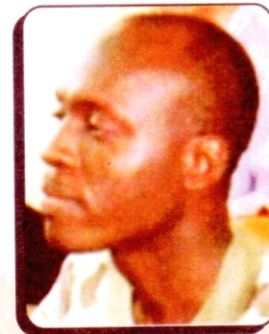


Neonatal Pemphigus: A Case Report Of A Baby Born To Mother With Pemphigus Vulgaris



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ABSTRACT

Neonatal pemphigus is a rare sequelae following maternal pemphigus. It is due to transfer of maternal IgG auto antibodies to desmoglein 3 to the neonate through the placenta when the mother is affected with pemphigus.

We describe this case of neonatal pemphigus in a baby born to a 35year old mother who was managed for pemphigus in pregnancy.

Key words: pemphigus,antibodies,neonatal

INTRODUCTION

PEMPHIGUS vulgaris is a blistering skin disorder of auto-immune aetiology. This rare intra-epidermal blistering disease is characterized by auto immunity to specific proteins [1]. It has a general worldwide annual incidence between 0.1 and 0.5 per 100,000 in the general population [2].

Neonatal pemphigus is also rarely reported with few cases documented. It is known that the baby acquires the antibodies through the placenta when the mother is affected with pemphigus. The implicated antibodies are IgG auto antibodies to desmoglein 3 [3]. Neonatal pemphigus has never been reported to persist beyond neonatal period and progress to adult disease. The neonatal disease seldom requires specific treatment.

The first case of neonatal pemphigus was documented in 1975 and there have been other cases since then [4]. The clinical presentation of neonatal pemphigus also varies from widespread denuded skin to only oral involvement which is observed at birth. There have also been cases of preterm deliveries in some mothers with pemphigus vulgaris [5].

There are reports of neonatal pemphigus vulgaris in newborns of mothers with no active disease as well as cases of non affected neonates born from mothers with highly active disease. [6, 7, 8, 9]

CASE PRESENTATION

A 35year old Nigerian woman was admitted into the hospital at 21weeks of gestation following complaints

of bleeding per vagina and was subsequently diagnosed as Type IV placenta praevia. She had a background history of subtle recurrent mouth ulcers for about 3years which resolved with use of prednisolone tablets but with no definite diagnosis made as at then. She had no history of blisters on the skin prior to this hospital admission.

It was after a month on admission that she began to have blisters on her scalp and on her groin. The blisters rapidly worsened with secondary bacterial infection (Fig 1). She had biopsy done on the lesions on her scalp and in her mouth and the result was in keeping with pemphigus vulgaris. Histology showed marked suprabasal acantholysis with dyskeratosis and hyperkeratosis. The suprabasal epidermis showed atypia, with papillary elongation into the upper dermis and eosinophilic spongiosis in the lower epidermis. This acantholytic cleft was overlain by nuclear debris and few atypical keratinocytes.

Other basic tests were carried out on her e.g. complete blood count, Electrolytes and they were all within normal limits.

She was placed on tablets prednisolone 20mg daily, clindamycin 150mg qds for one week based on wound swab mcs which yielded significant growth of *Staphylococcus aureus*. The oozing scalp wound was also washed with potassium permanganate daily.

Diprosone ointment (Bethamethasone dipropionate) was prescribed for the lesions on her groin and these resolved within two weeks of treatment.

She remained in stable clinical condition following resolution of her skin and oral lesions though she was kept on admission for bed rest and fetal wellbeing monitoring.

She was eventually delivered of a live female infant weighing 3.25kg by elective caesarian section at a gestational age of 37weeks.

At birth, the baby was found to have denuded skin on the gluteal area, some blisters on the neck and erythematous patches on the scalp with some erosions around the patches. (Fig 2, 3)

Dermatology review of the baby was of a diagnosis of neonatal pemphigus and mother was counseled on the self limiting nature of the condition. The wounds were cleaned with normal saline and emphasis was on adequate fluids and calories for the baby. However at 3 days after birth the skin lesions showed signs of impetiginisation and a topical antibiotic mupirocin was added and used for five days. No steroid preparations were applied. The skin lesions on the baby had healed remarkably by 7th day when mother and child were discharged in stable clinical condition. By the time baby was reviewed again at 3weeks after birth, all the skin lesions had healed completely (Fig 4).

DISCUSSION

Pemphigus vulgaris is a disease that affects patients in the fourth, fifth and sixth decades [10]. Neonatal pemphigus is rare but babies born to mothers with pemphigus vulgaris may display clinical, histological and immunopathological features of pemphigus vulgaris [11]. Skin biopsy was not done in the baby born to the mother with pemphigus vulgaris in this case simply due to preference of the mother to defer such a procedure. However the skin lesions which were seen in the baby right after birth were classical.

Although reports of fetal prematurity and death have been documented in babies of mothers with pemphigus vulgaris, the baby born in this case was delivered at term in spite of the dual morbidity suffered by the mother i.e. placenta praevia type IV and pemphigus. This may be attributed to the close monitoring and management given by the team of specialists who attended to her.

The antibodies which are responsible for neonatal pemphigus are known to be of maternal origin as only 5% of circulating IgG in the neonate is produced by its own immune system and these antibodies are responsible for the blisters and erosions [6]. The effect of these maternally acquired antibodies is transient and this is why neonatal pemphigus has never been reported to persist beyond neonatal period to progress into adulthood [3]. This case presented further highlights this well held fact. The mothers of these babies are usually worried and knowledge of this fact will help in counseling the parents of such children and help avoid use of medications which may eventually prove to be harmful.

Immunofluorescence was not done in this case because the technology is not readily available in the country for skin histology. We had to use our clinical acumen in addition to the basic histology in our low resource

setting. This is a shortcoming in our health system which requires appropriate intervention

The management of neonatal pemphigus is different from adult pemphigus in the sense that steroids are not required in the management of neonatal pemphigus[12]. This case also emphasises that neonates do not require steroids in the

management of neonatal pemphigus. This would help to avoid the side effects that may arise due to unnecessary exposure to steroids. Attention to fluids and electrolytes is however mandatory and should be emphasised.

The role of topical antibiotics in the management of neonatal pemphigus is controversial.¹² Topical mupirocin was administered in this case when the lesions showed signs of being secondarily infected. It is advised that antibiotics be used when their use is justified.

This case has been documented to further strengthen the body of knowledge as regards neonatal pemphigus. The purpose therefore is to emphasise the transient and self limiting nature of this condition so that neonates born with this condition can be well managed without exposure to drugs which may not be required hence avoiding anxiety in the mothers as well as unwarranted side effects of medications.

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Fig. 1 Showing infected lesion of pemphigus on mother's scalp



Fig. 2: Neonate with ulceration in gluteal region



Fig 3: Neonatal pemphigus with ulcer on the lower neck



Fig 4: Healed lesions of same neonate at 3 weeks of life managed without steroids

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