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CASE REPORT

Neonatal lupus erythematosus presenting as Stevens-Johnson syndrome

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ABSTRACT

Neonatal lupus erythematosus (NLE) is a rare acquired condition caused by the transplacental passage of maternal autoantibodies. It is characterized by cutaneous, cardiac, hepatobiliary, hematological and neurological involvement. Cutaneous findings of NLE are variable, but few reports in the literature describe the presence of erosions or epidermal loss in NLE. Herein, we describe the case of an infant with NLE presenting as Stevens-Johnson syndrome (SJS), with cardiac, hematologic, hepatobiliary and neurologic abnormalities. The characteristic features by which to differentiate SJS-like NLE from infant SJS include subacute course, cardiac abnormalities, positive serology tests and direct immunofluorescence, histopathologic findings with melanin incontinence, periadnexal infiltrates, and mucin deposit. Though rare, it is important to consider lupus erythematosus as a potential cause of acute syndrome with focal epidermal necrosis or pan-epidermolysis.

Introduction

Neonatal lupus erythematosus (NLE) is a rare acquired condition caused by the transplacental passage of maternal anti-Ro (SS-A), anti-La (SS-B), or anti-U1RNP antibodies. Cutaneous involvement occurs in roughly 70% of infants; congenital heart block is the most common extracutaneous manifestation, occurring in approximately 60% of cases.[1] Other systemic findings including hematologic, hepatobiliary and neurological abnormalities can also be found.

The mean age for the development of this skin disease is 5–6 weeks, although around 20% of patients have cutaneous eruptions at birth.[2] Cutaneous findings of NLE are variable, but the most typical is annular or polycyclic, scaly erythematous plaques occurring mainly on the face and scalp. These lesions usually resolve spontaneously within weeks to months, leaving residual dyspigmentation and atrophic or scarring lesions. However, only four reports in the literature describe the presence of erosions or epidermal loss in NLE.[2–5] In this report, we describe the case of an infant with NLE presenting as cutaneous Stevens-Johnson syndrome (SJS)-like eruptions, as well as with associated cardiac, hematologic, hepatobiliary and neurologic abnormalities.

Case report

A 5-week-old baby boy presented with a progressive skin rash over his scalp, face, trunk and four limbs for two weeks (Fig. 1A and B). He was delivered at 30 weeks gestation via cesarean section due to bradycardia with pericardial effusion and weighed 1698 g. Congenital atrioventricular block was diagnosed at birth and a permanent pacemaker was implanted the following day. Temporary empiric antibiotics including ampicillin, gentamicin, oxacillin and cefotaxime were used in the first week for prophylaxis of possible infection due to preterm delivery of the patient. The mother had a history of two miscarriages and was diagnosed as highly suspect for LE due to the presence of positive autoantibodies (ANA, anti-ds DNA, anti-Ro, anti-La). Although not meeting the diagnostic criteria of SLE, anti-phospholipid syndrome or mixed connective tissue

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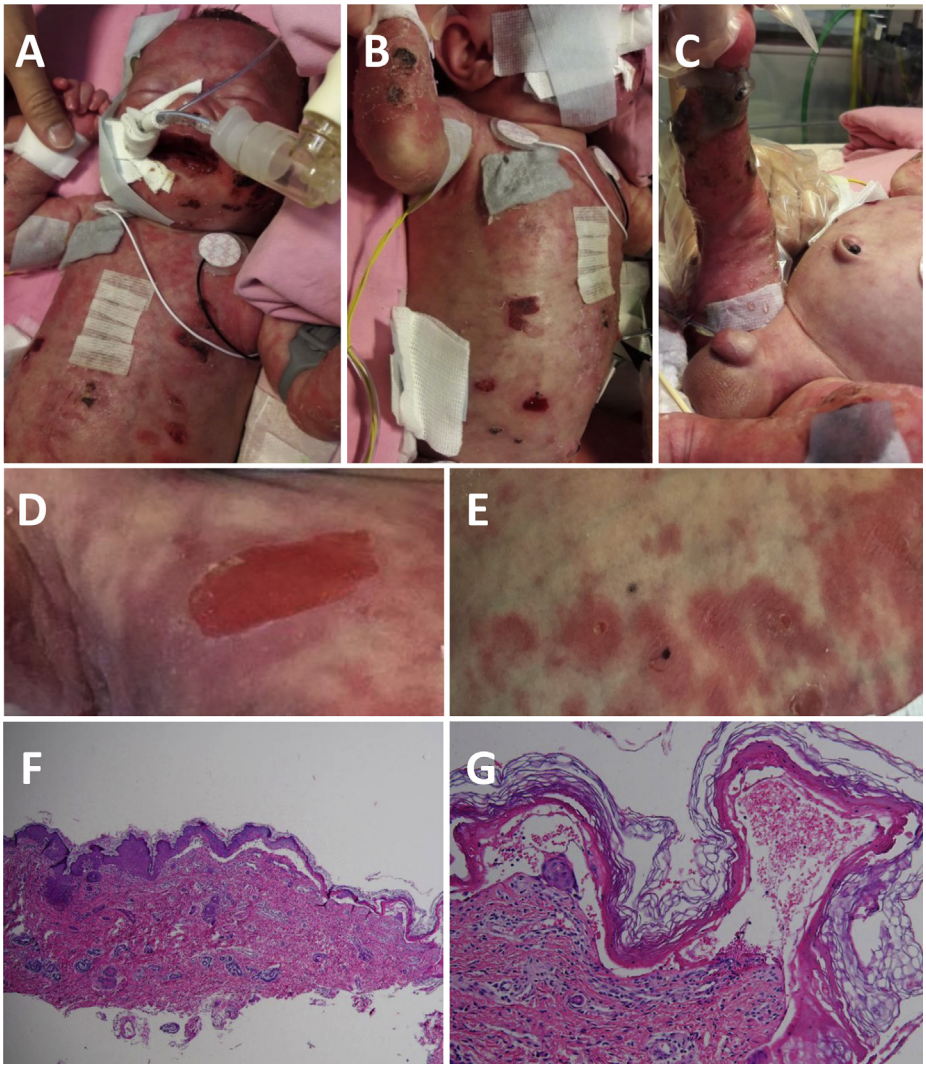

Fig. 1 Multiple reticulated erythematous to dark red patches with crusts on face (A), trunk (B), and multiple flaccid bullae on right lower leg (C). Central sheet-like erosions (D) and denudation (E) were found. Histopathological examination showed basket weave hyperkeratosis, many dyskeratotic cells, vacuolar degeneration of basal cells, partially detached necrotic epidermis, and lymphocytes at interface and around the adnexal structures (F, H&E x40; G, H&E x200).

disease, she had started to receive therapy of oral corticosteroids and hydroxychloroquine three months before delivery because of the immunologic abnormalities which suggested LE.

Physical examinations of the infant revealed multiple reticulated erythematous to dark red patches with central sheet-like erosions and crusts (Fig. 1A and B) on the scalp, face, trunk and buttock, as well as multiple flaccid bullae on the right lower leg (Fig. 1C). The lesions over the upper and lower extremities, especially on the palms and soles, were confluent with purplish change. Meanwhile, mucosal lesions with lip erosions and genital involvement were also found. The lesions had developed two weeks earlier, with annular and polycyclic lesions with erythematous margins and lighter-colored centers on the trunk and lower limbs.

Laboratory tests showed mild thrombocytopenia (103,000/mm^3), elevated liver function (AST/ALT (118/48 IU/L)), and hyperbilirubinemia (direct/total bilirubin (2.4/3.1 mg/dl)). Further tests indicated that the child was positive for anti-Ro, anti-La antibodies, negative for anti-U1RNP antibody and hypocomplementemia (C3: 48.90 mg/dl and C4: 5.76 mg/dl). Abdominal ultrasonography results were normal. Cranial ultrasonography revealed a left subependymal hemorrhage and right subependymal cyst.

A skin biopsy was obtained from one dark red patch on the infant's abdomen. Histopathology examination showed basket weave hyperkeratosis, partially detached necrotic epidermis, many dyskeratotic cells, vacuolar degeneration of basal cells and lymphocytes at interface and around the adnexal structures (Fig. 1D and E). No evidence of mucin deposits in the dermis was noted. The direct immunofluorescence results were negative. The histopathological findings were suggestive of NLE in the context of clinical features, image and laboratory abnormalities.

The patient was then treated with systemic steroids for the following 19 days. Aquacel-Ag dressing and betamethasone-gentamicin cream were prescribed for skin care. The lesions healed with residual mottled dyspigmentation and slightly atrophic lesions within two weeks. The blood dyscrasia disappeared and hepatobiliary function returned to normal by fourth months old.

Discussion

Herein, we report a case of NLE as the unusual presentation of SJS. NLE is a multisystem disease with variable cutaneous

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