Coexistence of annular polycyclic, morpheaform and atrophic lesions in neonatal lupus erythematosus

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Accepted 18 October 2022

DESCRIPTION

Neonatal lupus erythematosus (NLE) occurs due to transplacental transfer of autoantibodies in newborns of mothers with clinical or subclinical collagen vascular diseases. Anti-Ro/SSA antibodies are strongly associated with NLE. Anti-La/SSB and anti-U_1-RNP antibodies are less frequent. Cutaneous and cardiac manifestations are prominent of NLE. Nearly half of the cases show either cutaneous or cardiac features, and 10% show both. Skin lesions may be congenital or develop within 12–16 weeks postpartum. Commonly, the lesions are characterised by erythematous scaly papules or plaques with annular or polycyclic configuration principally affecting the face and scalp, followed by the trunk and extremities. Characteristic periorbital involvement is described as 'raccoon eyes' sign. These lesions usually resolve within a year. Uncommonly, vitiligo-like, morpheaform, erythema multiforme-like, atrophic-telangiectatic, purpuric and discoid-atrophic lesions are seen. Acral papular and bullous lesions have also been described. Cardiac involvement usually develops in the second trimester and is characterised by irreversible complete heart block necessitating pacemaker in almost all the cases. A transient thrombocytopenia occurs in 20% of the cases accounting for the purpuric lesions.[1–4] Here, we report a case of NLE with various types of skin lesions in a newborn of an asymptomatic mother who was incidentally found to have systemic lupus erythematosus (SLE).

A full-term male neonate was brought with lesions on the face and trunk since birth. On examination, multiple erythematous papules with annular, arcuate and polycyclic margins were seen on the abdomen (figure 1A). Also noted on the abdomen was a smooth atrophic hypopigmented plaque with polycyclic erythematous margin, central telangiectasia and areas of brown pigmentation reminiscent of morphoea (figure 1A). Bilateral periorbital erythematous-hypopigmented patches were seen (figure 1B) and the back revealed multiple hyperpigmented atrophic plaques (figure 1C). Rest of the cutaneous and systemic examination was normal. Dermoscopy of the morphea-like lesion showed white globules and structureless areas, telangiectatic vessels, follicular plugging and brown pigmented lines and structureless areas (figure 2), supporting the diagnosis.[5] Examination of the mother revealed asymptomatic erosions on the hard palate without any other cutaneous or systemic abnormalities. Diagnostic workup of the baby revealed anaemia (8.7 g/dL), thrombocytopenia (92 000 cells/mm^3) and normal cardiologic evaluation. Both the mother and baby were positive for Ro/SSA and La/SSB antibodies. Mother's haemogram revealed anaemia (9.6 g/dL), leucopenia (3700 cells/mm^3) and raised erythrocyte sedimentation rate (90 mm/hour). Urinalysis was normal. Based on the findings in the baby and his mother, a diagnosis of NLE was established.

Congenital atrophic lesions in NLE are very rare and are indicative of intrauterine disease with an irreversible outcome.[6] Morphea-like lesions are

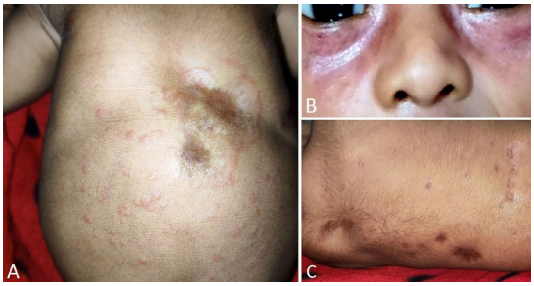

Figure 1 Erythematous papules with arcuate polycyclic margins and an atrophic hypopigmented plaque with polycyclic erythematous margin, central telangiectasia and areas of brown pigmentation on the abdomen (A) bilateral periocular erythematous-hypopigmented patches (B) and multiple hyperpigmented atrophic plaques on the back (C).

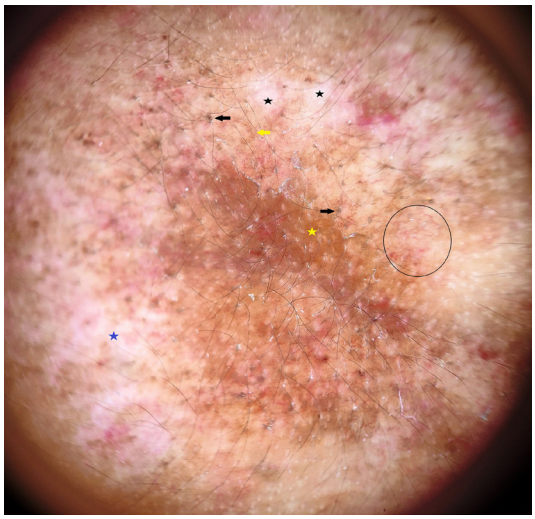

Figure 2 Dermoscopy of morphea-like lesion showing white globules (black stars) and structureless areas (blue star), telangiectatic vessels (black circle), follicular plugging (black arrows) and brown pigmented lines (yellow arrow) and structureless areas (yellow star). (x10, Polarised dermoscopy using DermLite DL3, 3Gen, San Juan Capistrano, California, USA)].

To cite: Kulkarni S, Adya KA, Janagond AB, *et al*. *BMJ Case Rep* 2022;**15**:e252434. doi:10.1136/bcr-2022-252434

also rare.[6 7] Despite the rarity, a diagnosis of NLE was prompted by the characteristic periocular lesions and the accompanying annular polycyclic erythematous papules. Further workup led to the diagnosis of NLE in the baby and incidental detection of SLE in the mother. Hence, NLE should be considered for congenital atrophic or morphea-like lesions with or without the classical lesions in babies of mothers with or without overt manifestations of collagen vascular diseases.

Learning points

- Neonatal lupus erythematosus (NLE) occurs due to transplacental transfer of autoantibodies in newborns of the mothers with clinical or subclinical collagen vascular diseases.
- Typical cutaneous lesions of NLE are erythematous scaly annular or polycyclic lesions predominantly involving the photoexposed areas. Rare manifestations include vitiligo-like, morpheaform, erythema multiforme-like, atrophic-telangiectatic, purpuric and discoid-atrophic lesions.
- Knowledge of the typical and atypical cutaneous lesions of NLE presenting at birth or developing in the early postnatal life is important for considering NLE among the differential diagnoses and to carry out further diagnostic workup in the baby and mother to establish the diagnosis of NLE and to detect any subclinical collagen vascular disease, respectively.

Contributors SK: data collection, data analysis, manuscript preparation, manuscript review. KAA: data analysis, manuscript preparation, manuscript review. ABJ: data analysis and manuscript review. AI: data analysis and manuscript review.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Consent obtained directly from patient(s).

Provenance and peer review Not commissioned; externally peer reviewed.

Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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