Generalized Reticulated Scars and Milia in a Neonate: A Quiz

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A 2-month-old boy presented with erosive and blistering lesions at birth that healed spontaneously within 1 week to form reticulated scars and milia on his face, trunk (Fig. 1A and B) and extremities (Fig. 1C). His oral mucosa, scalp, nails, palms, and soles were spared, and no associated extracutaneous features were found. No new lesions developed during the neonatal period. There was no consanguinity between the parents. The patient was born at term of an uneventful pregnancy by caesarean section. His birth weight was 3,950 g. The placenta, amniotic fluid, and umbilical vessels were normal. The results of routine investigations were normal. Serological tests for syphilis and herpes simplex virus were negative.

What is your diagnosis? See next page for answer.

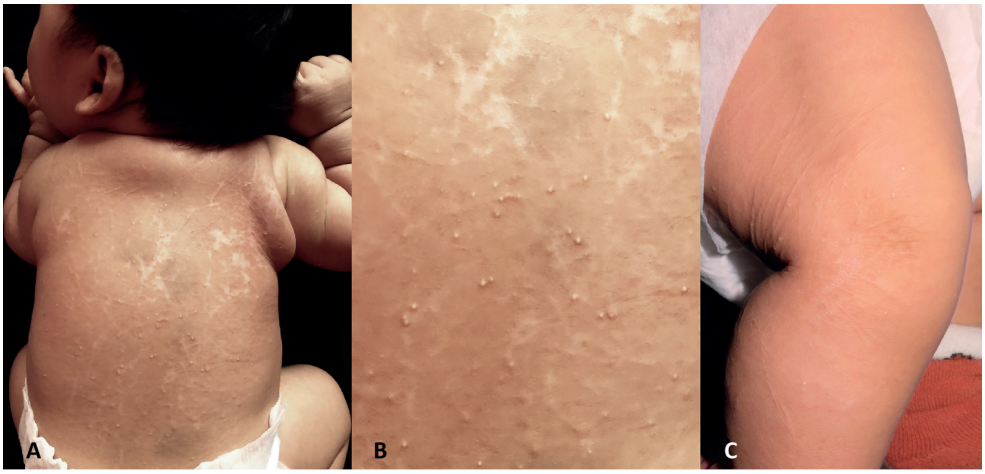

Fig. 1. Clinical photographs. (A, B) Generalized reticulated scars, milia and partial depigmentation on the back. (C) Reticulated scars and milia along the long axis of the leg.

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ANSWERS TO QUIZ

Generalized Reticulated Scars and Milia in a Neonate: A Commentary

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Diagnosis: **Congenital erosive and vesicular dermatosis with milia formation**

Congenital erosive and vesicular dermatosis (CEVD) is a rare disorder of unknown origin presenting at birth with vesicles, erythema, erosions, ulceration, crusts, and fissures affecting up to 75% of the body surface area (1). It usually affects preterm children. Fewer than 40 cases of CEVD have been reported in the literature since 1985 (2). The pathogenesis of CEVD remains unknown, but proposed aetiopathogeneses include birth trauma, premature rupture of membranes, intrauterine infections, chorioamnionitis, amniotic adhesions, purulent or discolored amniotic fluid, and a developmental defect involving unusual healing of premature skin (3). Some cases involving similar findings in siblings may provide insight into the pathogenesis of CEVD, such as whether there is a possibility of genetic predisposition or any recurrent intrauterine pathology occurring during subsequent pregnancy. In future, genetic analysis may contribute to a further understanding of this condition (4).

CEVD is characterized by extensive superficial erosions, vesicles, ulcerations, and crusts with relative sparing of the palms and soles. The lesions heal rapidly within the first few months of life, leaving characteristic reticulated scars. Scarring often follows the lines of cleavage on the trunk and long axis of the extremities. Scalp involvement consists of cicatricial diffuse alopecia in affected areas, which may improve with time. Associated features include neurological defects, ocular abnormalities, hypoplastic or absent nails, scarring alopecia, tongue atrophy, and erosions (5). Whether these associations are related to prematurity or are part of a syndrome remains unclear. Histopathological features are variable and non-specific; they are related to the stage of the disease.

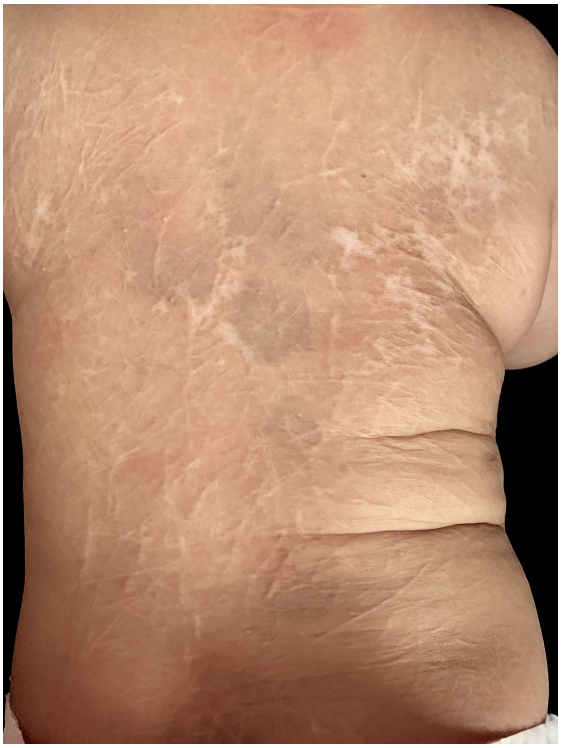

Fig. 2. Most of the milia disappeared spontaneously after 5 months.

Milia may be primary or secondary. Primary milia arise spontaneously and may be present at birth. Secondary milia represent retention cysts that occur in the setting of a proliferative epithelium resulting from trauma (6). The milia in the current patient appeared to be secondary rather than primary because they were limited to the erosive and reticulated scar sites.

The diagnosis of CEVD in the present case was based on the clinical evolution and the typical features of reticulated supple scarring and exclusion of other cutaneous diseases characterized by erosions, blisters, or vesicles at birth. The patient had congenital skin erosion, blisters, and milia formation, requiring us to distinguish the lesions from epidermolysis bullosa. Epidermolysis bullosa was ruled out because the patient had no family history of this condition; developed no new trauma-induced lesions affecting the nails, skin, or mucous membranes since the neonatal period; and exhibited characteristic reticulated scars.

The prognosis of CEVD is generally excellent, with rapid healing, despite the initial appearance, and most patients develop no more lesions after the neonatal period. To the best of our knowledge, the current report describes the first case of CEVD with milia formation. After 5 months, most of the patient's milia had spontaneously disappeared (Fig. 2).

REFERENCES

1. Cohen BA, Esterly NB, Nelson PF. Congenital erosive and vesicular dermatosis healing with reticulated supple scarring. Arch Dermatol 1985; 121: 361–367.
2. Kong BY, Mancini AJ. Congenital erosive and vesicular dermatosis with reticulated supple scarring. Arch Dis Child Fetal Neonatal Ed 2018; 103: F78.
3. Tlougan BE, Paller AS, Schaffer JV, Podjasek JO, Mandell JA, Nguyen XH, et al. Congenital erosive and vesicular dermatosis with reticulated supple scarring: unifying clinical features. J Am Acad Dermatol 2013; 69: 909–915.
4. Srinivas SM, Mukherjee SS, Hiremagalore R. Congenital erosive and vesicular dermatosis healing with reticulated supple scarring: report of four cases. Indian J Dermatol Venereol Leprol 2018; 84: 73–75.
5. Hazarika N, Vathulya M, Joshi PP, Bhatia R. Congenital erosive and vesicular dermatosis in a young girl: a diagnostic dilemma. Int J Dermatol 2019; 58: e212–e214.
6. Langley RG, Walsh NM, Ross JB. Multiple eruptive milia: Report of a case, review of the literature, and a classification. J Am Acad Dermatol 1997; 37: 353–356.