

Congenital erosive and vesicular dermatosis: an atypical presentation of a rare dermatosis

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DESCRIPTION

Congenital erosive and vesicular dermatosis (CEVD) is a rare condition of unknown origin. Classically, it presents at birth with erythema, vesicles, erosions, crusts and fissures, involving over 75% of the skin.¹ The lesions heal spontaneously within 10 days–3 months, leaving behind a pathognomonic reticulate scarring.^{1–3}

The authors present the case of a boy, born prematurely at 25 weeks' gestation by an emergency caesarean delivery to a 31-year-old woman (gravida 1, para 0). The pregnancy had been uneventful, with adequate follow-up. The newborn was admitted to the neonatal intensive care unit for severe respiratory distress syndrome (hyaline membrane disease). Although he did not present with any skin changes at birth, after a few weeks, a small erythematous plaque, with hypopigmented



Figure 2 Scar tissue at 3 years old, note the peculiar shape and cribriform surface.



Figure 1 Small erythematous plaque, first observed in the neonatal intensive care unit.

regions and a cribriform surface, was noted on his left flank (figure 1). There was no apparent traumatic cause and serological tests of syphilis, hepatitis, toxoplasmosis, rubeola, cytomegalovirus and herpes simplex virus (HSV) 1 and HSV2 revealed negative. The infant had favourable evolution and was discharged from the unit after 94 days.

He presented to the dermatology outpatient clinic 10 months after his birth. On his left flank, he had a 3 cm cicatricial-like cribriform plaque, with elevated pink tissue surrounded by a whitish perilesional halo, as well as a smaller identical lesion on the periumbilical region. A punch biopsy of the larger lesion revealed scar tissue and small foci of granulomatous foreign body reaction.

Considering the exclusion of possible differential diagnosis, the course of the dermatosis, the histopathology and the typical scarring, a diagnosis of CEVD was made.

There are less than 40 cases of CEVD reported. Aetiological hypotheses include intrauterine infections, amniotic adhesions, birth trauma and a developmental defect with unusual healing in premature skin.^{1,4,5} It is strongly associated with prematurity, although a few cases have been reported in full-term infants.^{2,3} Other associations include nail abnormalities, hyperthermia/hypohidrosis, maternal chorioamnionitis, alopecia, neurodevelopmental and ophthalmological abnormalities, and tongue atrophy. These patients may also be prone to post-natal herpetic superinfections.¹

Differential diagnosis comprehends amniotic construction bands, congenital infections, epidermolysis bullosa, incontinentia pigmenti, focal



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dermal hypoplasia, aplasia cutis congenita and autoimmune bullous diseases. The exclusion of these differentials and the development of the characteristic supple-reticulated scarring establish the diagnosis.¹

The histopathological findings depend on the stage of the disease. In early inflammatory lesions, there are reports of epidermal necrosis, subepidermal vesiculation and an eroded epidermis with a predominantly neutrophilic or mixed dermal infiltrate, while late lesions show scar tissue.¹

Our patient presented with atypical clinical features, such as the development of the lesions after birth (which, to the authors' knowledge, was reported only in two other cases) and the limited extent of skin involvement.^{2,3} At the current time, he is 3 years old (figure 2) and keeping regular massages of the cicatricial tissue, as to maintain the skin elasticity.

The overall prognosis of CEVD is good, and most patients do not develop new lesions after the neonatal period. Nevertheless, the cosmetic impact of the permanent scarring should be addressed.^{2,4}

Learning points

- Congenital erosive and vesicular dermatosis (CEVD) is a rare dermatosis of the neonatal period that heals with a pathognomonic reticulate pattern.
- CEVD is strongly associated with prematurity.
- CEVD should be considered in the differential diagnosis of non-infectious erythematous plaques and erosions in neonates.

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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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