Segmental, congenital erosive and vesicular dermatosis: mimicking neonatal herpes zoster – a report of a case and review of the literature

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Sri Lanka Journal of Dermatology, 2017, **19**, 36-38

Abstract

Congenital erosive and vesicular dermatosis (CEVD) is an extremely rare disorder of unknown etiology that presents at birth. Initial presentation is with intact vesicles, erythema and extensive crusted erosions, which often involve greater than 75% of the skin surface. Lesions typically heal rapidly, leaving scars with a distinctive supple and reticulated texture. Todate only 28 cases have been reported. Herein we report a case of CEVD involving a segment of skin on left side of the abdominal wall and back. To our knowledge this is the first case of segmental localization of this disease.

Introduction

Congenital, erosive and vesicular dermatosis (CEVD) is an extremely rare disorder of unknown etiology. It often presents at birth, with intact vesicles, erythema and extensive crusted erosions, involving greater than 75% of the total skin surface. Lesions typically heal relatively rapidly, leaving behind scars with a distinctive supple and reticulated texture over the trunk, limbs, scalp and tongue with relative sparing of the face and volar surface of upper limbs. The etiology of the condition remains unclear. The proposed theories include amniogenic theory, vascular theory and non-hereditary intrauterine events. Infants affected with this condition usually are premature and may have abnormal nails and patchy alopecia. Todate only 28 cases have been reported.

Case report

Our patient was a baby girl born to non-consanguineous healthy parents, at 38 weeks'of gestation by a cesarean section. The birth weight was 2.65 kg. At birth, she was noted to have multiple grouped vesicles and crusted erosions in an erythematous background in a segment of skin over left side of the abdominal wall and back (Figure 1). Baby was otherwise healthy.

The attending Paediatrician had made a diagnosis of herpes zoster and initiated treatment with intravenous Acyclovir. The mother denied having vesicular rash during latter part of her pregnancy. She also had a normal antenatal period. Eye examination showed a perimacular chorioretinitis and scarring. The eye lashes, the cornea and tearing were normal. Mucous membranes, hair and nails were not involved. There was no family history of similar skin problems.

Tzanck smear taken from a fresh blister fluid showed only neutrophils. It did not show multinucleated giant cells to suggest a diagnosis of herpes zoster. The biopsy of a vesicle showed a subepidermal blister with neutrophilic infiltration in the dermis. Complete blood count and biochemical analysis were normal. Serologic tests for toxoplasma, rubella, cytomegalovirus, herpes simplex and syphilis (TORCH and VDRL) were negative. The diagnosis of CEVD in our patient was based on characteristic clinical features, course of the disease and exclusion of other diseases which present in similar manner. New lesions continued to appear for a few weeks over the same area of the skin and sometimes over the scars. Few vesicles appeared later across the midline for a few months. The affected areas show marked hypopigmentation and depigmentation. All lesions healed completely within few months with characteristic residual reticulated supple scarring (Figure 2).

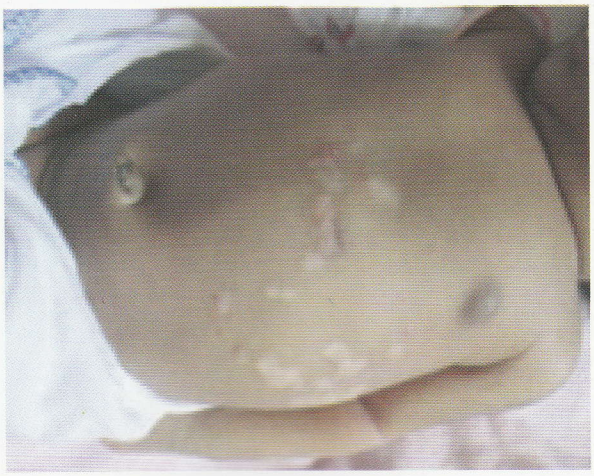

Figure 1.

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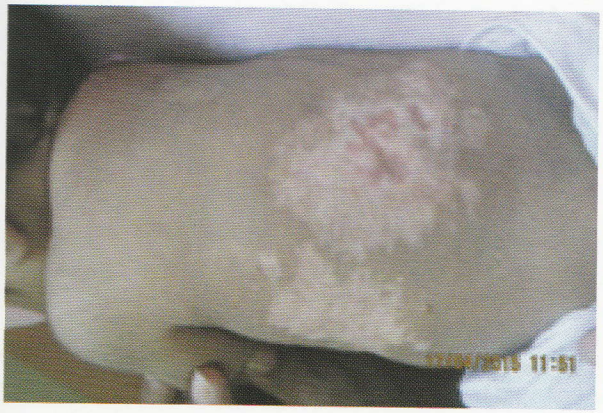

Figure 2.

Discussion

The first case of CEVD was reported in 1985[1]. Since then only few cases (less than 30) have been reported worldwide[12]. The disease is characterized by prematurity, lack of family history and consanguinity and clinical picture characterized by vesicles, erythema, crusted erosions, ulcerations, and fissures at birth. The erosions heal relatively rapidly, leaving behind characteristic supple, reticulated scarring often along the long axis of the extremities and the lines of cleavage on the trunk. There may be discrete round scars with an irregular border. Recurrent episodes of erosions or vesicles were noted in 6 cases, including our patient, most often over the scarred skin[7].

In most of the cases, more than 75% body surface area was affected. The trunk and extremities are most severely involved, with relative sparing of the face, volar surfaces and palms and soles[7]. In seven cases face was involved[8]. Scalp involvement consists of cicatricial localized and diffuse alopecia. Mucous membrane involvement was seen in four cases[11]. The associated ophthalmic involvement is common and the reported eye abnormalities include chorioretinitis, corneal scars, neovascularization, nasolacrimal obstruction, recurrent conjunctivitis, absence of eyelashes. Nail involvement was seen in 10 patients[11]. Reported nail abnormalities are residual total or partial hypoplasia or anonychia. These are independent of skin scarring in the immediate area. Heat intolerance is characteristic, with hypohidrosis of the affected skin and compensatory hyperhydrosis in healthy skin[9]. Dentition is always normal. Rarely it may be associated with neurological defects. The reported abnormalities are seizures, spastic cerebral palsy, structural and developmental abnormalities.

Histology of early inflammatory lesion shows epidermal necrosis, subepidermal bulla and diffuse interstitial neutrophilic dermal infiltrates[4]. There is no evidence of vasculitis or immune complex deposition[6]. Neutrophil-derived proteases may lead to destruction of tissues extensively causing scarring. Later on, histology reveals normal epidermis, few elastic fibers, high collagen, and no evidence of inflammation or neutrophilic infiltrate[1].

The pathogenesis is still not explained yet. But intrauterine infections, intrauterine trauma may be related[3]. Involvement of epidermal structures such as hair, nail, sweat gland and lack of family history may raise the possibility of sporadic onset variety of ectodermal dysplasia[9]. But, to date they have not been able to demonstrate any relevant genetic mutations. The involvement of the retina in some cases raises the possibility of structures of ectodermal origin being targeted. The fact that lesions heal rapidly, raises the possibility of targeted structures being eliminated, like in incontinentia pigmenti.

The prognosis is usually good. Recurrent new blisters limited to early childhood. But few cases have been reported with recurrent episodes beyond early childhood period[7]. Even though it is of benign nature, the scars can cause disfigurement, resulting in lowered self-esteem and possibly adverse psychological implications. Counseling may be necessary, especially in those with extensive or prominent scarring. Better understanding of its etiology may help to guide interventions towards prevention of its onset.

Our patient presented with segmental distribution of vesicles, crusted erosions which has led to an erroneous diagnosis of herpes zoster. Absence of maternal history of chicken pox and negative Tzanck smear excluded this diagnosis. Our patient had perimacular chorioretinitis and scarring which led to possible misdiagnosis of Toxoplasmosis.

As far as we are aware, our patient is the first case with a segmental distribution of this disease.

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