9. Lahmam Bennani Z, El Fekih N, Baccouche D, Khaled A, Zaglaoui F, Fazaa B. [Autoimmune progesterone dermatitis]. Ann Dermatol Venereol. 2012;139:832–5.
10. Rosato E, Farris M, Bastianelli C. Mechanism of action of ulipristal acetate for emergency contraception: A systematic review. Front Pharmacol. 2015;6:315.

S. Oscoz-Jaime,[a,*] M. Larrea-García,[a]
M.J. Mitxelena-Eceiza,[a] N. Abián-Franco[b]

[a] *Servicio de Dermatología, Complejo Hospitalario de Navarra, Pamplona, Navarra, España*

[b] *Servicio de Ginecología, Complejo Hospitalario de Navarra, Pamplona, Navarra, España*

*Corresponding author.
E-mail address: saioaoscoz@gmail.com (S. Oscoz-Jaime).
1578-2190/

Dilated Cardiomyopathy in a Child with Recessive Dystrophic Epidermolysis Bullosa☆

Miocardiopatía dilatada en una niña con epidermólisis ampollar distrófica recesiva

To the Editor:

Dilated cardiomyopathy (DC) is a progressive dilatation and impaired contractility of the left or both ventricles. Predisposing factors may involve familial/genetic, viral infection, autoimmune, nutritional deficit, iron overload, chronic anaemia and drugs.[1]

Sharratt et al. reported in 1986 the first case of DC in a patient with epidermolysis bullosa (EB).[2] Since then, the clinical association of EB and DC has been described in several case reports and case series.[2–6]

A 6-year-old child with severe generalized recessive dystrophic epidermolysis bullosa (RDEB). Treatment with daily cures and moisturizing was made. Her brother, her twin sister and her parents had no relevant medical history. The patient had enteral nutrition by a gastrostomy since 2 years ago. She had chronic anaemia treated with intravenous iron and periodic transfusions were required.

The patient was referred to the Emergency Department due to respiratory distress and influenza-like syndrome for four days.

Her general condition was bad. Her temperature was 36.4 °C, blood pressure 100/50 mmHg, heart rate 125 bpm, and oxygen saturation of 94% with room air. Cutaneous examination showed generalized erosions and syndactalia on her hands with a severe functional limitation (Figures 1, 2).

Electrocardiogram presented diffuse changes in repolarization. Chest radiograph revealed the presence of cardiomegaly and acute pulmonary oedema (Figure 3).

Echocardiogram showed severely dilated left ventricle with ejection fraction of 40%, mild tricuspid insufficiency and moderate pulmonary insufficiency.

The biochemical parameters showed glucose 113 mg/dL, C-reactive protein 120 mg/L, total proteins 9.8 g/dL, haemoglobin 9.2 g/dL, mean corpuscular volume 91.4 fl, 578000 platelets/mm^3, 15300 leucocytes/mm^3 (polymorphonuclear leucocytes 72.6%, lymphocytes 17.4%, monocytes 7.8%, eosinophils 2.1%, basophils 0.1%). Blood cultures were negative.

The patient was admitted to the Intensive Care Unit with a diagnosis of DC, congestive heart failure, acute pulmonary oedema and cardiogenic shock.

Treatment with high flow oxygen therapy, non-invasive mechanical ventilation, hydrochlorothiazide, spironolactone, enalapril, carvedilol and aspirine was initiated. The patient presented clinical improvement after 2 months of treatment. However, she had progressive worsening and was

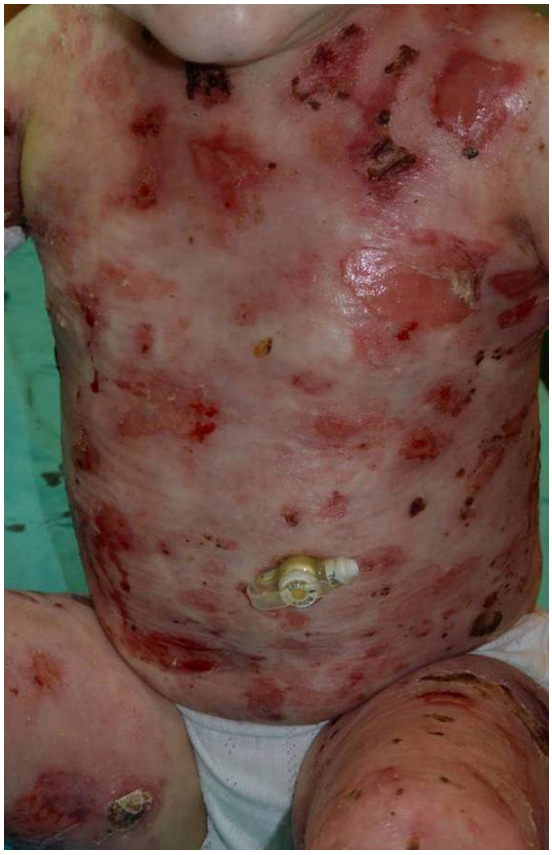

Figure 1 generalized erosions on her trunk and gastrostomy.

☆ Please cite this article as: Imbernón-Moya A, Maseda-Pedrero R, Feito M, de Lucas R. Miocardiopatía dilatada en una niña con epidermólisis ampollar distrófica recesiva. Actas Dermosifiliogr. 2019;110:81–83.

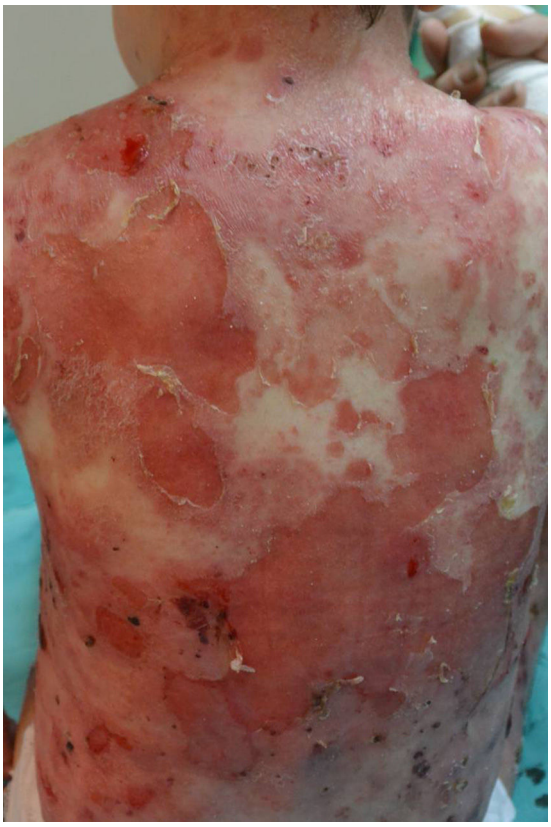

Figure 2 generalized erosions on her trunk.

followed up by the palliative care unit. Sshe died due to multiple organ failure nine months later.

Discussion

Multiple structural and functional alterations in the echocardiogram have been reported in RDEB patients including dilated aortic root, left ventricular hypertrophy, dilatation and dysfunction, increased left ventricular mass and depressed right ventricular systolic function.[3]

DC usually appears in cases of severe generalized RDEB. The most common EB subtypes associated with DC are RDEB and junctional EB-non-Herlitz.[4–6]

The age at diagnosis of DC in RDEB ranges from 2 to 28 years, with a variable ejection fraction from 9 to 45%. Mortality rate varies from 30-60% and death usually occurs within the first 3 months after diagnosis of DC. DC is usually detected at an advanced stage with low ejection fraction.[2–6]

Development of DC in RDEB is due to multifactorial disorders. Contributor factors may include chronic anaemia, iron overload, poor nutritional status, albumin and carnitine low levels, selenium and zinc deficiency, cardiotoxic drugs, viral infections, inflammatory conditions, structural defects in myocardium, chronic renal insufficiency and hypoaminoacedemia.[4–6] To date there is no evidence of mutation in type VII collagen or the basement membrane zone in the pathogenesis of DC.[5]

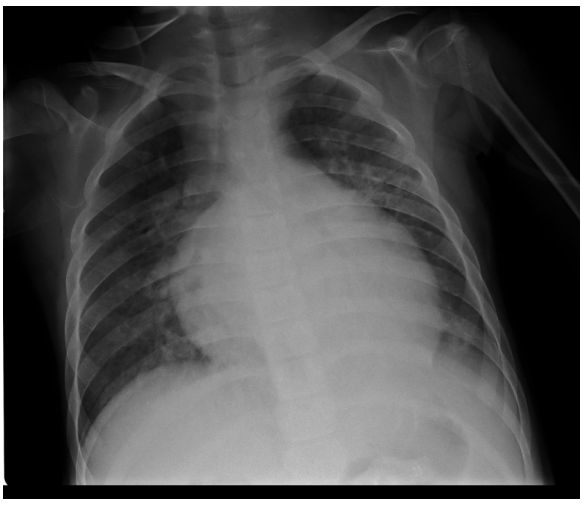

Figure 3 cardiomegaly and findings of acute pulmonary oedema.

History of prior viral infection is usually associated with a more favourable outcome.[4] We suggest that our patient presented DC due mainly to chronic anaemia, iron overload and nutritional deficiency. Viral infection of the upper respiratory tract was the trigger for the development of heart failure and death.

In conclusion, a RDEB patient who develops influenza-like illness and fever requires an urgent cardiac evaluation including electrocardiogram, chest radiograph and echocardiogram. Screening laboratory evaluations are also necessary including albumin, carnitine, selenium, zinc and haemoglobin levels and viruses serology.[5,6] A complete drug history is essential.[4] An early diagnosis and treatment can delay clinical progression and reduce morbidity and mortality.[4–6]

Conflicto de intereses

Los autores declaran no tener ningún conflicto de intereses.

Bibliografía

1. Gagliardi MG. Dilated cardiomyopathy in children. Acta Paediatr Suppl. 2006;95:14–6.
2. Sharratt GP, Lacson AG, Cornel G, Virmani S. Echocardiography of intracardiac filling defects in infants and children. Pediatr Cardiol. 1986;7:189–94.
3. Ryan TD, Lucky AW, King EC, Huang G, Towbin JA, Jefferies JL. Ventricular dysfunction and aortic dilation in patients with recessive dystrophic epidermolysis bullosa. Br J Dermatol. 2016;174:671–3.
4. Lara-Corrales I, Mellerio JE, Martinez AE, Greeen A, Lucky AW, Azizkhan RG, et al. Dilated cardiomyopathy in epidermolysis bullosa: A retrospective, multicenter study. Pediatr Dermatol. 2010;27:238–43.
5. Lara-Corrales I, Pope E. Dilated cardiomyopathy in epidermolysis bullosa. Dermatol Clin. 2010;28:347–51.
6. Batalla A, Vicente A, Bartrons J, Prada F, Fortuny C, González-Enseñat MA. Cardiomyopathy in patients with hereditary bullous epidermolysis. Actas Dermosifiliogr. 2017;108:544–9.

A. Imbernón-Moya,[a,*] R. Maseda-Pedrero,[b] M. Feito,[b] R. de Lucas[b]

[a] *Departamento de Dermatología, Hospital Universitario Severo Ochoa, Leganés, Madrid, Spain*
[b] *Departamento de Dermatología, Hospital Universitario La Paz, Madrid, Spain*

[*] Corresponding author.
E-mail address: adrian_imber88@hotmail.com (A. Imbernón-Moya).
1578-2190/