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Treatment of Harlequin Ichthyosis With Acitretin☆

Ictiosis arlequín tratada con acitretino

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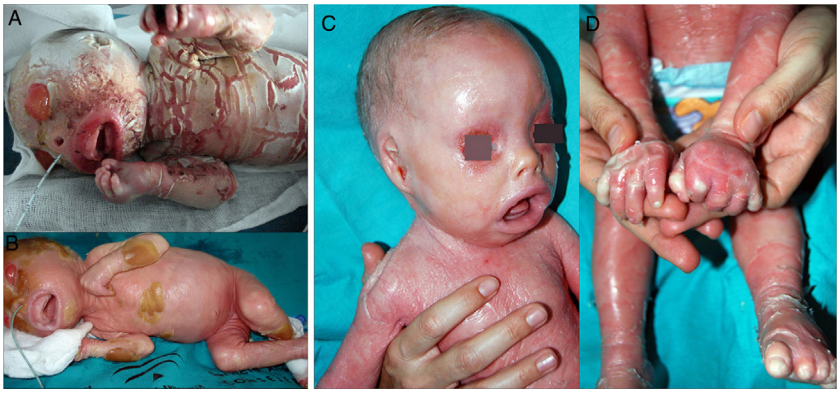

Figure 1

Harlequin ichthyosis is the most severe autosomal recessive congenital ichthyosis caused by homozygotic mutations of the *ABCA12* gene. About 200 cases have been published. We present the case of a premature newborn infant with typical characteristics of harlequin ichthyosis: thick yellowish-brown scales with deep fissures affecting the whole body surface, hypoplasia of all 20 digits, digital contractures, eclabium, ectropion, and madarosis (Fig. 1A). Skin biopsy taken from the trunk revealed a thick and compact orthokeratosis that extended up to dilated pilosebaceous units. Systemic treatment with acitretin was started on the ninth day of life at a dose of 0.5 mg/kg/d, increased to 1 mg/kg/d after 2 months. Tolerance and the clinical response were excellent (Fig. 1B). The infant presented progressive desquamation, with improvement of the ectropion, eclabium, and digital contractures, and was discharged from hospital at 8 months (Fig. 1, C and D). The systemic retinoids are associated with increased survival to 2 to 20 years of age. Acitretin is the drug of choice because of its shorter plasma half-life, which reduces side effects. Prior to its introduction, mortality was around 70% in the first weeks of life. However, it is difficult to carry out clinical efficacy trials because of the very low prevalence of harlequin ichthyosis and because neonatal intensive care (fluid and electrolyte support, topical emollients and keratolytic agents, debridement of constricting bands, monitoring for infection) is also crucial to the prognosis.

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