

Incontinentia pigmenti: a rare genodermatosis

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Infant of 6 months, female, referred by her pediatrician for skin lesions evolving for 2 weeks, with concept of a similar episode since birth evolving by relapses remission according to the mother, the questioning does not find a similar case in the family. The clinical examination objectified vesiculo-pustular lesions with some post-bullous erosions with unilateral crusts of linear localization occurring in affecting the left lower limbs, on the buttocks and on the upper limb (Figs. 1 - 5). The examination of the mucous membranes and integuments was normal and the rest of the examination did not show any dental malformation or neurological damage. The ophthalmologic examination was normal, Biological assessment was within the standards. We retained the diagnosis of Incontinentia pigmenti (Stage I).

Incontinentia pigmenti is a rare genodermatosis with dominant transmission linked to X, it manifests in

newborns, often female by a vesiculo-pustular rash, predominantly acral, with linear arrangement, along the Blascko line, it is a multisystem disease which also affects the teeth, eyes and nervous system, early



Figure 2: Linear vesiculopustular lesions on the buttocks.



Figure 1: Linear vesiculopustular lesions on the left lower limb, with some post-bullous erosions.



Figure 3: Linear vesiculo-crusts lesions on the upper limb.

How to cite this article: Palamino H, Belmourida S, Meziane M, Ismaili N, Benzekri L, Senouci K. Incontinentia pigmenti: a rare genodermatosis. *Our Dermatology Online*. 2021;12(e):e12.

Submission: 24.08.2020; **Acceptance:** 30.12.2020

DOI:10.7241/ourd.2021e.12



Figure 4: Post-bullous erosions with linear hyperpigmentation (after 5 days of local care)

diagnosis is necessary in order to detect and manage complications [1].

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be



Figure 5: Post-bullous erosions with linear hyperpigmentation with vesiculo- crusts (after 5 days of local care)

published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

REFERENCE

1. Minić S, Trpinac D, Obradović M. Incontinentia pigmenti diagnostic criteria update. Clin Genet. 2014;85:536-42.

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Source of Support: Nil, **Conflict of Interest:** None declared.