Abstract

Incontinentia Pigmenti is a rare genodermatosis, which is considered a hereditary alteration, linked to the X chromosome, with a dominant character. It occurs more frequently in women, and the main involvement is observed in tissues derived from the ectoderm, that is, it can be seen as abnormalities in the skin, teeth, hair, eyes and nervous system. We report a case of incontinentia pigmenti with cutaneous manifestations in a male newborn is presented. The case was confirmed through two biopsies evaluated in the pathological anatomy service of the Cayetano Heredia Hospital. Due to the broad spectrum of clinical presentation of incontinence pigmenti, it is recommended to consider this entity in the differential diagnosis when we are faced with predominantly vesicular skin lesions that follow a distribution along Blaschko lines. Incontinentia pigmenti is a rare disease in our country, and its diagnosis requires an adequate clinicopathological correlation, and knowledge of the different phases of the disease. The timely and early diagnosis and recognition of the entity will prevent associated complications at the systemic level.

Keywords: Incontinentia Pigmenti; Heredity; Females; Hyperpigmentation; Peru (source: MeSH NLM).