

Medical Reports & Case Studies

Image Article

Autosomal Dominant Lamellar Ichthyosis in a Cameroonian Family

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Abstract

Ichthyosis encompasses genetic disorders linked to disorders of skin differentiation, but can also be acquired. Neonatal Congenital Ichthyosis which, major risk is predisposition to infections due to inefficient cutaneous barrier, has many sub-type including lamellar Ichthyosis. Although the most described mode of inheritance is the recessive one, few cases of Autosomal Dominant Lamellar Ichthyosis (ADLI) are encountered in literature. Here we describe a similar mode in a Cameroonian family.

Keywords: Lamellar ichthyosis; Autosomal dominant; Cameroon

Image Description

A female newborn aged two weeks was referred from a rural setting for a "congenital dermatologic disorder" noticed since birth. She was born at term from a HIV positive treated woman without any gestational complications. On physical examination, she had palmoplantar hyperkeratosis with large scales covering the whole body including palms and soles, dry skin with desquamation and reduced eyes mobility (Figure 1). The child had neither a collodion membrane nor erythroderma. Otherwise the general state was conserved with normal vital and anthropometric parameters. We noticed similar cutaneous lesions concerning two siblings (all females) and an uncle on the mother's side (Figure 2). Considering the family genealogic tree and characteristics of cutaneous lesions, we thought of a Lamellar Ichthyosis with autosomal dominant mode of inheritance. Financial difficulties of the family didn't allow us to do genetic assessment. The child was treated with regular emollients, keratolytics and close monitoring of electrolytes without any evolutionary complications after 6 months. She was also followed for HIV exposition. Lamellar Ichthyosis is an autosomal recessive disorder in almost all cases. Autosomal dominant transmission is rare [1]. The congenital lesions are not life threatening but largely affects the quality of life of the affected people. Major risks in neonatal period are infections and dehydration which should be prevented with skin care and correct hydro-electrolytic balance [2].



Figure 1: Large scales covering the entire body in a newborn with Lamellar Ichthyosis.

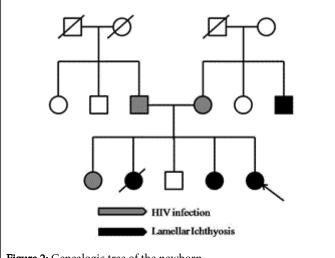


Figure 2: Genealogic tree of the newborn.

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