

Incontinentia pigmenti

Dr. Lelde Reinberga¹, Dr. Karīna Maļina², Dr. Sanita Žigure², Dr. Ieva Ozola²

1. Rīga 1st Hospital; Rīga Stradiņš University, 2. Rīga Stradiņš University; Children's Clinical University Hospital

Incontinentia pigmenti is an X-linked dominant genodermatosis, disorder that affects the skin, teeth, nails, hair, eyes and central nervous system. The classic manifestations are skin lesions that evolve through four stages. Disease is associated with alopecia, hypodontia, nail dystrophy, retinal changes and neurological findings-seizures, developmental delays and intellectual disability. Blistering in first months, followed by wart-like rash, then hyperpigmentation and blaschkoid hypopigmentation.

The aim of this clinical case report is to accent this disease for better understanding and faster recognizing that leads to adequate care and treatment.

11 days old female infant (born at 39 gestational weeks, birth weight 3620g, Apgar score 7/8) was admitted to the Children's Hospital Neonatology unit due to vesicular lesions that were present from birth and progressing. One month before delivery a mother had unspecified stomatitis (other infections she denies) Physical examination: baby's general condition was mild. T 36,8 C. Skin was very dry, hyperemic, with desquamation. On the left leg some small grouped pustules; no oedema. Other systems- without any pathology. Laboratory studies: normal

Skin culture: CONS, normal skin flora contaminant.

A consultation of dermatologist 2 days after hospitalisation: Skin lesions dynamics is negative. Skin was very dry, slightly erythematous, a little bit scaling. Multiple grouped, converging vesicles and pustules were located on the extremities, inguinal region and mons pubis

Recommended skin care program, continuous consultations- ophthalmologist (concl. neonatal conjunctivitis, dacryocystitis), neurologist (NSG without pathology), geneticist consultation.

A differential diagnosis – neonatal infection –, was initially considered because of progressing lesions after birth, mother's infection during pregnancy.

Conclusions: Disease can imitate neonatal infection due to skin lesions. It is important to remember about non-infectious etiology and recognize a genodermatosis of non-infectious cause. If the diagnosis is made precisely, the young patient would be protected from inadequate therapy.