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Images in...

Neonatal incontinentia pigmenti

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DESCRIPTION

A 3-day-old female baby born by normal delivery following an uncomplicated pregnancy presented with blisters on her legs. These blisters were present since birth and were diagnosed as staphylococcal skin infection and treated with oral flucloxacillin and the baby was discharged home. She was referred to our hospital on day 3 as the rash was getting worse. The baby was feeding well and showed no signs of sepsis. The baby was screened for sepsis and changed intravenous antibiotics. The inflammatory markers, blood culture and skin swab were normal.

The rash was characterised by linear vesicles, pustules and bullous lesion with erythema along the lines of Blaschko (figure 1). The rash followed a particular pattern, which is pathognomic for incontinentia pigmenti.

Incontinentia pigmenti is a rare, X-linked, dominantly inherited, disorder of skin pigmentation that often is associated with ocular, dental and central nervous system abnormalities.[1] The incidence is 1 case per 40 000. It usually affects females, as the male fetus does not survive. It results from deletion of the NEMO gene and half of these are new mutations. The skin manifestation is evident soon after birth, as an erythematous eruption with linear vesiculation, followed by a verrucous stage.[2] After a few months the verrucous growth drops off and leaves hyperpigmented areas. The management involves multidisciplinary team involvement with regular follow-up. The babies are also at risk of developing malignancies, which needs to be monitored.

It is important to think about incontinentia pigmenti in babies with typical distribution in absence of signs of sepsis.

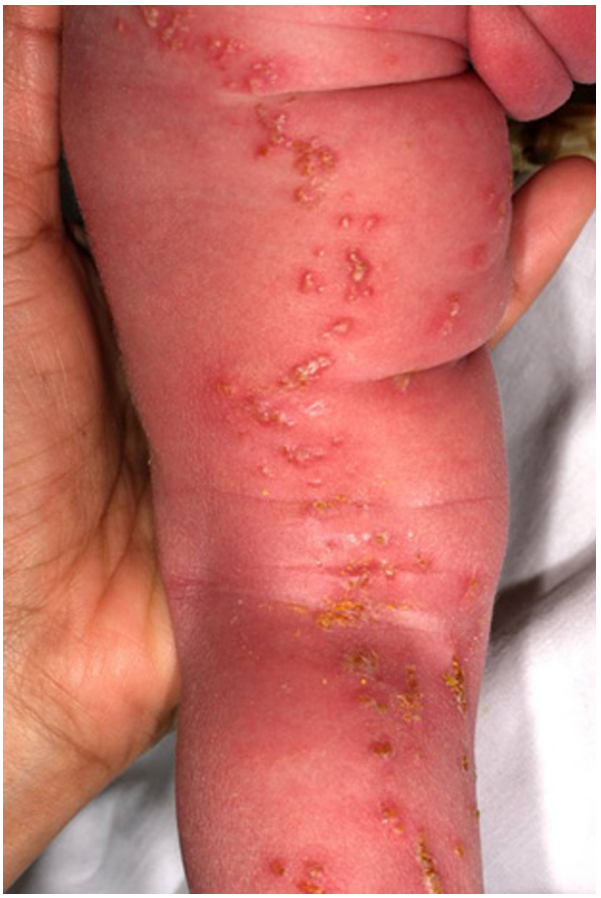

Figure 1 Incontinentia pigmenti along the lines of Blaschko.[3]

Competing interests None.

Patient consent Obtained.

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