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Image Article

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Hypomelanosis of Ito

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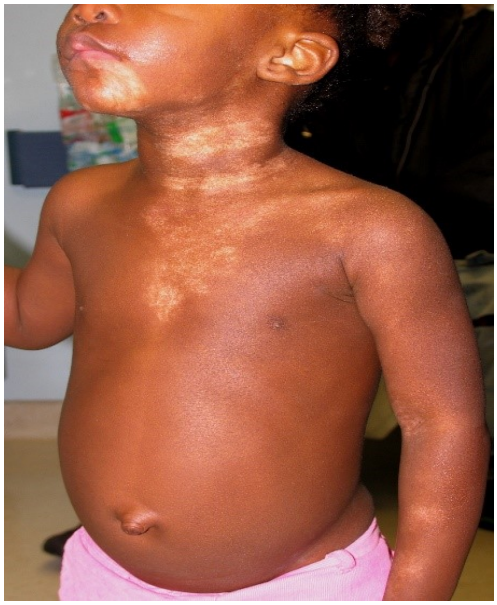
Case Report

A 2 year old previously healthy female presents to your clinic for a well child visit. Review of systems is negative. Physical examination reveals hypo pigmented areas localized to the left side of her face, neck, chest, and arm. Rest of her physical examination is unremarkable.



1. What is the most likely diagnosis?

Hypomelanosis of Ito. It is a rare sporadic neurocutaneous syndrome characterized by unilateral or bilateral hypopigmented whorled or streaked cutaneous lesions in a “marble cake pattern”. Skin lesions tend to run parallel to one another along the lines of Blaschko and are mostly distributed on trunk and extremities. Scalp, palms and soles are rarely affected. Skin changes appear either at birth or during infancy and remain unchanged throughout childhood but may fade or darken in adulthood. Information on age of onset, distribution of lesions and degree of pigment loss can provide vital cues about the underlying etiology in a patient with hypopigmented lesions [1-4].

2. What other associated abnormalities patient can have?

Around 33 to 94% percent of affected individuals may have extracutaneous manifestations mostly involving neurological and musculoskeletal system. These include: neurological (developmental delay, seizures, ataxia, hypotonia, neuropathy, deafness); musculoskeletal (scoliosis, cran-iofacial malformations, hemi-hypertrophy); strabismus, cleft lip/palate, dental anomalies and malformation of cardiac, renal and/or genitourinary tract. Neuroimaging findings can include cerebral or cerebellar atrophy, cerebral dysgenesis, or migrational abnormalities. A thorough history and physical examination is therefore imperative as management is mostly supportive and directed toward the specific symptoms and abnormalities that are apparent in each individual.

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