

Visual Treats in Dermatology

Streaks of hypopigmentation with hemihypertrophy in an infant

Karthick Kannan¹, Anupama Bains¹

¹Department of Dermatology, Venereology and Leprology, AIIMS Jodhpur, Jodhpur, Rajasthan, India.



***Corresponding author:**

Anupama Bains,
Department of Dermatology,
Venereology and Leprology,
AIIMS Jodhpur, Jodhpur,
Rajasthan, India.

whiteangel2387@gmail.com

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A 2-month-old male baby presented with multiple hypopigmented macules arranged in linear streaks, whorls, and bands in blaschkoid distribution predominantly over left side of body since birth [Figure 1]. There was a history of progressive increase in size of the left side of the body. Patient's mother denied history of any systemic complaints. On detailed examination, he also had macrocephaly and hemihypertrophy of the left side of the body. Girth of the left upper (16.5 cm) and lower limb (27.5 cm) was more than right upper (13.5 cm) and lower limb (23 cm). MRI brain showed polymicrogyria and thinning of corpus callosum. Ear, eye, and cardiac examination were normal. The patient fulfilled the diagnostic criteria of hypomelanosis of Ito.

Diagnostic criteria proposed by Ruiz-Maldonado *et al.*^[1]

1. Sine qua non: Non-hereditary cutaneous hypopigmented linear streaks or patches involving more than two body segments, appearing at birth or in the 1st months
2. Major criteria: One or more neurological or musculoskeletal anomalies
3. Minor criteria: Chromosomal anomalies, two or more congenital malformation other than nervous and musculoskeletal systems.

Definitive diagnosis: Criterion 1 (Sine qua non) plus 1 or more criterion 2 (Major) or 2 or more criterion 3 (Minor).



Figure 1: (a) Multiple hypopigmented macules arranged in linear streaks, whorls, and bands in blaschkoid distribution predominantly over left side of body seen. (b) Macrocephaly and hemihypertrophy of the left side of the body seen.

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Presumptive diagnosis: Criterion 1 alone or in association with 1 minor criterion.

Hypomelanosis of Ito is a rare neurocutaneous syndrome with non-hereditary early appearance of hypopigmented macules in blaschkoid distribution. CNS manifestations such as seizures, developmental delay, intellectual disability, hypotonia, and macrocephaly are seen in 90% of cases. Other manifestations include musculoskeletal involvement such as hemihypertrophy, ocular, dental, renal, and endocrine abnormalities. Hence, early diagnosis and referral to specialists become important in cases of hypomelanosis of Ito to prevent complications.^[2]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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2. Chamli A, Litaïem N. Hypomelanosis of Ito. In: *StatPearls*. Treasure Island, FL: StatPearls Publishing; 2022.

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