Hypomelanosis of Ito: An Unusual Association

Corresponding Author:
Dr. B M Shashikumar,
Assistant professor, Dept. of Dermatology, MIMS, Mandya, 571401 - India

Submitting Author:
Dr. B M Shashikumar,
Assistant professor, Department of Dermatology, Mandya Institute of Medical Sciences, Mandya, Dept. od
Dermatology, Mandya Institute of Medical Sciences, Mandya, Karnataka, 571401 - India

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Author(s): Shashikumar BM, Reddy RR, Harish MR

Abstract

Hypomelanosis of Ito is a neurocutaneous syndrome characterized by hypopigmented lesions occurring in streaks and whorls located on the trunk, head or extremities. Associated abnormalities occur in musculo-skeletal system, CNS, eyes, teeth etc. we report a case of hypomelanosis of Ito with rare association of alternate convergent squint and microcephaly.

Introduction

Hypomelanosis of Ito, also known as Incontinentia Pigmenti Achromians (IPA), is a rare neurocutaneous syndrome clinically characterized by macular hypopigmented whorls, patches and streaks resembling fountain spray splatters [1]. Associated abnormalities are thought to occur in 30-50% of patients with cutaneous lesions and includes defects in the musculo-skeletal system, CNS, eyes, teeth etc. We report a case of hypomelanosis of Ito with rare systemic associations.

Case reports

A 9-year-old girl presented with hypopigmented lesions over trunk, both upper limbs and both lower limbs of 6years duration. It was insidious in onset. Initially it was noted as small hypopigmented lesion over back which gradually increased in a linear fashion to involve all parts of the body. No history of itching. All her developmental milestones were normal. There was no history of seizures, vesiculobullous skin lesions or weakness of limbs. The girl was a full term normal delivery, the first child of a non-consanguineous marriage and with no history of birth trauma. The second sibling, 6-year-old boy, was normal.

General physical examination revealed normal milestones except for borderline mental retardation and microcephaly with head circumference of 46.5cms (expected 50cms). Dermatological examination revealed bizarre hypopigmented linear streaks bilaterally over the trunk giving a fountain spray splatters [Illustration 1] and similar linear lesions along the limbs following Blaschko's lines [Illustration 2]. Hair, nails, palps, soles and mucosae were normal. Ophthalmological examination revealed alternate convergent squint of 30°. Slit lamp and fundoscopic examination were normal. Other systems were normal. Radiological examination of the spine, CT scan of head, echocardiography and ultrasonography of abdomen was normal.

Discussion

Hypomelanosis of Ito is a neurocutaneous disorder characterized by a bizarre, bilateral, irregularly shaped leukoderma affecting the trunk and extremities and often associated with neurologic and musculoskeletal abnormalities. It is diagnosed in 1 per 8000-10000 unselected patients in general paediatric outpatient clinic and 1 out of every 790 in a paediatric dermatology clinic [2].

Though originally described as a purely cutaneous disease subsequent reports have included 33% to 94% association with multiple extracutaneous manifestations mostly of the central nervous and musculoskeletal systems leading to frequent characterization as a neurocutaneous disorder [3]. The hypomelanotic macules of Hypomelanosis of Ito are usually present at birth but may appear in early infancy or childhood. Eye abnormalities include microphthalmia, iris coloboma, heterochromia irides, pinpoint pupils and retinal pigmentary abnormalities [4]. Alternate convergent squint and microcephaly, the two unusual association were noted in this case. This case is present for rare presentation of the condition with unusual associations.

References

Illustrations

Illustration 1

Illustration 1: Shows bilateral hypopigmented streaks giving fountain spray appearance

Illustration 2

Illustration 2: Shows hypopigmented linear macules along Blaschko’s lines over lower limb
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