Hypomelanosis of Ito: An Unusual Association

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No conflict of interest
Hypomelanosis of Ito: An Unusual Association

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 6 years 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 46.5 cms (expected 50 cms). 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, palms, 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
Reviews

Review 1

Review Title: Hypomelanosis of Ito: An Unusual Association

Posted by Faculty Dr. Nooshin Bagherani on 22 Jul 2012 10:45:14 AM GMT

What are the main claims of the paper and how important are they?:
This manuscript has presented a rare neurocutaneous syndrome in association with unusual manifestations.

This article is invaluable. Although the association between hypomelanosis of Ito and alternate convergent squint and microcephaly is not a novel presentation, it is rare. Searching computerized PubMed database showed no "case report" article which directly presents this association altogether.

No

Yes

If a protocol is provided, for example for a randomized controlled trial, are there any important deviations from it? If so, have the authors explained adequately why the deviations occurred?
Nill

Yes

The authors could note the probable genetic associations.

The authors could note the probable genetic associations.

Rating: 7

Comment:
This article is invaluable.

Competing interests: None

Invited by the author to make a review on this article? : No

Have you previously published on this or a similar topic?: No

References:

Experience and credentials in the specific area of science:
Nill

How to cite: Bagherani N.Hypomelanosis of Ito: An Unusual Association [Review of the article 'Hypomelanosis of Ito: An Unusual Association ' by ].WebmedCentral 1970;3(7):WMCRW002108
Review 2

Review Title: REVIEW OF THE ARTICLE: Hypomelanosis of Ito

Posted by Dr. Constantino Ledesma-Montes on 19 Jun 2012 08:22:17 PM GMT

What are the main claims of the paper and how important are they?:

The main claim is to present the clinical features of a patient with Incontinentia Pigmenti.

It is an important matter for Dermatologists.

This case is an unusual association among Incontinentia Pigmenti Achromians and other developmental abnormalities.

Yes, there are few cases of this developmental disease with the anomalies present with the reported case.

If a protocol is provided, for example for a randomized controlled trial, are there any important deviations from it? If so, have the authors explained adequately why the deviations occurred?

NA

NA

NA

NA

Rating: 7

Comment:

It is an interesting and well done case report of a rare disease and unusual clinical associations.

Competing interests: None

Invited by the author to make a review on this article?: No

Have you previously published on this or a similar topic?: No

Experience and credentials in the specific area of science:

I am an oral and Maxillofacial Pathologist and few cases of this developmental disease assisted to my clinic.

How to cite: Ledesma-Montes C.REVIEW OF THE ARTICLE: Hypomelanosis of Ito[Review of the article 'Hypomelanosis of Ito: An Unusual Association” by ].WebmedCentral 1970;3(6):WMCRW001946
Review 3

Review Title: Hyomelanosis of Ito: An Unusual Presentation

Posted by Faculty Dr. Varadaraj V Pai on 18 Jun 2012 01:37:00 AM GMT

What are the main claims of the paper and how important are they?:
Describing an unique case of ITO and pretty successfully done

No

Very correctly placed

Yes

If a protocol is provided, for example for a randomized controlled trial, are there any important deviations from it? If so, have the authors explained adequately why the deviations occurred?
Case report protocol is perfect

Yes

Histopathology would help

Histopathology would help

Rating: 7

Comment: No

Competing interests: No

Invited by the author to make a review on this article? No

Have you previously published on this or a similar topic?: No

Experience and credentials in the specific area of science:
Am a practising dermatologist and have few such cases

How to cite: Pai V. Hyomelanosis of Ito: An Unusual Presentation [Review of the article 'Hypomelanosis of Ito: An Unusual Association ' by ]. WebmedCentral 1970;3(6):WMCRW001936
Review 4

Review Title: Reviewd

Posted by Dr. Pradeep V Bhagwat on 01 May 2012 05:27:32 AM GMT

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Rating: 7

Comment:
It is an excellent case report. Documented well.

Competing interests: Nil

Invited by the author to make a review on this article? : Yes

Have you previously published on this or a similar topic?: No

Experience and credentials in the specific area of science:
I have seen few cases earlier

How to cite: Bhagwat P. Reviewd[Review of the article 'Hypomelanosis of Ito: An Unusual Association ' by ].WebmedCentral 1970;3(5):WMCRW001758
Review 5

Review Title: Hypomelanosis of Ito: an unusual association

Posted by Lead Faculty Prof. Valcinir Bedin on 24 Apr 2012 09:59:58 PM GMT

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Rating: 8

Comment: As a case report it is very well documented and very explainable.

Competing interests: no

Invited by the author to make a review on this article? : No

Have you previously published on this or a similar topic?: No

Experience and credentials in the specific area of science: I am a dermatogy professor.

How to cite: Bedin V. Hypomelanosis of Ito: an unusual association[Review of the article 'Hypomelanosis of Ito: An Unusual Association' by ].WebmedCentral 1970;3(4):WMCRW001732
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