

Darier's disease with guttate leukoderma: correlation of dermoscopic and histological findings

POSTER
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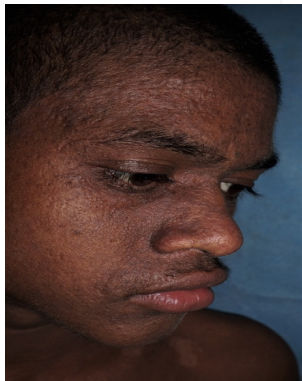


Background

- Darier's disease is an autosomal dominant disorder of keratinization caused by mutations in ATP2A2 gene.
- Characterized by greasy, hyperkeratotic papules predominantly in a seborrheic distribution.
- Guttate leukoderma has been described as a rare manifestation of Darier's disease .

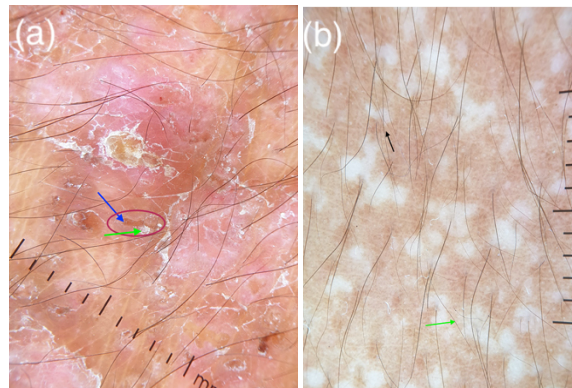
Clinical history

- Fifteen-year-old boy with multiple itchy hyperpigmented papules over face and trunk with hypopigmented macules over limbs for last 8 years.
- Exacerbation of itching over lesions in summers, especially with sweating.
- Hypopigmented macules were not preceded by any inflammatory lesions.
- Similar lesions were also noted in his mother.



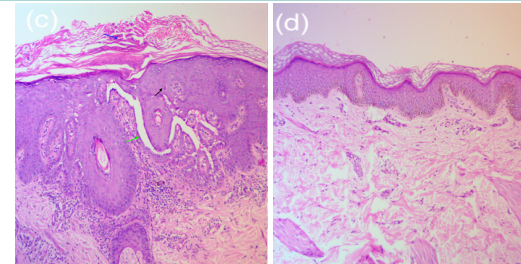
Clinical examination

- Multiple skin colored to hyperpigmented greasy papules and plaques over the face, predominantly at the centro-facial area and the upper trunk.
- Multiple hypopigmented macules, 1-4 mm in size, over bilateral upper limbs.
- Punctate depressions were seen on palms while nails and oral cavity were within normal limits.



Dermoscopic examination

- A. Facial papules - central brownish area (blue arrow) with a thin whitish halo (oval shape) and fine white scales (green arrow).
- B. Hypopigmented lesions- multiple depigmented areas (black arrow) of variable size and shapes, confluent at places (green arrow).



Skin biopsy

- C. Papule over chest - hyperkeratosis, parakeratosis, acanthosis, suprabasal cleft and dyskeratotic cells in form of corps ronds and grains.
- D. Hypopigmented macule - within normal limits.

Conclusion

- Brownish areas and fine white scales histopathologically correspond to marked hyperkeratosis and parakeratosis while a peripheral whitish halo correlates with marked acanthosis.
- Dermoscopic evaluation may help in early recognition of these lesions in a patient with Darier's disease which may allow earlier diagnosis and appropriate counselling to patients and family member.

References

1. Takagi A, kamijo M, ikeda S. Darier disease. J dermatol 2016;43:275-9.
2. Harb J, motaparathi K. Clinicopathologic findings of guttate leukoderma in darier disease: A helpful diagnostic feature. JAAD case reports 2018;4:262-266.