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.

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.

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).

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