DARIER'S DISEASE
(Case reports)

K. ANANDAM * AND C. T. HEMACHANDRA †

Summary

Three cases of Dariers Disease are reported. Histopathological aspects are presented. Other relevant aspects are discussed.

Introduction

Darier's disease is a rare familial disorder, inherited as an autosomal irregular dominant type, affecting both sexes in childhood or early adult life. It is characterised by the appearance of hyperkeratolic and crusted papules, that by confluence form crusted areas. When fully established, the disease affects temples, paranasal furrows, scalp, ears, sternal and interscapular regions, genital region and large skin folds. This condition was first described by Darier in 1883. Since then many cases have been described in world literature. Recently we had an opportunity to see three cases which are reported here.

Case Reports

Case No. 1

A Hindu male aged 20 years, presented with multiple scaly papular lesions in the seborrhoeic areas of 15 days' duration. There was no history of consanguinity among parents, and he was the only issue. He is married and he has one son who is free of skin disease. No history of drug intake was present.

Blood counts, urine examination and stool examination were normal. S.T.S. was negative.

Histopathology

Epidermis showed hyperkeratosis and some nucleated parakeratotic cells (grains). Granular and malpighian layers showed multiple splits and lacunae. Interspersed among the normal cells there were corps ronds possessing large round, homogenous deeply basophilic nuclei and homogenous deeply eosinophilic cytoplasm. There was irregular upward proliferation of papillae into lacunae resulting in formation of villi. (Figs. 1 & 2 Page No. 293)

Case No. 2 and 3

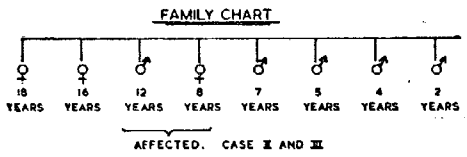
A 12 year old male child and his 8 year old sister presented with multiple papular scaly lesions on scalp, face, presternal areas, infrascapular regions and flexors of 2 days' duration. There was no history of drug intake. Four brothers and two sisters were living

* Civil Surgeon and Head of the Dept. of Venereology, Kurnool Medical College, Kurnool (formerly Asst. Prof. of Dermatology, Osmania Medical College, Hyderabad).

† Civil Surgeon and Head of the Dept. of Dermatology, Osmania Medical College, Hyderabad. (Presently retired).

Received for Publication on 23—12—1976

and healthy. Parents were nonconsanguinous. Mother had no abortions (family history appended below). Blood counts, urine-analysis and stool examination did not show any abnormality. S.T.S. was negative on all the children and mother.



Biopsy of skin lesions revealed typical histopathological features of Darier's disease.

Discussion

The study of these cases show that Darier's disease is a familial one. The definite mode of inheritance could not be determined with certainty in our cases. Onset was in early adulthood in one case while in other two it was during childhood. Two patients were males and one female. In none of the cases was any systemic abnormality detected. These cases are presented because of paucity of reports on such.

FALSE

Several mechanisms contribute towards the beneficial effect exhibited by beta carotene in photosensitive porphyrias. This agent strongly absorbs 400 nm light thus preventing the penetration of their rays and subsequent symptms. This effect plus the ability to efficiently trap singlet excited oxygen radicals and quenching free radicals of beta carotene makes this a therapeutic agent superior to Alpha tocopherol.

Reference : Moshell AN and Bjornson L : Photoprotection in erythropoietic protoporphyria : mechanism of photoprotection by beta carotene, J Invest Dermatol, 68 : 157, 1977.