

PAGETOID RETICULOSIS

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A case of pagetoid reticulosis (Woringer Kolopp disease) is reported. The diagnosis was confirmed by the typical histopathology. Review of the available literature failed to show report of any such case from India and hence this report.

Introduction

Pagetoid reticulosis is a rare form of cutaneous T cell lymphoma which exhibits a chronic localised nature and histologically characterised by a typical pagetoid mononuclear cells colonising the epidermis (epidermotropism).

Case Report

A 56 years old man was referred for a pruritic, slowly spreading verrucous plaque of the left foot of 8 years duration. Recurrent ulceration occurred over the lesion which responded temporarily to antibiotics. Antituberculous treatment was given for 7 months from elsewhere without any response.

On examination an irregular erythematous scaly plaque of 20 x 12 cm. in size was seen on the left foot involving the lateral surface of the dorsum, lateral three toes, and anterior two thirds of the sole of the foot. Margins of the lesion were circinate, raised and verrucous while the central part was relatively clear with tendency for healing. Areas of ulceration and crusting were also present. Depigmentation with spotty hyperpigmentation was noticed over the forefoot and involved the toes. The little toe was deformed with destruction of the nail. Regional lymph nodes were enlarged. There was no generalised lymphadenopathy or hepatosplenomegaly.

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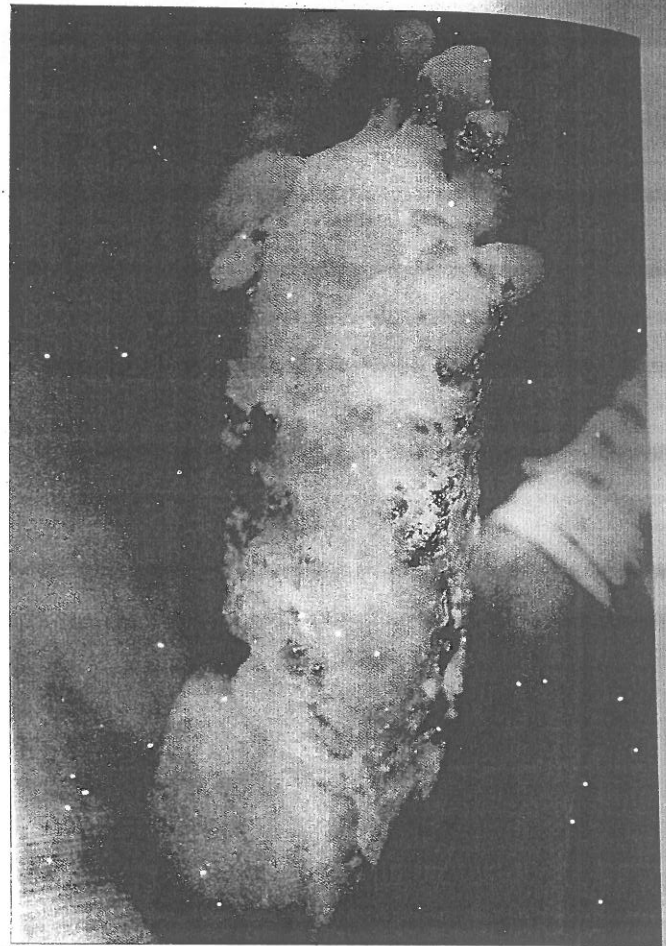


Fig. 1.

Routine blood and urine examinations were within normal limits. Peripheral smear did not show any abnormal cells. Mantoux test was negative. Chest X-ray was within normal limits and blood VDRL was non reactive. Regional lymph node biopsy showed only reactive hyperplasia. An incisional biopsy of the lesion was done.

Epidermis showed marked hyperkeratosis, irregular acanthosis and was infiltrated upto the stratum corneum by numerous atypical mononuclear cells. The cells

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this disease a variant of mycosis fungoides. It is different from Mycosis Fungoides in its clinical behaviour and histopathological picture. Electron microscopy has shown the atypical mononuclear cells to be indistinguishable from mycosis cells.

Tan et al consider pagetoid reticulosis, epidermotropic mycosis fungoides and mycosis fungoides to be part of the spectrum of mycosis fungoides, the first representing the early or benign end of the spectrum.² Immunohistochemical and enzymohistochemical studies have shown these cells to be of T lymphocytic and also histiocytic origin. For the time being it is best to regard Worringer-Kolopp disease as a form of cutaneous T cell lymphoma that may remain localised for many years.

Acknowledgement

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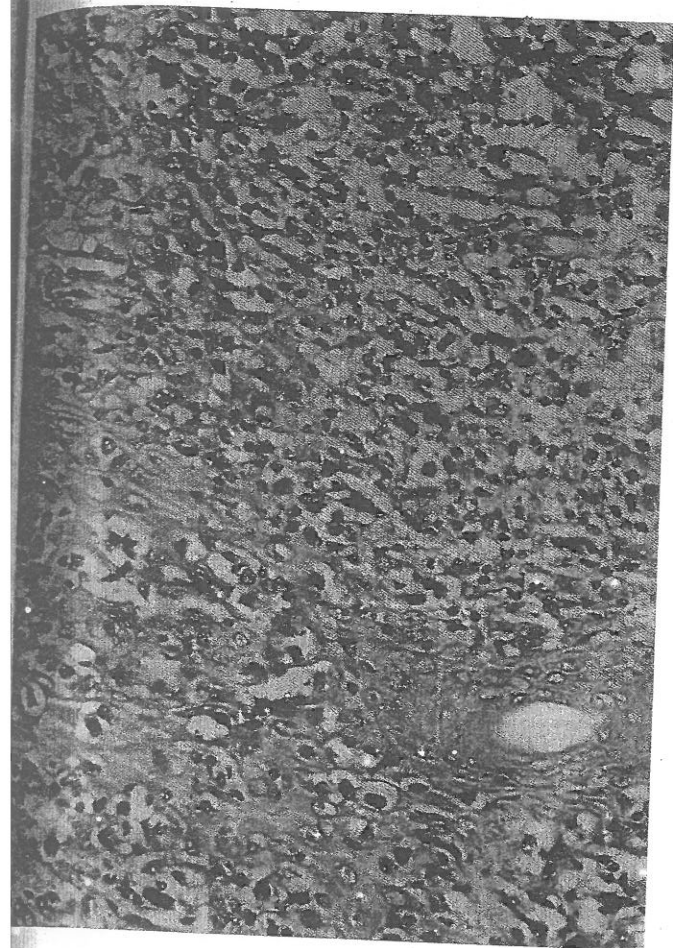


Fig 2

had large, hyperchromatic, atypical, irregular nuclei and abundant vacuolated cytoplasm forming a halo around the nucleus. Nuclei were arranged in nests which resembled a follicular abscess. Upper dermis contained collections of the same cells along with lymphocytes and plasma cells. This histological picture is consistent with Pagetoid Reticulosis.

The patient was referred to Regional Cancer Centre, Trivandrum for treatment where he received local irradiation with 4000 rads in 20 fractions and is showing good response on follow up.

Comment

This rare disease was first described by Worringer and Kolopp in 1939. Many consider