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CASE REPORT

A CASE OF CUTANEOUS T-CELL LYMPHOMA

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ABSTRACT

The peripheral T-cell lymphomas are a heterogeneous group of neoplasms that constitute less than 15% of all non-Hodgkin lymphomas in adults but the most common subtype of cutaneous lymphomas. It is characterized by the presence of erythematous plaques that evolve into ulcerated lesions, tumours throughout the skin or even bone marrow infiltration in advanced stages. We report a case of a 36 year old female who clinically presented with multiple, generalized, papulo- nodular skin lesions.

INTRODUCTION

The incidence of Mycosis fungoides (MF) is approximately six cases per million per year, accounting for about 4% of all cases of non-Hodgkin lymphoma[1]. It is distinguished from other cutaneous T cell lymphomas (CTCL) by its unique clinical and histologic features. The most common clinical feature is skin changes that are often pruritic and is characterized by heterogeneous cutaneous manifestations including patches, plaques, tumours, generalized erythroderma[2], poikiloderma, or rarely, papules, nodules[3]. Initially, it affects the skin in areas not exposed to the sun[4] and with the development of the disease lesions become infiltrated, as elevated erythematous plaques or reddish-brown, with well defined borders and eccentric contours. The gold standard for the diagnosis of MF is histopathology that can be complemented by clinical, molecular and immunopathologic presentation.

Case Report

A 36 year old female presented in our institute with multiple itchy lesions all over the body for 2 ½ yrs. Initially she developed small whitish lesion over lower limb, which was non-itchy and non-anesthetic, progressive and was diagnosed clinically as leprosy despite the slit skin smear was negative for acid fast bacilli. She was then given treatment for lepromatous leprosy for 1 year to which she did not respond. The skin lesions gradually spread all over the body and became lumpy in appearance. She had no addiction and her past medical-surgical history was unremarkable.

On examination numerous erythematous, soft papulo-nodular and plaque lesions of different sizes (1 cm to 3 cm in diameter) with erythroderma were present all over the body (fig 1, 2). It was more in face, neck, upper limbs, trunk and genitalia and less in number in lower limbs. No lesion was present on palm, sole and in oral cavity. Surface over the lesions was moist at axilla and genitalia with ill defined border and lesions were conglomerated, verrucous type without sensory impairment or peripheral thickening of nerves. There was no lymphadenopathy or organomegaly. Other system examination revealed no abnormality.

Complete blood count was within normal limit with normal ESR and without any abnormal cell in the peripheral smear. Her chest x-ray and USG whole abdomen were normal. Skin biopsy was done to find out the diagnosis and the histopathology showed thinning of epidermis with downward elongation of thinned rete ridges in some area. Dense lymphocytic infiltrate in the dermis with different size and different nuclear contour. Invassion of lymphocytes from dermis to epidermis (epidermatophism) without spongiosis (fig 3,4). Pautrier's micro-abscess was present in areas of section.

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Immunohistochemistry revealed, tumour cell expressed – CD 3, CD 5, CD 2 and CD 4 and immuno-negative for – CD 20, CD 30, CD 8 and TdT. Then the patient was diagnosed as cutaneous T cell lymphoma and referred to medical oncology department for treatment.

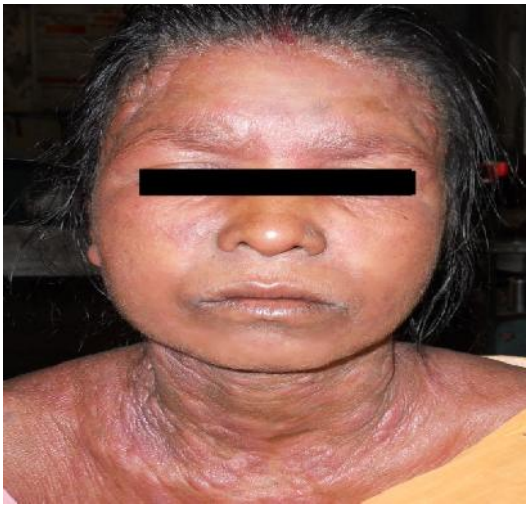

Figure 1 Papulo-nodular and plaque lesions of different sizes with erythroderma.

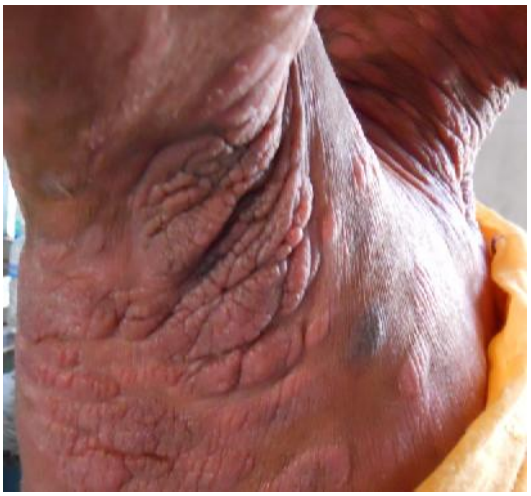

Figure 2 Conglomerated, verrucous type lesions at axilla.

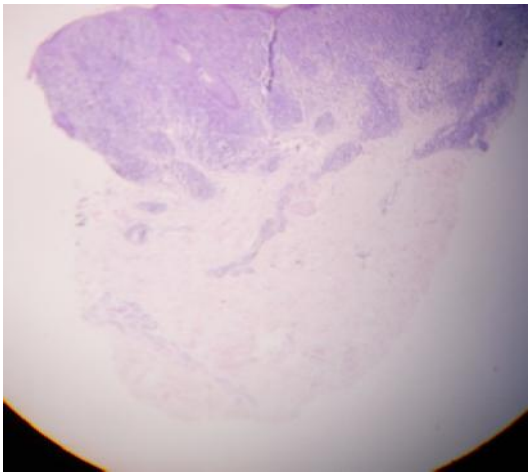

Figure 3 Dense lymphocytic infiltrate in the dermis with different size and different nuclear contour. Invassion of lymphocytes from dermis to epidermis(epidermatophism) without spongiosis.

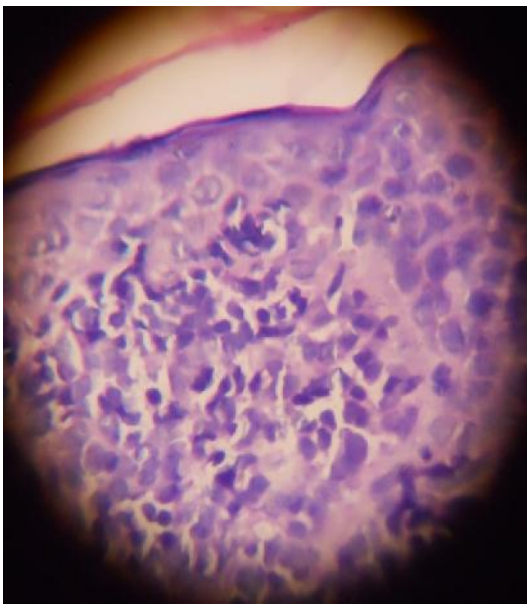

Figure 4 Atypical lymphocytic infiltrate in the dermis**.**

DISCUSSION

Mycosis fungoides is the most common type of cutaneous lymphoma of T- cells included in the classification of non-Hodgkin lymphoma. Histologically, MF is characterized by the presence of Sézary-Lutzner cells (T helper cells), which form clusters in the superficial dermis and invade the epidermis in small cell groups[5]. It is more common in the age group of 55-60 years of age, being more common in male patients and with rare incidence in childhood and young adults[6]. Skin biopsy with routine histology is the single most important laboratory tool that will assist the clinician in establishing the diagnosis. Skin biopsies demonstrate small to medium-sized atypical mononuclear cells with cerebriform nuclei infiltrating the upper dermis among epidermal keratinocytes (epidermotropism) or forming intraepidermal aggregates (Pautrier microabscesses). Pautrier microabscesses are pathognomonic, yet uncommon[7] and spongiosis, or the collection of fluid in the epidermis, is not seen. Immunophenotyping is used to support or confirm results of the routine histology[8]. For staging it is taken into account the evolution of the disease in the skin (T), the lymph node status (N), visceral (M) and blood involvements (B). In early stage it is treated with topical chemotherapy (Nitrogen mustard or carmustine), topical corticosteroids, phototherapy or radiotherapy (x-ray or electrons) localized or on the whole body surface. In later stages, it can be tried total skin irradiation as a form of curative or palliative treatment[9] and combined chemotherapy is generally used in case of unequivocal lymph node or systemic involvement, or in cases with widespread tumor-stage[10]. Prognosis of MF depends on the staging, mainly concerning the extension and skin involvement type and the presence or absence of extra cutaneous disease.

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