To the Editor:

Mycosis fungoides presenting as exfoliative dermatitis

Sri Lanka Journal of Dermatology, 2003, 7, 43-44

Exfoliative dermatitis (ED) frequently seen in clinical practice, is often due to common problems such as psoriasis, eczema and drug reactions. ED due to mycosis fungoides (MF) is rare.

A 52 year old male presented to the dermatology out patients' clinic in year 2001 with ED. Skin biopsy was done, but it was non specific. This patient was treated for ED with topical emollients and topical steroids, and was followed up in the clinic. He was lost to follow up for sometime.

Two years later he was admitted to the dermatology ward with exacerbation of ED and he was found to have pripheral lymphadenopathy with hepatomegaly. Skin biopsy was repeated again, it showed atypical mononuclear cells infiltrating the epidermis and dermis. Suggesting mycosis/ fungoides. He had mild leukocytosis with 40% lymphocytes. Blood picture showed occasional sezary cells (2 in number). Lymph node biopsy showed dermatopathic lymphadenopathy with atypical lymphocytes. Bone marrow biopsy was normal. Ultra sound scanning of abdomen showed liver enlargement; 3 cm below the costal margin, with normal echogenicity. Ultimately, a diagnosis of mycosisfungoides - stage IV, was made.

MF is a cutaneous T cell lymphoma, which is a slowly progressing chronic condition. It is commoner among elderly people with a male to female ratio of 2:1[2]. Aetiology is not known.

MF shows various clinical presentations like macular/patchy lesions, plaque lesions, nodular lesions, erythrodermic and poikilodermatous patterns[1]. Erythematous macular patchy lesions mainly occur appear on the trunk than limbs but in dark skinned people hypopigmented patches may occur. Plaque lesions may occur in two forms such as limited plaque lesions (involving less than 10% of body surface area) and extensive plaque lesions (involving more than 10% body surface area)[1]. Nodular lesions may develop on these plaques, and sometimes, these nodules may ulcerate. Erythrodermic and poikilo-dermatous patterns are less common[1]. MF may present with systemic involvement which shows a poor prognosis[2]. Pruritus and burning sensation are common symptoms.

Histology of MF shows epidermotropism and Pautrier's abscess. Spongiosis is uncommon. Dermis shows monomorphic cellular infiltrate with atypical lymphocytes[1].

Atypical lymphocytes of MF express various chemokine receptors, particularly CCR4, CXCR3, CXCR4, in the early stages of the disease. These receptors may contribute to chemotaxis and the pathogenesis of the disease[5].

MF can be staged according to TNM classification from stage I to stage IV, and every stage is subdivided into A and B except stage III[1].

Topical therapy such as potent steroids, nitrogen mustard, BCNU (carmustine) are effective in early stages of MF with better response rate[1]. Narrowband UVB phototherapy is successful in early stage of MF (stage 1A and IB) like PUVA therapy[7]. Erytherodermic patients do not tolerate PUVA well. Total skin electron beam treatment is also effective in MF[1]. Photodynamic therapy and photopheresis are also used in MF[1] systemic chemotherapy is limited to the advanced stages (stage III and IV). Low dose methotrexate, is effective in erytherodermic MF[1]. Cyclophosphamids, daunorubicin, vinscritine may also be used. A recent study shows, Pentostatin IV treatment is effective in advanced stages of MF[3]. Alemtuzumab (campath -1H) can be used as salvage treatment in advanced stages of MF/sezary syndrome[4].

This case highlights the need to do serial biopsies in cases of exfoliative dermatitis if the aetiology of ED is not clear. It also highlights that exfoliative dermatitis may be a manifestation of mycosis fungoides without sezary syndrome.

References

1. Mackie RM. Cutaneous lymphoma and lympositic infiltrates. In: Champion RH, Burton JL, Burns DA, Breathnach SM eds. Textbook of Dermatology 6th ed Oxford, Blackwell science 1998; 2375-2399.
2. Odom RB, James WD, Berger TG, eds. Cutaneous lymphoid hyperplasia, cutaneous Tcell lymphoma, other malignant lymphomas and allied disseases. Andew's diseases of the skin. 9th ed, New York WB. Saunders, 2000; 921-929.

3. Beridon AMT, Giles F, Durvic M, Fayad L, Kurzrock R. Phase II study of pentostatin in advanced T cell lymphoid malignancy, update of M.D. Anderson cancer series. *Cancer* 2004; **15**: 342-349.

4. Capalb S, Declin M, Dargemio M, Liso A, Diomode D, Garofalo L, Liso V. Mycosis fungoides / sezary syndrome - a report of three cases treated with campth - 1H as salvage treatment. *Med Oncol* 2003; **20**: 389-396.

5. Kallimich T, Mche JM, Qin S, Sterry W, Kroczeki RA. Chemokine receptor expression on neoplastic and reactive T cell in the skin at different stages of mycosis fungoides. *J Invest Dermatol* 2003; **12**: 1045-1052.

6. Bolli N, Di Lanni M, Simonetti S, Cerroni L, Liso A, Falini B, Tabilo A. Treating to concurrent B cell and T cell lymphoid neoplasm with Alemtuzumab monotherapy. *Lancet Oncol* 2004; **5**: 64-65.

7. Gathers RC, Cherschun LS, Malik F, David P, Fivenson, Lim H. Narrowband UVB phototherapy for early stage mycosis fungoides. *Journal of American Academy of Dermatology* 2002; **7**: 191-197.

S A Uwyse, *Registrar*, **S P W Kumarasinghe**, *Consultant Dermatologist, Dermatology Unit, North Colombo Teaching Hospital, Ragama.*