Cutaneous T-Cell Lymphoma (Mycosis Fungoides)

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Introduction

Cutaneous T cell lymphoma (CTCL) is an uncommon fatal skin condition in which there is an abnormal neoplastic proliferation of T cell lymphocytes [1]. The entity varies considerably in clinical presentation, histological appearance, immunophenotype and prognosis of its variants [2,3]. Mycosis fungoides (MF) is the commonest subtype of CTCL. It may develop de novo or may persist in one stage for years or slowly progress from patch to plaque stage and later to a nodule. Spread to other organs is most likely in this stage, leading to a rapid, progressive, downhill course [3]. Fatality is usually associated with sepsis. We present two cases of MF.

Case Report 1

A 30 year old serving soldier, was admitted with the complaints of gradually increasing nodular swellings over the shoulder of two months duration. Dermatological examination revealed multiple sized infiltrated erythematous scaly plaques over the trunk and extremities. The swellings were three in number and the largest measured 8x6 cm in size (Fig.1). The skin overlying the swellings was red, thin and stretched. One of the swellings had ulceration over its surface. The patient was febrile. Ultrasonography, contrast enhanced computed tomography (CECT) chest and abdomen showed multiple enlarged lymph nodes in the right para tracheal and tracheal regions. There was a mass lesion over the neck of the pancreas measuring 5.3 x 4.2 cm. Both the kidneys were normal in size but showed multiple poorly enhancing focal mass lesions. The skin overlying the tumour on biopsy revealed characteristic features of MF with Pautrier's microabscesses (Fig.2), with dense polymorphous inflammatory infiltrates in the dermis containing a few atypical small and medium sized lymphocytes with cerebriform nuclei (Fig. 3). The patient was diagnosed in advanced tumour stage of MF affecting internal organs and transferred to oncology center for further management.

Case Report 2

A 90 year old patient, presented with an oval shaped red, itchy patches over the skin of feet. The lesions were waxing and waning for last three months but progressing in size. Vital parameters were within normal limits. Skin examination showed a few small macules and plaques with surface showing hyperkeratosis. Skin biopsy showed polymorphous inflammatory infiltrates with scant atypical lymphocyte having cerebriform nuclei expanding the dermis (Fig. 4). There was apparently no intraepidermal and superficial dermal infiltration appreciated under low power magnification however high power examination revealed small and medium sized malignant T-cell with characteristic cerebriform nuclear contour lined up along the basal layer. The patient was diagnosed in nonepidermotropic stage of MF, however involvement of internal organs could not be ascertained, as the patient was lost to follow-up.

Fig. 1 : Patient with three swellings on the shoulder

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Fig. 2 : Skin showing Pautrier's microabscess in epidermis and dense polymorphous inflammatory infiltrates in the dermis.

Discussion

MF has been classified as a cutaneous form of T-cell lymphoma and the diagnosis is made on biopsy. Approximately 75-80% of primary cutaneous lymphomas are CTCL and rest are represented by cutaneous B cell lymphomas [2]. Revised European American Lymphoma classification has classified CTCL, as a low-grade T-cell lymphoma [4]. It should be distinguished from other histologic types of T-cell lymphomas that involve skin such as anaplastic large cell lymphoma, peripheral T-cell lymphoma, adult T-cell leukemia/lymphoma and subcutaneous panniculitic T-cell lymphoma [5]. The accurate evaluation of neoplastic cells can be made by determining ultra structurally the value of the nuclear index, which is not practical in all laboratories [6]. So is DNA ploidy determination through flow cytometry and molecular studies for the identification of T-cells [3]. In the last few decades, MF and Sezary's syndrome (SS) were the only known types of CTCL. This entity now based on the clinical, histological, and immunophenotypical criteria, also includes follicular cell lymphoma, T-cell lymphoma and T-cell leukemia. SS is a variant of MF, which presents clinically by infiltrative erythroderma with pruritus, lymphadenopathy, and presence of small and large (Sezary's) cell in the peripheral blood [3].

The aetiology of MF is not known, but in some it is associated with pre existing contact dermatitis. A few unsubstantiated reports of its association with retrovirus

Fig. 3 : Atypical small and medium sized lymphocytes with cerebriform nuclei.

Fig. 4 : Polymorphous inflammatory infiltrates with scanty atypical lymphocytes with cerebriform nuclei, epidermis not involved.

infection similar to human T cell lymphotropic virus (HTLV-1), associated with adult T cell leukemia/lymphoma have been reported [8]. It is more common in men than in women. Majority of the cases are adult and elderly people [3].

MF has a plethora of clinicopathological manifestations having three distinct stages: premycotic, mycotic and tumorous. In the premycotic stage the skin is erythematous, scaly and pruritic but at times it thins out and may not itch. It may be difficult to distinguish clinically from psoriasis/parapsoriasis and discoid eczema [1,3]. Initially the microscopic findings may not be diagnostic as it may represent as chronic non-specific dermatitis. These lesions disappear spontaneously or enlarge slowly. In the mycotic stage, patches may be thickened and resemble psoriasis. Histopathology is characteristic with polymorphous inflammatory cells with small or medium sized lymphocytes, having cerebriform nucleus. They infiltrate dermis and later invade epidermis to form Pautrier's abscesses. In the advanced stage,

the irregular lumps may ulcerate and the disease may become systemic involving other organs. The pathologist has to distinguish it from many benign conditions, such as lymphomatoid papulosis, cutaneous lymphoid hyperplasia and Jessner's lymphocytic infiltration of the skin.

In first case, the diagnosis of MF was entertained on clinical examination and there was no difficulty in establishing the diagnosis on biopsy. In the second case, the skin biopsy showed linear accumulation of lymphocytes associated with basement membrane zone (lentiginous pattern). There was diffuse dermal infiltrate with relative sparing of superficial dermis and epidermis. In view of the presence of atypical lymphocytes with convoluted hyper chromatic nuclei, the diagnosis of nonepidermotropic MF was made.

MF is a cutaneous T cell lymphoma where architecture pattern of T cell show characteristic epidermotropism in the initial stages of the disease whereas advanced stages develop as a consequence of exclusive dermal involvement by nonepidermotropic, cytologically uniform atypical malignant T- cells [9].

The mean survival following diagnosis of MF, is dependent on the stage. Patients with stage I, may have mean survival of 20 or more years whereas more than half of the patients in stage III/IV die in less than five years [7,10]. Upto two third of the patients of MF of skin have involvement of the lymph nodes and internal organs. The commonest cause of death in these cases is sepsis due to *P aeruginosa* or *S aureus* infection. For treatment of MF/SS, topical and systemic steroids, chemotherapy, localized/electron beam radiotherapy, oral retinoids and UVB phototherapy, photopheresis, and PUVA photochemotherapy are recommended, depending upon the stage. Unlike other lymphomas the

symptoms are usually controlled with treatment but the disease is not curative.

Conflicts of Interest

None identified

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