Mycosis fungoides - case report

Dr. Bader Faiyaz Zuberi[1] M.B.B.S., Registrar Medical Unit - III, Civil Hospital Karachi
Dr. Mukhtar Azeem Mirza, M.B.B.S. (Kar) F.R.C.P. (London); F.R.C.P (Edin), F.C.P.S. (Pak); D.T.M.&.H (Eng). Professor Medical Unit - III, Civil Hospital Karachi
Dr. M.U. Islam, M.B.B.S.; M.R.C.P. (UK); F.R.C.P. (Edin) D.C.H. (GLA), Associate Professor Medical Unit - III, Civil Hospital Karachi
Dr Syed Hamid Zaki, M.B.B.S.; D.D.V. (Aus)
[1]*For correspondence and reprints*

Abstract

Case report of Mycosis Fungoides; and T-cell Lymphoma of skin. The patient underwent extensive workup and posed diagnostic enigma for years. It is characterized by the infiltration of the skin with plaques and nodules composed of T-Lymphocytes.

Key Words

MYCOSIS FUNGOIDES
T-CELL LYMPHOMA
CUTAINEOUS LYMPHOMA
SKIN TUMORS

Case Report

T.U.H. a 48 years old school teacher was admitted in CHK with the complains of reddish patches all over the body since childhood, ulcers on left foot since 22 months, and an ulcer over the sternum since 15 months. He had consulted many doctors for his ailment. There was itching in the red patches which was severe in the night. The patches were initially limited to the face especially over nose, lips and ears. They gradually increased and spread to all over the body. In 1967 he was admitted in the All India Institute of Medical Sciences where he was investigated thoroughly including for Leprosy, but nothing conclusive came up and he was discharged on multivitamins. In 1983 he came to Pakistan and here too consulted many doctors. 22 months back he developed 2 ulcers on his left foot with pussy discharge which refused to heal. 15 months back he developed similar ulcer over the sternum. Mantoux Test was done which was negative. He was given steroids (Deltacortil 6 tabs/day) for 6 weeks considering the diagnosis of immune allergy dermatosis. There was no improvement. Mantoux was repeated which came positive with 12 mm of induration, anti-tuberculous therapy was started which failed too. He was then refered to the Marie Adeliade Leprosy Center Karachi where they did specific tests for leprosy and concluded that he was not having Leprosy. At this stage he came to the OPD of CHK and was admitted in M.U. - III.

On examination his pulse was 78/min., BP-135/80 mm of Hg, temp.- 99.0° F. Except for the anaemia there was nothing significant in the general examination. His whole skin was covered with erythematous plagues with fine scales and pruritus was present. On the lower limb there were multiple small nodules with ulcerations which were secondarily infected. The ulcer over the sternum measured 5 x 3 cm while that on left foot measured 5 x 8 cm. Both the ulcers were having a necrotic base with areas of granulation tissue, the margins were irregular. There was nothing significant in the systemic examination.

His investigations showed Hb = 11.0 gm%, TLC = 6,500/cu mm, DLC = normal ESR = 88 mm, Peripheral film = Normocytic normochromic no atypical cells seen, Urine DR = Normal, ANA = negative. Anti ds DNA = negative, Mantoux Test = 12 mm, VDRL = negative, Ultrasound abdomen = normal.

Keeping in view of history and previous lab. reports a provisional diagnosis of Mycosis Fungoides was made.

Several biopsies were taken from the edge and base of both ulcers. The histopathological findings were consistent with the diagnosis of **MYCOSIS FUNGOIDES.**

The patient was found to be in stage T_4 N_0 M_0 B_0. Chemotherapy was started with Tab Chlorambucil 5 mg once daily. The patient responded very well and after 6 months of therapy; his erythematous patches were cleared and the ulcers on the foot and sternum were healed. His latest ESR was 30 mm of Hg.

Discussion

Mycosis Fungoides (MF) is a rare disease. It is a Cutaneous T-cell Lymphoma characterized by the infiltration of the skin with plaques and nodules composed of T-lymphocytes. The literature was reviewed. The cause of MF is un-

known. At present, there are three current theories concerning its actiology. These relate to; antigen persistence; a role for retroviruses; and a relationship to industrial exposure to the carcinogens especially related to petro-chemical, textile, metal and machine industries[1].

The histological picture of fully developed MF is that of a dense lymphocytic infiltrate which occupies the papillary dermis and extends up into the epidermis forming Pautrier micro-abscesses. The lymphocyte marker studies in MF have established that the majority of the lesions biopsies bear the CD4 antigen the marker for the T-helper subset[2,3].

Staging[4]

Cutaneous involvement (T)

T_0 Suspicious Lesions

T_1 Plaques involving <10% of skin

T_2 Plaques involving > 10% of skin

T_3 Tumors Present

T_4 Erythroderma

Lymph nodes (N)

N_0 Normal

N_1 Palpable. Pathologically not involved

N_2 Not palpable. Pathologically MF

N_3 Palpable & pathologically MF

Viscera (M)

M_0 No visceral spread

M_1 Visceral spread present

Peripheral Blood (B)

B_0 No atypical circulating cells

B_1 Atypical circulating cells present.

Clinical Features

Mycosis Fungoides can present in four different forms.

Plaque MF develops slowly usually first involving the buttocks. There are round or oval plaques with fine scales, erythema and pruritus. Secondary infection is common.

In **Tumer d'emblee** form the patient develops large nodules without prior presence of the plaques.

In the **Erythrodermic** form there is rapid development of erythroderma progressing to lymphadenopathy and peripheral blood involvement.

The **Poikilodermatous MF** is characterized by wide spread poikiloderma usually involving trunk, breast and buttocks severely.

The clinical variety of MF does not carry prognostic significance once correction is made for the disease.[5]

Treatment

1. **Tropical steroids and UVB.** It is effective in the plaque stage of the disease especially during the winter.
2. **Photochemotherapy (PUVA).**[6] Many studies have shown the efficacy of the photochemotherapy regarding the symptomatic and clinical response.[7]
3. **Tropical Nitrogen Mustard.**[8] This drug has also shown good results, it is dissolved in water and painted over the entire body once or twice weekly. Use of nitrogen mustard in the ointment base is more stable and economical.[9]
4. **Chemotherapy.** The chemotherapy is usually given as a single drug therapy with chlorambucil. In contrast to the lymphomas; the results of chemotherapy in MF are disappointing; but our patient responded very well to chemotherapy. No difference in the disease free interval or survival was found in the patients receiving electron-beam plus chemotherapy and those receiving topical therapy.[10]
5. **Electron-beam Therapy.** It gives good symptomatic relief in the later stages of the disease. The usual dose is 20-30 Gy over a period of 4-6 weeks.[11]
6. **Interferons.**[12] The role of interferon is under evaluation.
7. **Photopheresis.** In this type of therapy the patient ingests psoralen and is attached to the cell separator in which the cells are sequestrated. These cells are then passed in a very thin tubing through a UVA beam and the cells are then returned to the patient.

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