MYCOSES FUNGOIDES

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Introduction

Mycosis Fungoides is an uncommon, chronic, poorly understood fatal disorder, first described by Alibert in 1806 and later by Bazin in 1851, originates in the reticulo-endothelial cells of the skin; lymph nodes and internal organs get involved secondarily. References in Indian Literature are very few. This is the first case at Medical College Hospital, Nagpur.

Clinical features of the case

A 52 year old male presented with the complaints of itching, red patches with scaling and cutaneous swellings of various sizes of 6 months in duration. It started with itching all over the body and urticarial patches for which he was treated with local applications and anti-histaminics. Four years ago he was admitted to the skin-ward of this hospital for urticaria, was treated for the same and discharged. Clinical examination during this admission revealed multiple swellings varying from size of a pea to a lemon (Fig. 1 & 2) distributed randomly over face, chin, shoulder, back, thighs and legs. In addition there were a large number of plaques, papules and nodules distributed all over the body. Quite a few of these were red but other signs of inflammation were absent. Scaling was present only over a few lesions. Axillary and cervical lymph nodes were enlarged, firm non-matted, fixed to skin, non-tender and in right axilla ulceration was present. Inguinal lymph nodes were also enlarged moderately on both sides. Liver and spleen were only just palpable. Other systems were normal.

Investigations

Hb–80%; T. L. C. 4,200/C.mm.; D.L.C.–P–60%, L–36%, E–4%. No immature cells were seen in peripheral smear. Bone marrow was Normoblastic with normal Myeloid Erythroid ratio; Blood for L.E. was Negative; Blood K.T. and V.D.R.L. were Negative; Post Meal (1½ hours) Blood Sugar 133.3 mgm.%; Mantoux test Negative, Chest X-Ray was normal.

Biopsies were performed twice from the cutaneous nodules. First one was from the face and second from the nodule on pectoral region. Sections from the nodule showed marked atrophy of the epidermis and dense infiltration in dermis by lymphocytes, mononuclears, with occasional plasma cells and eosinophils Few large monocytic cells with hyperchromatism and mitosis were scattered diffusely. The picture was of cellular pleomorphism. Histopathologically a diagnosis of Mycosis Fungoides was confirmed. (Fig. No. 3).

On receiving this report Endoxan 200 mgm. daily intravenously was begun. A total of 2.8 gms. was administered in 14 days. The regression of the lesions was only marginal. But itching and erythema subsided. However, when

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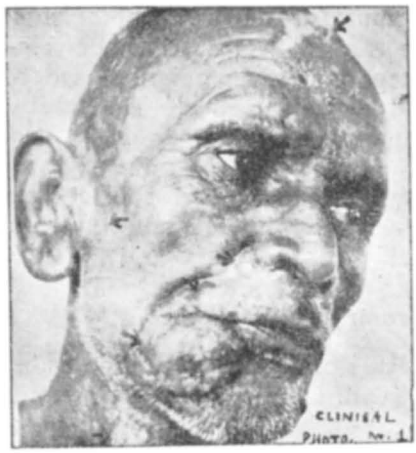

Fig. 1

Fig. 2

Clinical photographs showing multiple swellings on the skin of varying sizes situated over face, chin, shoulder, back, thighs and legs.

liver biopsy and a follow-up skin biopsy (to see the result of treatment) were planned, the patient went home on request to see his ailing aged mother and did not return for further follow-up.

Discussion

Mycosis Fungoides is more common in males. The history of the disease may be as long as 20 years. In the first

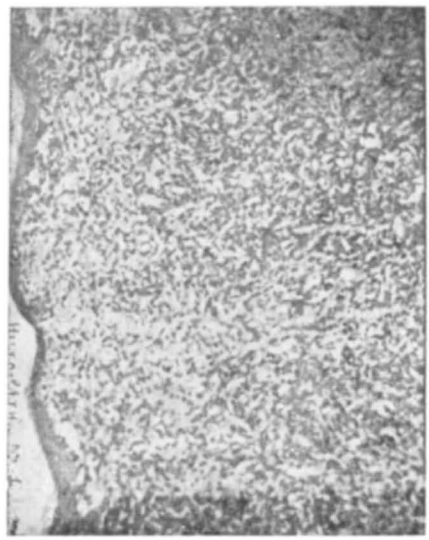

Fig. 3

Photomicrograph of cutaneous nodule shows dense cellular infiltration in the deeper tissue. The picture is of cellular pleomorphism.

pre-mycotic stage the cutaneous eruptions may be missed for non-specific dermatitis, psoriasis, eczema, urticaria etc., but they fail to respond to the usual treatment for these disorders. Vesicles and itching may be present. The second or Infiltration stage is characterized by thickening of skin with duskyred hue, scaling and itching. Histology at this stage reveals pautrier's Abscesses with pleomorphic cellular infiltration. Last is the tumour stage (in which our patient was admitted). Involvement of nodes is most common, however internal organ involvement may be as high as 94%[3]. Involvement of liver, spleen, kidney, heart, G. I. tract, endocrines, Central Nervous system, bones and testes have been reported[9]. Respiratory system may be involved[2, 7, 8, 9]. Various types of the disorder are also recognized. (i) Generalised Erythroderma (ii) Tumour d' emblee' in which tumour stage develops suddenly on the normal skin and histopathology is frankly malignant. (iii) Isolated plaques with bullous eruptions, (iv) Purpuric lesions, (v) Poikiloderma atrophicans vasculare

showing atrophic epidermis and infiltrate of mycosis (vi) Se'zary Syndrome-characterized by lymphocytic leukemoid reaction.[2, 10, 6] Visceral involvement preceding cutaneous lesions has not been reported.[2] Bone marrow involvement is rare. Immunological reactivity in mycosis fungoides has been extensively studied by Blaylock et al. Its relation with other lymphomas has also been discussed by various workers.[2]

Prognosis

Prognosis is uniformly poor and death results due to respiratory infection, septicemia, result of treatment, Intercurrent illness or inanition and exhaustion.[8]

Treatment

Various forms of treatment-varying from topical application of fluocinolone acetonide[5] to systemic administration of cytotoxic drugs.[11] Apart from nitrogen mustard and cyclophosphamide, methotrexate, Velban, Actinomycin-D, Azotepa and streptonigrin, have been found to be effective. The results of treatment are rarely dramatic. Most of the patients die within 3 years on Lymphnode and visceral involvement. Five-year survival rate is only 15 percent.

Summary

A case of Mycosis Fungoides is being presented. The patient was admitted for itching, scaling and multiple cutaneous swellings with hepato-splenomegaly and lymphnode involvement. Histopathology confirmed the diagnosis. Response to treatment with Endoxan was unsatisfactory

Acknowledgment

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REFERENCES

1. Blaylock W K: Normal immunological reactivity in patients with the lymphoma mycosis fungoides, Cancer, 9 : 233, 1966.
2. Block J B: Mycosis Fungoides-Natural History and Aspects of its relationship to other malignant lymyhomas, Amer Jr Med. 34 : 228, 1963.
3. Cawley E P: Is mycosis fungoides a reticulo endothelial neoplastic? A M A Arch Derm 64 : 255, 1951.
4. Cecil-Loeb: Text book of Medicine, 1967, 1903.
5. Eugene F: Therapy of mycosis fungoides with topically applied Fluocinolone acetonide under occlusive dressing, Cancer, 19 : 237, 1966.
6. Lewer W F: Histopathology of skin, 1961, P 615.
7. Pillsbury D M, Shelley W B, & Kiligman S M: Dermatotogy 1956, P 1089.
8. Rook A, Wilkinson D S & Ebling F J: Text book of Dermatology, 1968.
9. Singh, Shurvir & Shah D R: Mycosis Fungoides-Case Report J Assn Phys India, 13 : 209, 1965.
10. Taswell H F et al: Se'zary Syndrom-A Malignant reticulemic erythroderma J A M A 177 : 465, 1965.
11. Wright C et al: Observations on the use of Cancer therapeutic agents in patients with mycosis fungoides; Cancer, 17: 1045, 1964.