MYCOSIS FUNGOIDES

An Analysis of Thirteen Cases

J. F. DORSEY, M.D., and E. W. NETHERTON, M.D.
Department of Dermatology

THIRTEEN patients with mycosis fungoides have been seen at the Cleveland Clinic. Nine were men and 4 women. The youngest patient to develop the disease in this series was a 22-year-old woman and the oldest a woman aged 71.

Twelve of the patients were white and 1 was colored. Occupation was not significant. The duration of the disease before being seen at the Clinic varied from 1 month to 10 years.

One patient developed dermatitis venenata which was followed by a generalized exfoliative dermatitis on which lesions of mycosis fungoides developed 4 years later. Another patient had "psoriasis" for 30 years and then developed mycosis fungoides. One patient had mycosis fungoides of the d'emblee type. Nine complained of pruritus and in 3 cases this was severe. Sensation of burning was present in 1 case. Two patients were asymptomatic. Enlargement of the superficial lymph nodes was observed in 6 patients.

On admission to the Clinic 2 patients had exfoliative dermatitis; the others presented nodules, erythematous scaly plaques, ulcers, papules, and macules. Concomitant diseases were syphilis, psoriasis and carcinoma of the esophagus.

Diagnosis is often difficult. The clinical diagnosis of mycosis fungoides or lymphoblastoma was entertained in 11 cases on entrance to the Clinic. In 1 case syphilis was first considered; in the patient with the d'emblee type of lesion, three possibilities were first considered in differential diagnosis: (1) neoplasm, (2) factitia dermatitis, and (3) melanoma.

In 10 cases biopsy showed histologic pictures consistent with mycosis fungoides. Three were diagnosed as lymphoblastoma histologically but as mycosis fungoides clinically.

Complete blood studies were done on all patients. Initially all were within normal limits. Three of the patients in the series had sternal marrow examinations. All were reported as nonspecific chronic inflammation. In 2 cases lymph nodes were removed and examined but were reported as nonspecific.

All of the patients received roentgen therapy at some time during their illness. This appeared to be the one method that could be depended upon to bring a temporary improvement in most cases. There was, however, great variation in individual response. Two patients became resistant to roentgen therapy after a while. One patient had approximately a total of 11,000 r of

superficial roentgen therapy and three courses of nitrogen mustard but continued to develop lesions when treatment was stopped.

Sodium arsenate, potassium bismuth tartrate, testosterone, manganese, urethane, paraminobenzoic acid, stibanose, nitrogen mustard and transfusions were used. Two patients received three courses and 1 patient received two courses of nitrogen mustard with temporary clinical and symptomatic relief. As in the other cases reported in the literature, those patients who were given nitrogen mustard developed leukopenia and a decrease in hemoglobin and red blood cells. It was also noted that the effects from subsequent courses of nitrogen mustard were not as great as the effect from the initial course. One patient received little relief from a second course of nitrogen mustard and was given roentgen therapy for a while after which a course of nitrogen mustard was repeated. The patient responded rapidly to the therapy temporarily and then relapsed. Remissions were never complete clinically with the drug and did not last longer than 2 months. Stibanose was not as spectacular in its results as nitrogen mustard. Two out of 3 patients noted a little improvement subjectively.

Of the 13 patients, 5 have died, 6 are living and 2 have disappeared from observation. The causes of death were carcinoma of the esophagus, pneumonia, and 1 was reported to have died of leukemia. Cause of death in the other 2 is unknown. One patient has had the disease for 14 years and is still living.

Discussion

Mycosis fungoides is a chronic disease of unknown etiology, primarily of the skin, occasionally with later involvement of the internal organs; characterized by remissions, exacerbations and various clinical stages. It occurs in the latter decades of adult life, predominantly in white men.

The name is a misnomer as the disease is not of mycotic origin.

Mycosis fungoides has been included as a member of the lymphoblastoma group, which consists of Hodgkin's disease, leukemias and lymphosarcoma. Some authors have felt that these are clinical pictures of the same disease process.

Signs and symptoms vary with the stage of the disease. Severe pruritus, pricking and burning may be present long before lesions appear in the skin. Systemic symptoms as fever, anorexia, and weakness, usually do not appear until later.[1]

The disease may begin as an exfoliative dermatitis, universal erythroderma, erythematous eczematoid eruption, poikiloderma,[2,3] and later develop infiltration of the skin with the formation of tumors and ulcers. The onset can be of the d'emblee type, in which tumors develop on the normal skin without the eczematoid phase. Any part of the cutaneous surface may be involved and Oliver[4] mentions some cases in which there were metastases to the lungs, pleura, mediastinum, tracheal mucosa, epicardium, soft palate, lymph glands, and skeletal musculature.

Death may occur from 6 months to 40 or more years after onset of the disease. According to Montgomery,[1] "The average duration of life was about 5 years and longer with treatment, which is much greater than in the other types of lymphoblastoma." Spontaneous involution of tumors has occurred without treatment, and then the tumors have reappeared years or months later.

The histopathologic changes vary in the different stages of mycosis fungoides. In the eczematoid stage, there is acanthosis and micro-abscesses containing lymphocytic cells. The cutis contains a polymorphic infiltrate consisting of eosinophils, endothelial cells, plasma cells, polymorphonuclear leukocytes, lymphocytes, pseudogiant cells, histiocytes, reticulo-endothelial cells, connective tissue cells with karyorrhexis and pyknosis of nuclei. The tumorous stage resembles a reticulum cell sarcoma as the cells usually appear more uniform and monomorphic. There is definite increase of reticulum fibers in both stages and there is a proliferation of the endothelial cells lining the blood vessels in the cutis, which has been considered as proof that mycosis fungoides is a disease of the reticulo-endothelial system. It is also transformed to other lymphoblastomas, especially lymphosarcoma according to Montgomery.[1]

Numerous drugs and forms of therapy have been used but the results have all been temporary.

Summary

Of 13 cases of mycosis fungoides diagnosed at the Cleveland Clinic 1 is still living who has had the disease for 14 years. There is no permanent satisfactory treatment for mycosis fungoides at present. Roentgen irradiation seems to be the best temporary method of treatment. Nitrogen mustard in our experience was not as beneficial over a period of time as superficial roentgen therapy, and its toxic effects limit its value as a therapeutic measure in this disease.

References

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