

## MYCOSIS FUNGOIDES - TUMOR D' - EMBLEE

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### Summary

Two cases of mycosis fungoides, tumor d'-emblee are reported and discussed.

Mycosis fungoides first reported by Alibert<sup>1</sup> in 1806 as "Une eruption furfuracee", is a relatively rare, chronic itchy dermatoses. Bazin in 1851 named it "Mycosis fungoides" and described its three stages - the premycotic, the plaque and the tumor stage, now well recognised. Mycosis fungoides runs a progressive course with infrequent remissions and is usually fatal. It is generally considered to be a type of lymphoma which affects the skin primarily and predominantly, but in the late stages, the internal organs may be involved. Histopathology of first or premycotic stage is not characteristic but it is usually diagnostic in both plaque and tumor stages.

Vidal and Brocq<sup>2</sup> described the tumor d'emblee form of mycosis fungoides, in which only the tumor stage is seen clinically. It is a rare type of mycosis fungoides and the authors are aware of only 47 published case reports in literature: 40 cases collected and documented by Bluefarb<sup>3</sup>, 5 cases by Block et al<sup>4</sup> and one case each by Saint-Andre P. et al<sup>5</sup> and Singh and Shah<sup>6</sup>. From India, only two cases have been reported: one by Basu et al<sup>7</sup> included in Bluefarb's<sup>8</sup> series of 40 cases and the other by Singh and Shah<sup>6</sup>. The authors wish to place on record two

additional cases of mycosis fungoides-Tumor d'emblee, from the Indian sub-continent.

### Case Report I

A 52 years old male was admitted to the Dermatologic service of Irwin Hospital, New Delhi on 30—10—1970, with multiple nodular swellings and tumors on the body of six years duration; one of them destroying his left eye four years back. One of the tumors was excised and others treated by x-ray therapy at different hospitals and showed temporary regression to irradiation. Six months prior to admission in Irwin Hospital, the swellings had relapsed again. There was no loss of appetite and weight was maintained.

Examination revealed an elderly man with multiple, nodular swellings and tumors on the scalp, forehead, face, neck, abdomen and thighs, of varying sizes, ranging from 1 cm x 1 cm to 7 cm x 5 cm (Figs. 1 and 2). Skin overlying the bigger swellings was slightly erythematous and shiny. Most of the swellings and tumors were firm, while a few were soft and mildly tender. Two ulcerated areas, each of 6 cm x 4 cm on the medial side of left thigh and just above umbilicus, with necrotic floor, were also present. There was no significant lymphadenopathy. Systemic examination did not reveal any abnormality. Prostate and adenexa were normal.

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**Fig. 1**

Case I — Nodular and tumor masses affecting the face and neck

**Fig. 2**

Case I — Nodular swellings on the abdomen

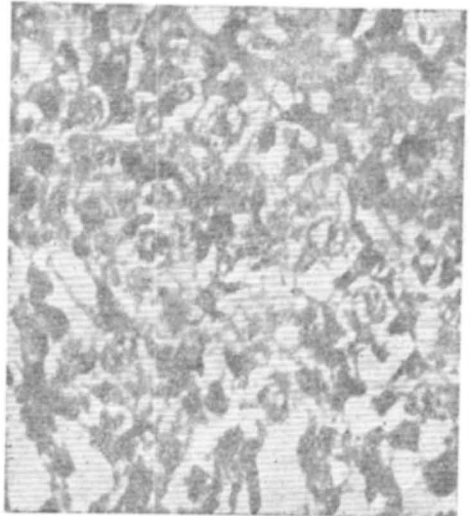
### Investigations

Hb. 14 gm%. Total leukocyte count 9000/cmm. with Polymorphs 64%, Lymphocytes 30%, Eosinophils 5% and Monocyte 1%. E.S.R. was 100 mm/1st hour. B.T.-2 minutes 30 seconds; C.T.-4 minutes 40 seconds. Platelet count-140,000/ cmm. Serum proteins-Total 6.9 gm%; Albumin 3.1 gm%; Globulin 3.8 gm%; Ratio 0.8. Stools and urine-N.A.D. S.T.S.-non-reactive. Sternal bone marrow picture-no abnormal cells seen. X-ray chest and E.C.G. were normal.

Histopathological picture of skin biopsy from one of the nodular lesions on abdomen was that of reticulum cell lymphoma, showing preponderance of immature reticulum cells and only few inflammatory cells (Fig. 3).

### Treatment and Progress

Nitrogen mustard, a total of 40 mg; 10 mg. by I.V. drip on alternate days was given but without any effect. On 14—12—1970, radiation therapy was started and a total of 1400r was given on each site, on face, neck and abdomen. A dramatic regression in all the

**Fig. 3**

Case I — Photomicrograph of one of the nodular lesions showing dermal infiltrate, consisting mainly of immature reticulum cells and few inflammatory cells. (H. and E. x 100).

lesions was seen, but soon they started growing again along with some fresh lesions. These again responded temporarily to irradiation (1000 r) combined with Cyclophosphamide (100 mg) daily

by oral route. Subsequently, Cyclophosphamide, intralesionally as well as I.V., 200 mg on alternate days, till a total of 2,600/ mg, was also tried but without any effect.

On 15-4-1971, patient developed left sided hemiplegia with supranuclear facial palsy and later became delirious and comatosed. Skin lesions had increased in size, many of them ulcerating and bleeding profusely. He progressively went downhill, terminally developed lung infection and finally died on 3-5-1971.

Partial autopsy was done — a portion of spleen, liver and lymphnodes were taken out. Gross examination showed areas of infarcts in spleen only. Histopathology revealed a normal architecture of spleen: in the red pulp, there was marked infiltration of plasma cells; many of them were seen in clusters. There was proliferation of reticulum cells, some of them were quite big (Fig. 4). Several small and large areas of infarcts were also present. (Fig. 5). Liver and lymphnode did not show any pathology.

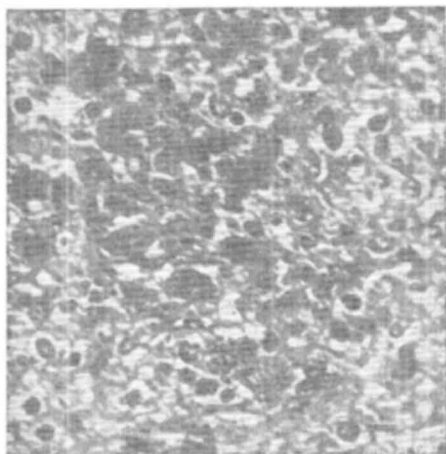


Fig. 4

Case I — Photomicrograph of spleen showing infiltration of red pulp by plasma cells and proliferation of immature reticulum cells (H. and E. x 100).



Fig. 5

Case I — Photomicrograph from one of the areas of infarcts in spleen. (H. and E. x 40)

#### Case Report II

A 47 years old female, attended the skin O.P.D. of Irwin Hospital on 16-11-1970, with the history of a slowly progressing, mildly tender, tumorous swelling in the left groin of 2 years duration. It was cauterized twice at another hospital and finally excised about a year back, when she remained free for 7 months and then it slowly recurred at the same site.

She also had a dry ichthyotic skin since childhood, which improved in summer and relapsed in winter, sparing the flexures. Family and personal history were non-contributory.

Examination revealed an apparently healthy woman of past middle age. There was no lymphadenopathy. Spleen and liver were not palpable. Various other systems were normal.

Cutaneous examination showed near the left groin, a skin coloured swelling, 7 cm. x 1 cm., with a healed incisional

scar in its centre and two nodules, each 1 cm. x 1/2 cm. at its periphery. They were mildly tender and soft in consistency. Overlying skin and underlying structures appeared normal. Medial side of the left knee had a skin coloured asymptomatic, nontender papular lesion, 1/2 cm. x 1/2 cm. The rest of the skin was normal except for mild degree of ichthyosis, sparing the flexures.

### Investigations

Hb. 15.8 gm%. Total leucocyte count 4000/cmm, with Polymorphs 59%; Lymphocytes 39%; Monocytes 1% and Eosinophils 0%. Montoux test—16 mm and persisted for 2 weeks. X-ray chest was normal. Aspirated pus from left groin lesion—smear examination showed a few gram positive cocci but culture was sterile. No A.F.B. or deep mycosis could be demonstrated.

### Histopathology

(1) Papular lesion from medial side of left knee was excised. Histopathology was that of Fibromatosis of Dermis.

(2) Histological examination of skin biopsy from groin lesion was inconclusive.

Patient was given 40 gm. of Streptomycin and I.N.H., followed by injection Acromycin (100 mg. I/M, twice daily for 2 weeks), but there was no appreciable improvement. On 9—3—1971, whole of the lesion from medial side of the left thigh with sufficient healthy skin was excised. The wound healed by primary intention in 2 weeks time.

Histopathologic picture of excised tissue was consistent with mycosis fungoides (Fig. 6). It showed pleomorphic infiltrate, consisting of plasma cells, lymphocytes and reticulum cells in the dermis and subcutis. Some of the reticulum cells were big with vesicular nucleus and prominent nucleoli: large

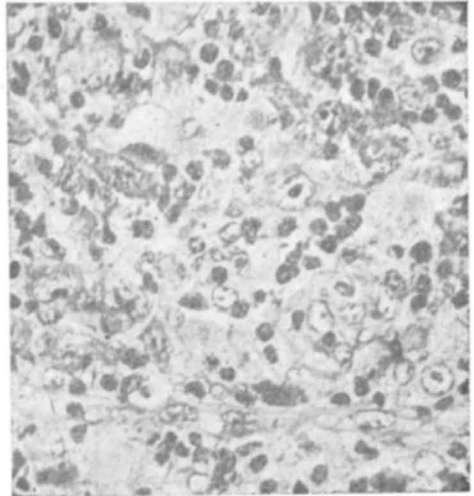


Fig. 6

Case II — Photomicrograph of skin, showing dense and pleomorphic infiltrate in the dermis. Immature reticulum cells, mycosis cells, plasma cells and lymphocytes are seen. (H. E. x 100)

number of mycosis cells with angular or binucleate nucleus were also seen. No Pautrier "microabscess" could be seen in the epidermis. Soon the patient left for Madras (South India), where she received "Cobaltbomb" on left groin; exact dosage not known. Patient has since been coming regularly for checkup. She was last seen in November, 1972. She had no complaints except for cancerphobia. She was given a thorough checkup and found normal.

### Discussion

Mycosis fungoides is a confused disorder and has been variously defined by different schools having different concept of it. Many do not even regard it as a disease entity. Symmers<sup>8</sup> has called mycosis fungoides a "Clinical and pathologic non-existent" condition and stated that cases reported as mycosis fungoides represented histopathologi-

cally "Hodgkin's disease, reticulum cell sarcoma or lymphosarcoma"; while others (Highman<sup>9</sup>, Berman,<sup>10</sup> Cawley et al,<sup>11</sup> and Clendenning et al.<sup>12</sup>) are of the view that mycosis fungoides is a clinical entity only, not a histopathological one. Clendenning et al<sup>12</sup> observed that all this confusion was, because emphasis was placed on histopathology in what was essentially a clinical entity. Opinion as to whether mycosis fungoides (Tumor d'emblee) represents a nosologic entity, is still more fluid. Bluefarb<sup>3</sup> and Samman<sup>13</sup> regard all such cases as lymphosarcoma and reticulum cell sarcoma respectively. In the first case, though the patient was diagnosed as a case of mycosis fungoides tumor d'emblee on clinical grounds, still histopathologically, cell type was mainly immature reticulum cells with only few inflammatory cells and in the second case, the inflammatory infiltrate was pleomorphic with many reticulum cells. These histological findings can be explained on the basis of Fraser's<sup>14, 15</sup> postulations that mycosis fungoides from the very beginning is a reticulum cell lymphoma and all other cells represent merely a defence reaction of host against the invading tumor cells as is seen in early cases of squamous cell carcinoma and melanoma also. Thus, in the early stage of the disease, the number of tumor cells is small and inflammatory defence reaction very pronounced. As the disease advances, the number of tumor cells increases and inflammatory cells fall and finally only the tumor cells proliferate. This may explain, why the first biopsy from the groin of Case No. II, was inconclusive. The involvement of spleen by tumor cells in Case No. I, shows that systemic involvement had occurred. The reported incidence of specific visceral involvement has varied considerably, depending upon the concept of mycosis fungoides. Ormsby

and Montgomery<sup>16</sup> and Bluefarb<sup>3</sup> are of view that such involvement is rare, while Cawley et al<sup>11</sup>, Gall<sup>17</sup>, Block et al<sup>4</sup>, and Rosai and Spiro<sup>18</sup> found visceral mycosis fungoides in 80%, 46%, 82% and 62.5% respectively. In Case No. I, the tumor cells infiltrating the spleen were different from the reticulum cells seen in skin biopsy. Similar observations have been made by Block et al<sup>4</sup>, who believe that once visceral metastasis have occurred, histopathological examination of these metastasis show morphologic changes that can be identified as malignant lymphoma but often cannot be classified further. However, the concerted view of the authors is that all lymphomas are related and multifocal in origin, though very infrequently metastasis can take place. The existence of lymphomas of different cell types in the very same patient, not only in different biopsy specimens but in deeper cuts from the same biopsy specimen is not uncommon. Further more, the possibility of transformation of one form of lymphoma to another, particularly into more anaplastic types cannot be excluded. Thus, the different types of tumour cells in the spleen and skin biopsies is quite understandable. Left sided hemiplegia in Case No. I might have been due to involvement of brain by mycosis cells, though brain specimens could not be studied histopathologically. There are several reports of C. N. S. involvement in mycosis fungoides (Block et al<sup>4</sup>, Reed and Cummings<sup>19</sup>, and Rosai and Spiro<sup>18</sup>).

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## TOPICAL RETINOIC ACID IN THE TREATMENT OF ACNE VULGARIS

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### Summary

Retinoic acid was administered in 45 patients with grade I or grade II Acne Vulgaris. In 10 patients it was compared in a clinical study to sulfur — Resorcinol Shake lotion. Good to excellent response was obtained in 22 patients (48.8%) while fair response was noted in 9 patients (20.0%). In 14 patients (31.1%) there was no change. Retinoic acid was found to be effective in reducing the number of comedones to about 68-70% in both grades of Acne. When compared with the shake lotion it was found to be more effective in reducing the number of comedones (70% with Retinoic acid compared to 50% with shake lotion).

Burning of the skin, smarting pain and irritation were the main side effects noted. 9 out of 45 patients discontinued the medication on their own as a result of side effects. This was the major problem in continuing the therapy.

Retinoic acid has a significant effect on the removal of comedones as compared to the other available methods of treatment. Whilst it has only a moderate effect, in reducing papular lesions, its effect on the pustules and cysts is virtually negligible.

Vitamin A has been used in the treatment of Acne Vulgaris since many years. Straumjform (1943) first reported apparent clinical benefits after administration of Vitamin A. This was believed to be due to reduction in follicular hyperkeratosis. Cornbleet et al (1944) however, could find no abnormality in the plasma vitamin A level of patients with Acne. Lynch (1963) did a double blind trial in which he could not find any advantage of oral administration of Vitamin A over a placebo.

Patients with Ichthyosis, Darier's Disease, Pityriasis Rubra pilaris, Ichthyosiform erythroderma and Psoriasis were given Vitamin A by mouth. All except the patients with Psoriasis improved clinically after 2 to 3 weeks. Although the mechanism is mysterious the therapeutic efficacy of Vitamin A is not disputed. To avoid the hazards of systemic toxicity many trials of the topical application of vitamin A were undertaken with either Vitamin A alcohol or its palmitate. The results of these trials were conflicting and disappointing.

This paper was presented at the 26th All India Conference of Dermatologists, Venereologists and Leprologists held in January 1973 at Udaipur.

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Stuttgen and Von Beer (1962) appear to have been the first to apply Vitamin A acid (Retinoic acid) which is not known to occur in tissues but can be derived from oxidation in vitro from the aldehyde, which does occur naturally. These authors reported favourable results in Psoriasis and Ichthyosis