MYCOSIS FUNGOIDES - PAPULAR PHOTOSENSITIVE ERUPTION WITH BONE MARROW INVOLVEMENT AND GRANULOMATOUS HISTOLOGY

Ajita Bagai (MD),
S D Shenoi (DVD, MD),
Sandra A (MD, DNB),
P Ravikala V Rao*(MD)
Address for correspondence:
Dr Shrutakirthi D Shenoi

Key words:
Mycosis fungoides, granuloma, photosensitivity.

Case Report

A 26 year old female presented with intermittently pruritic, discrete to confluent erythematous papules with photosensitivity over the face, neck, limbs and trunk since 3 years (Fig 1). There were lichenified lesions with follicular prominence over the lower legs and forearms. Lesions tended to wax and wane but never subsided completely. Bilateral recalcitrant verruca vulgaris were present over the extremities. Discrete firm mobile non-tender lymphnodes were palpable in the cervical, axillary and inguinal regions. Systemic examination was normal.

Investigations were as follows: Hb - 10gm%, total leukocyte count - 1,56,900 cumm, ESR-24 mm/hr, platelets - 2,69,000 cumm. Peripheral smear showed markedly increased abnormal lymphocytes (98%) with adequate platelets. Chest X-ray, ECG, Ultrasound & HIV ELISA were normal. Skin biopsy from leg lesion showed dense infiltration of hyperchromatic pleomorphic lymphoid cells in upper dermis with occasional mitotic figures, epidermotropism (Fig 2) and Pautrier's microabscess. The cells were CD3 positive and CD20 negative. Biopsy from forearm showed an epitheloid granuloma with Langhans giant cells (Fig 3). No acid fast bacilli were seen. Lymph node biopsy from axillary region showed partial loss of architecture with paracortical expansion by proliferation of abnormal lymphoid cells with convoluted nuclei.

Additional Professor, Department of skin & STD, Kasturba Medical College, Manipal 576 119, Karnatakam Inda,
Ph : 08252-71201 Ext. : 2276
Department of Skin & STD; Department of Pathology, Kasturba Medical College, Manipal, Karnataka, India 576 119.*

Bone marrow aspiration revealed 31% atypical lymphocytes with convoluted nuclei, coarse chromatin and a scanty rim of cyto plasm.

A diagnosis of mycosis fungoides stage IV A was established & patient received I.V. cyclophosphamide 1000 mg, vincristine 1.8 mg, adriamycin 70 mg in 500 ml of dextrose normal saline (DNS), once every 3 weeks & oral prednisolone 25 mg qid for 5 days at three weekly intervals for a total of 8 courses. A repeat bone marrow aspiration showed only 7% atypical lymphocytes following which she received 7 courses of 10 mg oral Chlorambucil monthly for two weeks. She is presently being followed up & her skin lesions continue to wax and wane and the verrucae over the limbs have been resistant to multiple sittings of cryotherapy, electro-dessication, phenol cautery and levamisole.

DISCUSSION

Cutaneous T cell lymphomas (CTCL) generally spare the marrow until the disease is far advanced. Salhany et al[1] found marrow involvement in approximately 20% of patients with CTCL, many of them having an advanced skin stage and decreased survival rate. At initial staging bone marrow involvement has been found in 2-10% of patients with mycosis fungoides [2,3]. Graham et al[4] demon strated that atypical bone marrow lymphoid aggregates portends a bad prognosis.

Our patient had only papular lesions with bone marrow, lymph nodes & peripheral blood showing lymphomatous infiltration. Although visceral involvement is rare in early skin stage patients,[5] Graham et al[4] demonstrated cytologically atypical lym phoid aggregates or infiltration of bone marrow in 19% of patients with early stage. Hence they recommend that bone marrow biopsy to be done in early skin stage patients also.

Our patient in addition had photosensitivity and epitheloid cell granulomas associated with Pautrier's microabscess and epider motropism. Granulomatous mycosis fungoides (GMF) is a rare histo logic variant of cutaneous T-cell lymphoma first described by Ackerman in 1970[6] Clinical significance is unknown and it has been suggested that the granulomatous reaction may represent a chronic tissue reaction to an unidentified antigen which may confer some protection against tumour progression.[6,7]

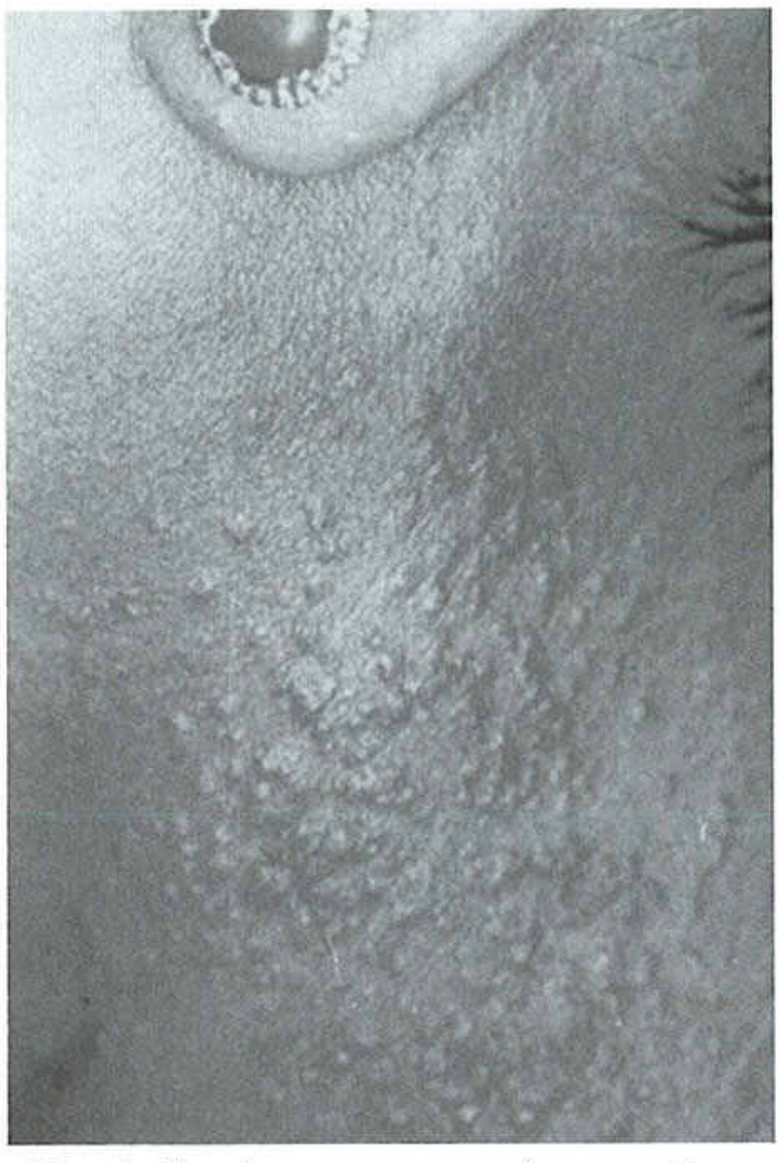

Fig. 1: Erythematous papular eruption over the neck.

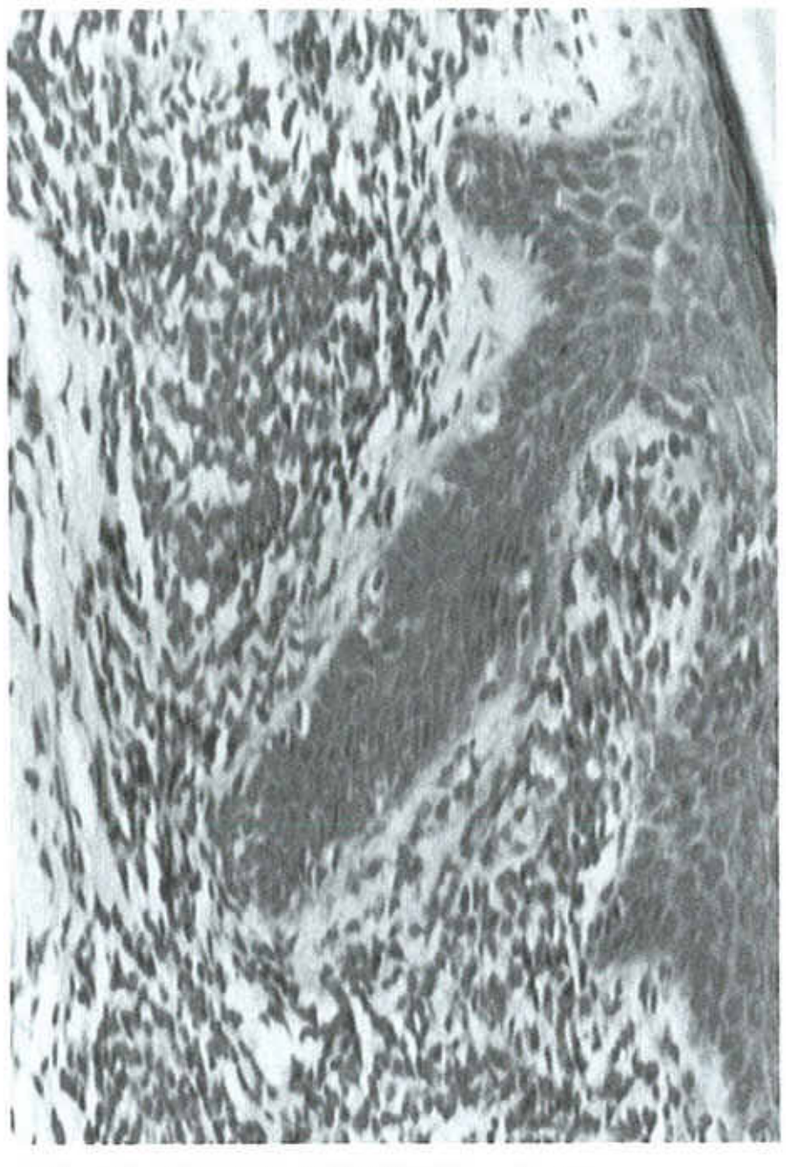

Fig. 2: H & E (20 X) showing epidermotropism and Pautrier's microabscess.

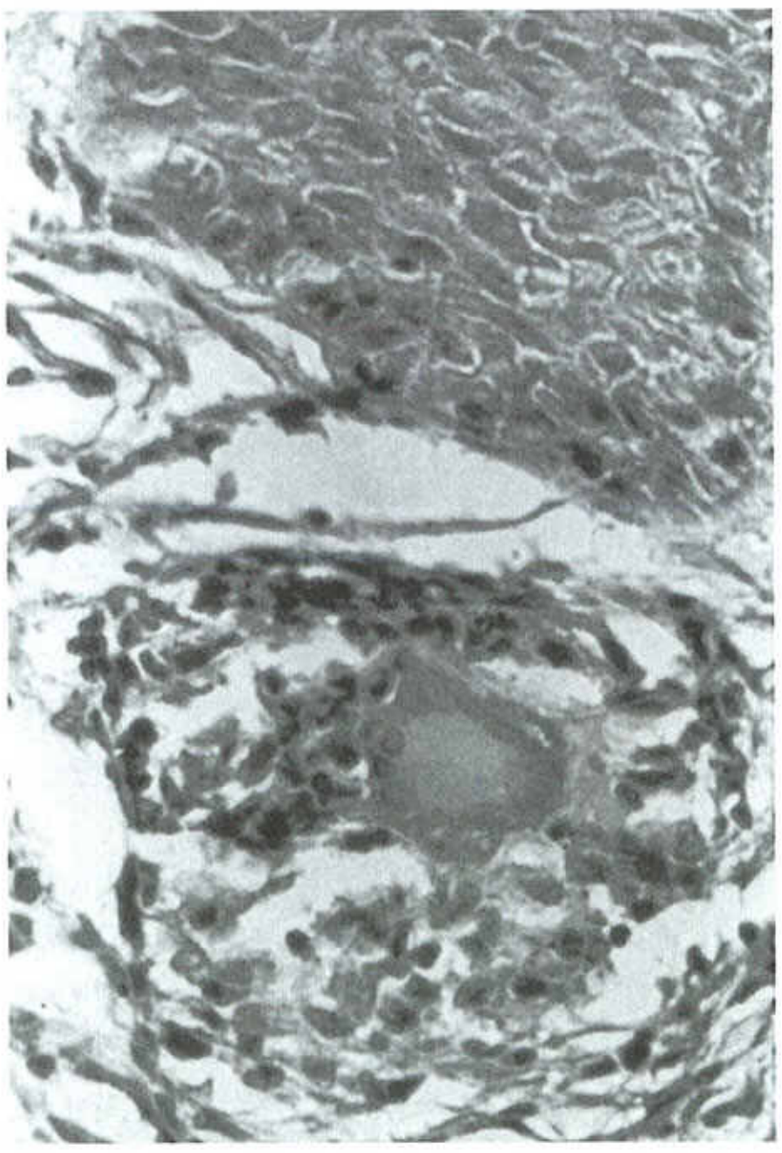

Fig. 3: H & E (20 X) showing epitheloid granuloma with Langhan giant cell.

REFERENCES

1- Salhany K.E, Greer J.P, Cousar J.B. et al. Marrow involvement in Cut T cell lymphoma. A clinico histopathological study of 60 cases. Am J Clin Patho. 1989; 92 (6): 747 - 754.

2- Bunn P.A, Huberman M.S, Whang Peng J et al. Prospective staging evaluation of patients with cutaneous T-celllymphoma: demonstration of high frequency of extracutaneous dissemination. Ann Int Med 1980; 93: 223 - 230.

3- Merlo C.J, Hoppe R.T, Abel E. et al. Extracutaenous mycosis fungoides. Cancer 1987; 60: 397 - 402.

4- Graham S.J, Sharpe R.W, Steinberg S.M. et al. Prognostic implications of a bone marrow histopathological classification system in mycosis fungoides and sezary syndrome. Cancer 1993; 72 (3): 726 - 734.

5- Sausville E.A, Eddy J.L, Makuch R.W. et al. Histopathological staging at initial diagnosis of mycosis fungoids and sezary syndrome. Ann Intern Med 1988; 109: 372 - 382.

6- Ackerman A.B, Flaxman B.A. Granulomatous mycosis fungoides. Br J Dermatol 1970; 82: 397 - 401.

7- Papadavid E, Yu R.C, Bunker C. et al. Tumour progression in a patient with mycosis fungoides. Br J Dermatol 1996; 134: 740 - 743.