

Case Report

Cutaneous T-Cell Lymphoma with Monoclonal Plasmacytosis

Rokuro MIMURA and Yoshitaka MORI

Department of Pathology, Hyogo College of Medicine, Nishinomiya, Japan

Summary

A 75-year-old man with a well-established cutaneous T-cell lymphoma (mycosis fungoides) had a monoclonal gammopathy IgA- λ type. Histologically infiltrate of plasma cells with an IgA- λ type was coincidentally present around atypical lymphocytes in the dermis. Some cases were reported as Sézary's syndrome or mycosis fungoides or cutaneous T-cell lymphoma in association with a monoclonal gammopathy. But proliferation of plasma cells related mycosis fungoides has not been described. This association suggests the production of cytokine by lymphoma cells.

Key words: Cutaneous T-cell lymphoma, mycosis fungoides, monoclonal plasmacytosis, monoclonal gammopathy

Introduction

Cutaneous T-cell lymphomas (mycosis fungoides and Sézary's syndrome) primarily involve the skin. Most of the neoplastic T-cells are typical of CD4-positive helper/inducer T-cells. Some cases were reported as Sézary's syndrome¹⁻⁴⁾ or mycosis fungoides⁵⁻⁸⁾ or cutaneous T-cell lymphoma⁹⁾ in association with a monoclonal gammopathy. Some of these cases complicated myeloma,^{1,2,7)} which showed diffuse proliferation of atypical plasma cells in the bone marrows. But most of them had benign monoclonal gammopathy unrelated to the lymphoma. We presented a patient with mycosis fungoides in association with a monoclonal gammopathy and plasmacytosis. This association lends support to the relation between T-helper-cell malignant proliferation and plasma cell proliferation.

Report of a case

A 66-year-old man presented in 1982 with a itching erythematous rash on the face, and was diagnosed as eczematous dermatitis. In the ensuing 8 years the

patient had a recurrent skin rash. In 1990, erythematous nodules developed on the face and the trunk, and were biopsied. The diagnosis was cutaneous T-cell lymphoma with monoclonal plasmacytosis. There were also palpable lymphadenopathies in the bilateral cervical and axillary regions. Biopsies of the enlarged lymph nodes performed also showed infiltration of neoplastic T-cells and plasma cells.

Bone radiograph revealed no punched out lesion. A computed tomographic scan of his chest and abdomen was performed, which failed to reveal any evidence of mediastinal and retroperitoneal lymphadenopathies.

His laboratory analyses disclosed the following values: red blood cell count, $433 \times 10^4 / \mu\text{l}$; white blood cell count $47 \times 10^2 / \mu\text{l}$, with a differential cell count of 22% band cell forms, 47% segmented forms, 2% eosinophils, 8% monocytes, and 21% lymphocytes; and platelet count $20 \times 10^4 / \mu\text{l}$.

Serum immunoglobulin assay showed IgG of 1510 mg / dl (normal, 750 to 1700); IgA, 1890 mg / dl (normal, 100 to 440); and IgM, 143 mg / dl (normal, 160 to 295). Serum protein electrophoresis levels showed M-protein (IgA- λ), and there was no evidence of Bence-Jones protein in the urine. The titer of anti-HTLV-1 antibody was less than x5. The results of the remainder of the patient's biochemical profile were within normal limits. The patient was treated with combination chemotherapy. However, he died of pneumonia on January in 1991.

Histology.

Several specimens of facial erythematous rash showed features of typical mycosis fungoides: The epidermis showed slight acanthosis and there was slight hyperkeratosis (Fig. 1). In the upper dermis infiltration of small-sized lymphocytes with irregular shaped nuclei was present, and within the epidermis small numbers of atypical, irregular lymphocytes were found (Pautrier's microabscess). In addition to the epidermis, follicular epithelium was involved. In the periphery of the dermal infiltrate plasma cells also were present (Fig. 2).

A biopsy specimen of skin erythematous nodule revealed a massive dermal cellular infiltrate consisting of lymphocytes that were medium-sized possessing irregular nuclei and were surrounded by plasma

Address for Correspondence: Rokuro MIMURA,
Department of Pathology, Hyogo College of Medicine,
Mukogawa-cho 1-1, Nishinomiya 663, Japan

cells(Fig. 3). And a heavily infiltrate of atypical lymphocytes was seen in the epidermis(Fig. 4).

The lymph node biopsy specimen disclosed that the cortical architecture of the lymph node was destroyed and a marked infiltration of large numbers of

pleomorphic medium-sized lymphocytes was seen in the paracortical region(Fig. 5). A proliferation of plasma cells was also present(Fig. 6). The post capillary venules appeared to be accentuated or increased in number, but no features of dermatopathic

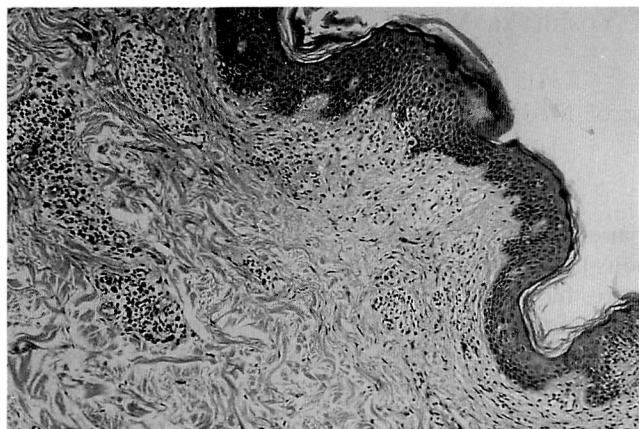


Fig. 1. Skin: The epidermis shows slight hyperkeratosis and mild acanthosis. A slightly perivascular infiltrate is seen in the superficial dermis.

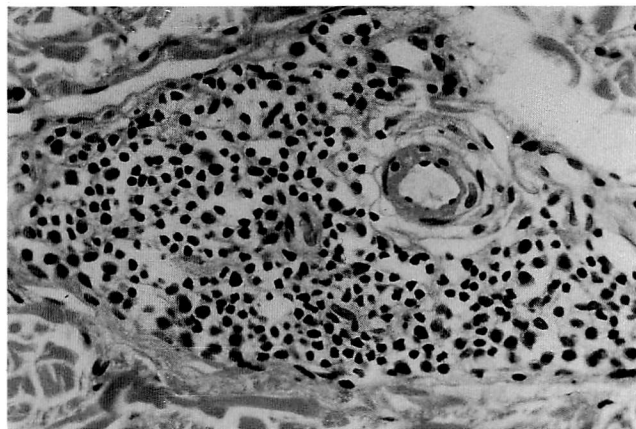


Fig. 2. Skin: Within the superficial dermis is a lymphocytic infiltrate and plasma cells also are found.

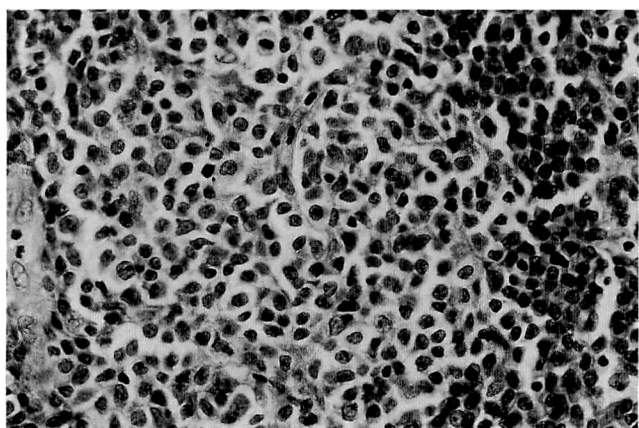


Fig. 3. Skin nodule: In the dermis was occupied with a intense cellular infiltrate, that is composed of atypical lymphocytes and plasma cells.

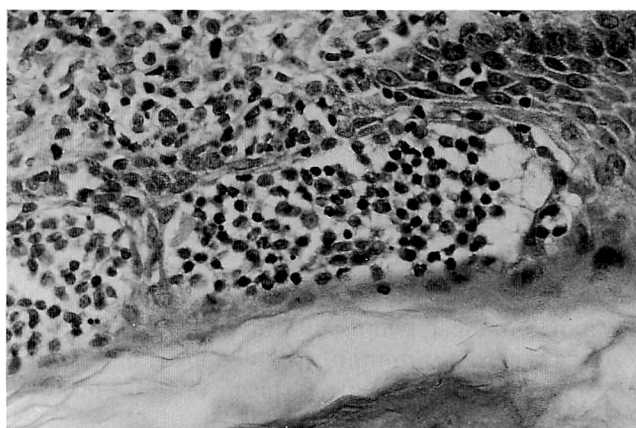


Fig. 4. Skin nodule: Within the epidermis (a hair follicle) are collection of atypical lymphocytes. And they shows nuclear hyperchromatism and irregularity.

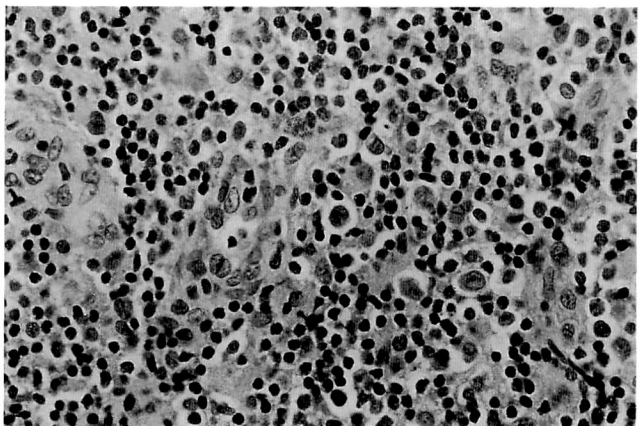


Fig. 5. Lymph node: A infiltration of the paracortical region by pleomorphic lymphocytes.

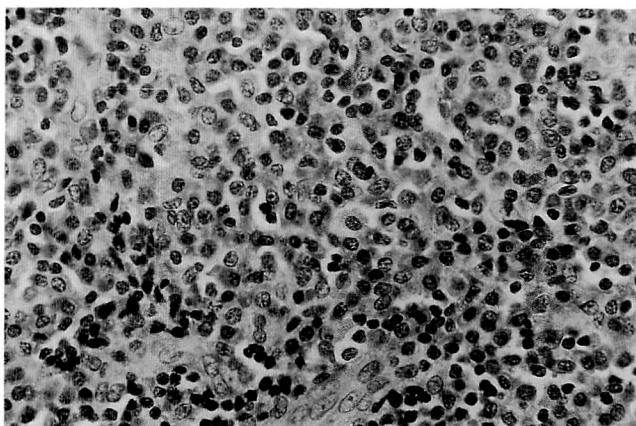


Fig. 6. Lymph node: A infiltration of plasma cells are seen.

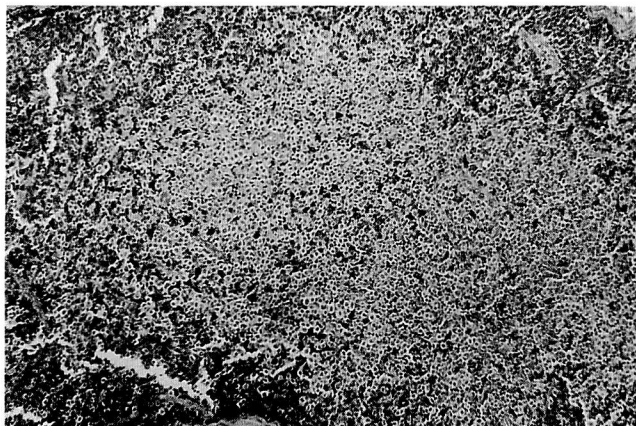


Fig. 7. Immunostain of skin nodule: The proliferative plasma cells are positive with anti-lambda antibody.

lymphadenopathy were seen.

Immunostain performed on skin and lymph node specimens showed that the pleomorphic lymphocytes had T cell marker (UCHL-1) and plasma cells had monoclonal intracytoplasmic immunoglobulin (IgA- λ) (Fig. 7).

Comment

We presented a patient with cutaneous T-cell lymphoma (mycosis fungoides) in association with a monoclonal gammopathy and plasmacytosis. Histologically skin biopsy specimens showed typical features of mycosis fungoides: The epidermis showed slight acanthosis and there was slight hyperkeratosis. In the upper dermis infiltration of small-sized lymphocytes with irregular shaped nuclei was present, and within the epidermis Pautrier's microabscess were found. In addition to the epidermis, follicular epithelium was involved.

The lymph node biopsy specimen disclosed that the cortical architecture of the lymph node was destroyed and a marked infiltration of large numbers of pleomorphic medium-sized lymphocytes was seen in the paracortical region. And the post capillary venules appeared to be accentuated or increased in number, but no features of dermatopathic lymphadenopathy were seen. The histological appearance of the lymph node was in accordance with previous reports in Sézary's syndrome rather than mycosis fungoides.

Characteristic feature in this patient was that infiltrate of plasma cells with the same type of serum immunoglobulin (IgA-lambda) was coincidentally present around atypical T-lymphocytes both in the skin and in the lymph node.

The frequency of monoclonal gammopathy in the general population is related to age and the incidence of monoclonal gammopathy unrelated to the lymphoma has been noted as high as 4.8% over the age of 80

years. The occurrence of monoclonal gammopathy has been previously reported in 17 cases of cutaneous T-cell lymphoma. In three of 17 cases the monoclonal gammopathy was related to myeloma, and one showed atypical plasmacytosis in the bone marrow. But in 13 of 17 cases no proliferation of plasma cell were reported and most of them had benign monoclonal gammopathy unrelated to the lymphoma.

The importance of this interesting association between cutaneous T-cell lymphoma and monoclonal gammopathy is controversial. We presented a patient with mycosis fungoides in association with a monoclonal gammopathy and plasmacytosis. This association lends support to the relation between T-helper-cell malignant proliferation and plasma cell proliferation.

Reference

- 1) Kovary PM, Suter L, Macher E, Niedorf H, Grundmann E, Lukitsch O, et al. Monoclonal gammopathies in Sézary syndrome: A report of four new cases and a review of the literature. *Cancer* 1981; 48: 788-92.
- 2) Venencie PY, Winkelmann PK, Puissant A, Kyle RA. Monoclonal gammopathy in Sézary syndrome: Report of three cases and review of the literature. *Arch Dermatol* 1984; 120: 605-8.
- 3) Shimamura R, Kaneko T, Morioka E, Shibuya T, Ishibashi H, Niho Y. Sézary syndrome associated with hepatocellular carcinoma and monoclonal gammopathy-A case report. *Rinsho Ketsueki* 1989; 30: 1067-73.
- 4) Borish L, Dishuck J, Cox L, Mascali JJ, Williams J, Rosenwasser LJ. Sézary syndrome with elevated serum IgE and hypereosinophilia: Role of dysregulated cytokine production. *J Allergy Clin Immunol* 1993; 92: 123-31.
- 5) Palermo A, Moretti S, Difonzo EM, Fattorossi. Mycosis fungoides with monoclonal gammopathy, hypereosinophilia, and hyper-IgE. *Dermatologica* 1984; 169: 194-6.
- 6) Venencie PY, Winkelmann PK, Friedman SJ, Kyle RA, Puissant A. Monoclonal gammopathy and mycosis fungoides: Report of four cases and review of the literature. *J Am Acad Dermatol* 1984; 11: 576-9.
- 7) Ruffieux C, Stalder M, Tran-Thang C. Mycosis fungoides and monoclonal gammopathy. Fortuitous or causal association? *Dermatologica* 1984; 168: 170-4.
- 8) Lawrence CM, Marks JM, Burrige A, Moss C, Ive FA, McGibbon D, et al. The nature of mycosis fungoides. *Q J Med* 1986; 58: 281-93.
- 9) Joyner MV, Cassuto JP, Dujardin P, Barety M, Duplay H, Audoly P. Cutaneous T-cell lymphoma in association with a monoclonal gammopathy. *Arch Dermatol* 1979; 115: 326-8.